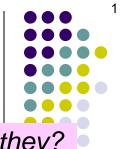
Q

## Ocular-Surface Immunology



The names of these two cell types are easily confused—what are they?

#### This one is:

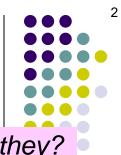
- --A type of dendritic cell residing in the ocular surface epithelium
- --An antigen-presenting cell (APC)
- --Described as the 'immune sentinels of the ocular surface'

cell

#### This one is:

- --A type of giant cell found in granulomas
- --Associated with TB
- --Horseshoe-like arrangement of nuclei

\_\_\_\_cell



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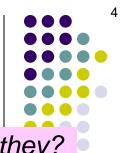
How did dendritic cells come to be called 'dendritic'? Does it have something to do with dendrites on the cornea, eg, as in HSV epitheliopathy?

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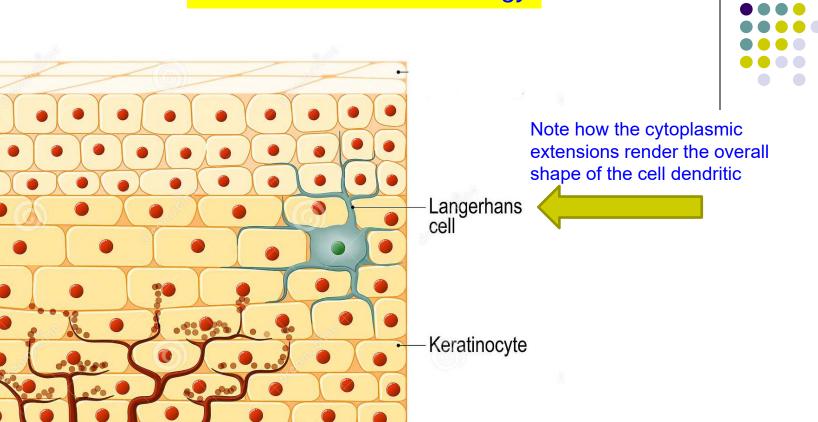
How did dendritic cells come to be called 'dendritic'? Does it have something to do with dendrites on the cornea, eg, as in HSV epitheliopathy?

No, it is because the cells have cytoplasmic extensions jutting out which give the cell the appearance of the dendrite-end of a neuron

-- ASS bolated With 1 b

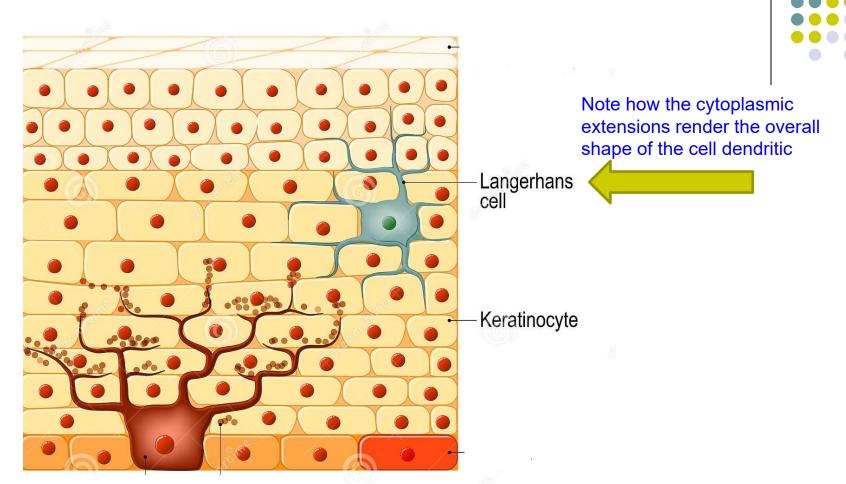
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**An important aside**: The term *dendritic* is also used in the *BCSC* to describe the overall shape of a cell type



Melanocyte

**An important aside**: The term dendritic is also used in the BCSC to describe the overall shape of a melanocyte



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Langerhans cell

Are Langerhans cells found in the conj epi, the corneal epi, or both?

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Regarding the cornea: Are Langerhans cells found anywhere in particular? Yes—they are found at the periphery (they will migrate inward if the central cornea is irritated)

is cell



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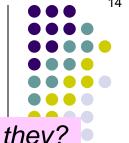
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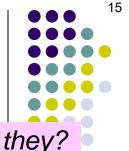
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n <u>the conj epi, the corneal epi</u>

The conj and cornea comprise the ocular surface. In terms of immunology, what is the key difference between their epithelia?





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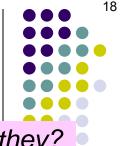
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What does this mean, 'immunologically privileged'?

It means the immune reaction to foreign antigens on the corneal surface is less than that of other tissues (including the conj)



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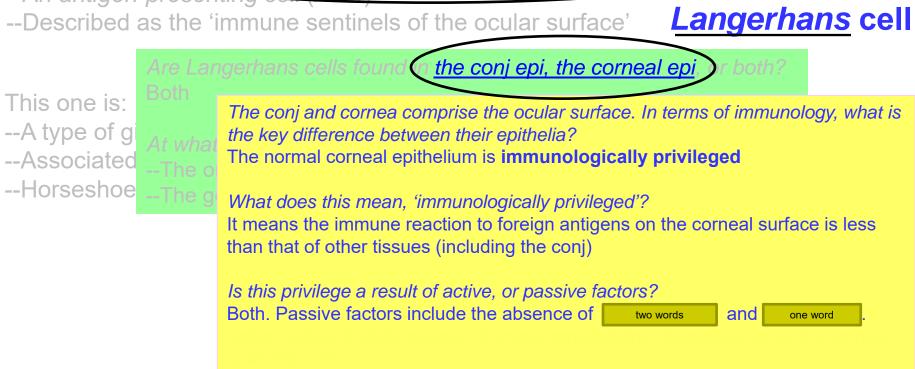
Is this privilege a result of active, or passive factors?



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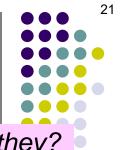
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Both. Passive factors include the absence of blood vessels and lymphatics.





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Active factors include release of immunomodulating chemoagents by the corneal epithelial cells.



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For an organized inflammatory response to qualify as granulomatous, it must contain a specific type of inflammatory cell. What cell type is it?



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What is a giant cell?

It is an aggregate (the formal term is \_\_\_\_\_\_) of histiocytes



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It is an aggregate (the formal term is syncytium) of histiocytes



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What is the histologic hallmark of a giant cell?



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What is a giant cell?

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What is the histologic hallmark of a giant cell?
It is multinucleated—the myriad nuclei of the involved histiocytes are all visible within it



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What type of immune cell aggregates to form a giant cell?



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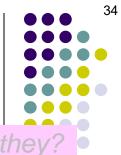
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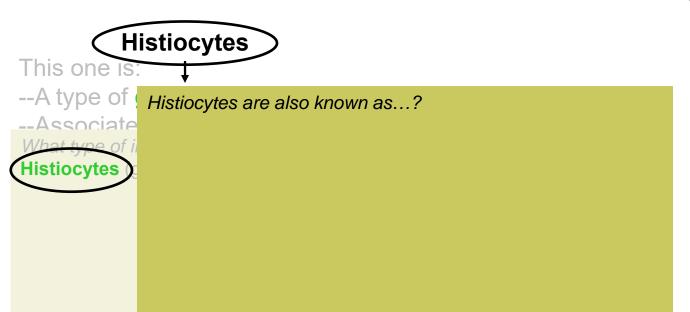


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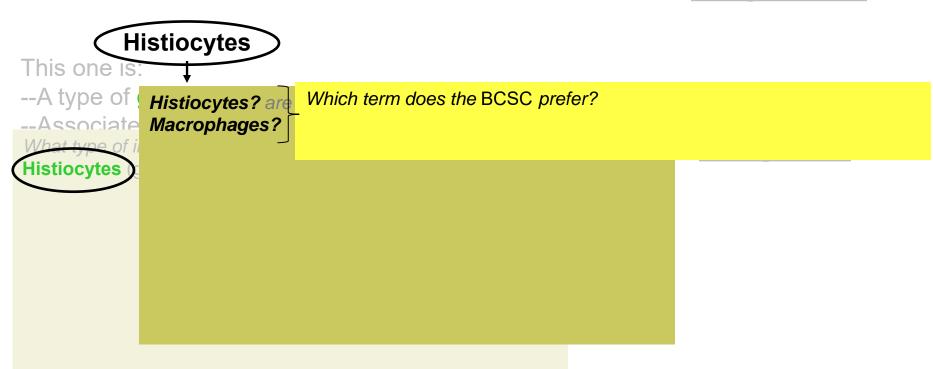




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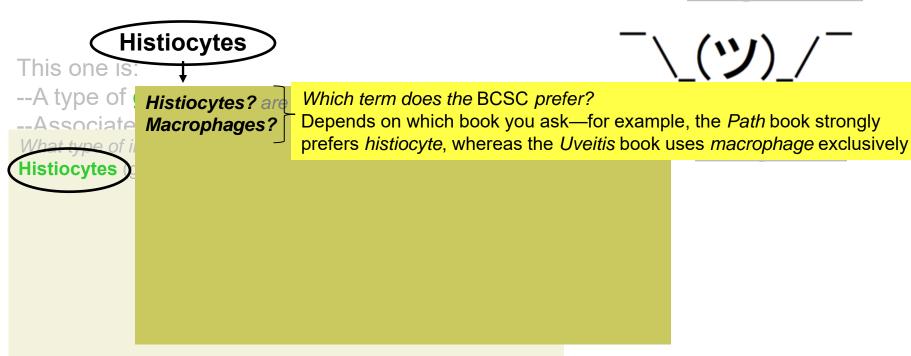




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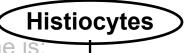


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--Associate Macrophages

Histiocytes

Histiocytes derive from what type of cell?

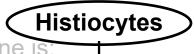


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What does derive mean here? How does a monocyte become a histiocyte?

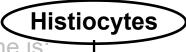


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Histiocytes derive from what type of cell? Monocytes

What does derive mean here? How does a monocyte become a histiocyte?

Monocytes are found within the vasculature. When a monocyte leaves the circulation and settles down within tissue, it becomes (ie, is re-classified) as a histiocyte.



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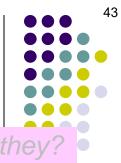
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**Histiocytes** 

--A type of Histiocytes involved in giant-cell formation have a characteristic appearance— --Associate specifically, they look like another cell-type. What cell do they look like? (It is type of an extremely common cell type, one with which you are well-acquainted.)

ans cell





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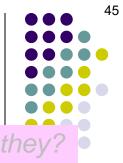
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By what name are these epithelial cell-looking histiocytes known?





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By what name are these epithelial cell-looking histiocytes known? They are known as 'epithelioid histiocytes.' All giant cells are composed of epithelioid histiocytes.



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What type of immune cell aggregates to form a giant cell? Histiocytes (geez, we just said that)

Histiocytes belong to what broad subset of immune cells?

Langhans cell

Next question



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What type of immune cell aggregates to form a giant cell? Histiocytes (geez, we just said that)

Histiocytes belong to what broad subset of immune cells? Myeloid cells



The names of these two cell types are easily confused—what are they?

This one is:

- -- A type of dendritic cell res Myeloid ocular surface epithelium
- --An antigen-presenting cell (APC)
- --Described as the 'immune sentinels of the ocular surface' Langerhans cell

Histiocytes This one is:

--A type of **giant cell** found in granulomas

-- Associated with TR

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What are the other members of the myeloid subset?



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This one is: -- A type of dendritic cell res Myeloid e ocular surface epithelium --An antigen-presenting cell (APC) Langerhans cell --Described as the 'immune sentinels of the ocular surface' PMNs, etc Histiocytes This one is: --A type of **giant cell** found in granulomas -- Associated with TR What type of immune cell aggregates to form a giant cell? Histiocytes (geez, we just said that) Histiocytes belong to what broad subset of immune cells? Myeloid cells What are the other members of the myeloid subset? PMNs, eos, basos, mast cells, and... two words [You definitely know this cell type]



The names of these two cell types are easily confused—what are they?

This one is:

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Histiocytes

PMNs, etc

This one is:

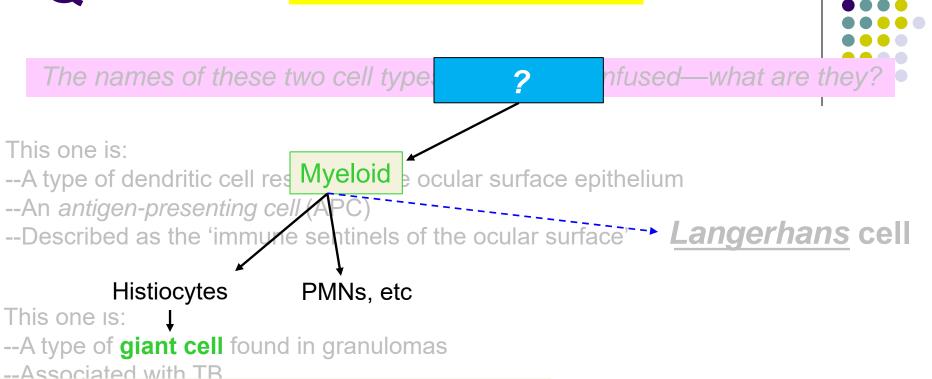
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What are the other members of the myeloid subset? PMNs, eos, basos, mast cells, and...Langerhans cells

Myeloid cells belong to what very broad subset of immune cells?

Langhans cell

52



The names of these two cell type. Leukocytes

nfused—what are they?

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Histiocytes

PMNs, etc

This one is:

--A type of **giant cell** found in granulomas

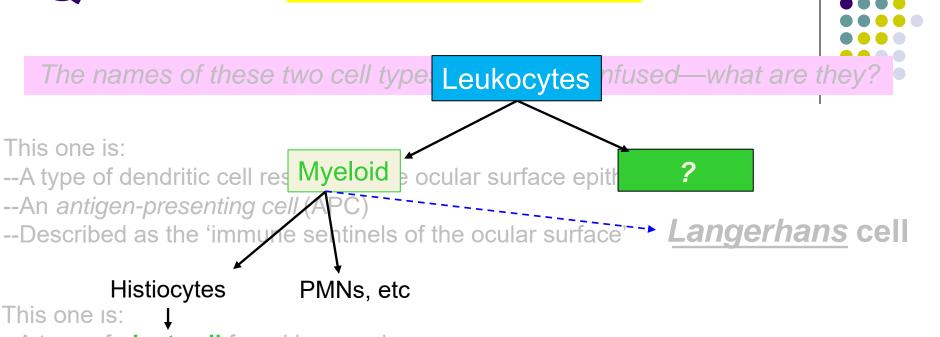
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What are the other members of the myeloid subset? PMNs, eos, basos, mast cells, and...Langerhans cells

Myeloid cells belong to what very broad subset of immune cells? Leukocytes (as in, WBCs)



This one is:

--A type of **giant cell** found in granulomas

-- Associated with TR

What type of immune cell aggregates to form a giant cell? Histiocytes (geez, we just said that)

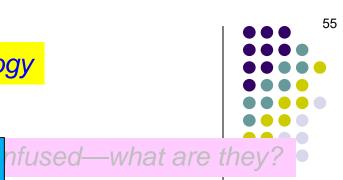
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What are the other members of the myeloid subset? PMNs, eos, basos, mast cells, and...Langerhans cells

Myeloid cells belong to what very broad subset of immune cells? Leukocytes, ie, WBCs

What is the other very broad subset of leukocytes?

54



This one is:

--A type of dendritic cell res Myeloid coular surface epith Lymphoid

The names of these two cell type. Leukocytes

--An antigen-presenting cell (APC)

-- Described as the 'immune sentinels of the ocular surface' Langerhans cell

Histiocytes

PMNs, etc

This one is:

--A type of **giant cell** found in granulomas

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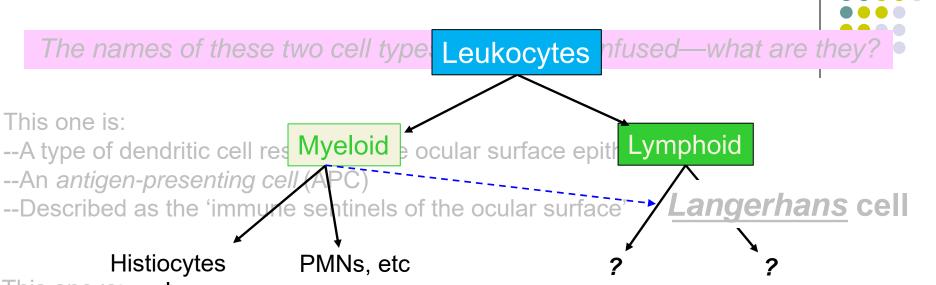
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What is the other very broad subset of leukocytes? Lymphoid cells



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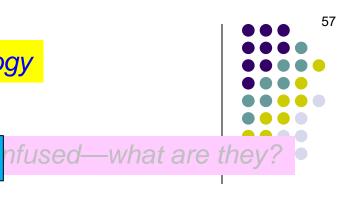
What are the other members of the myeloid subset? PMNs, eos, basos, mast cells, and...Langerhans cells

Myeloid cells belong to what very broad subset of immune cells? Leukocytes, ie, WBCs

What is the other very broad subset of leukocytes? Lymphoid cells

56

What are the two broad subsets of lymphoid cells?



This one is:

--A type of dendritic cell res Myeloid coular surface epith Lymphoid

--An antigen-presenting cell (APC)

--Described as the 'immune sentinels of the ocular surface'

The names of these two cell type. Leukocytes

Histiocytes

PMNs, etc

B cells

T cells

Langerhans cell

This one is:

--A type of **giant cell** found in granulomas

-- Associated with TR

What type of immune cell aggregates to form a giant cell? Histiocytes (geez, we just said that)

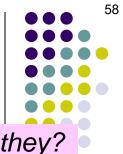
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Myeloid cells belong to what very broad subset of immune cells? Leukocytes, ie, WBCs

What is the other very broad subset of leukocytes? Lymphoid cells

What are the two broad subsets of lymphoid cells? B cells and T cells



The names of these two cell types are easily confused—what are they?

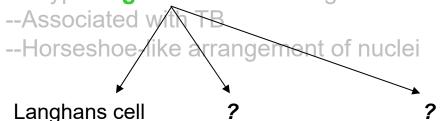
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Langerhans cell

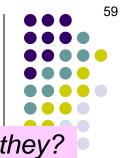
#### This one is:

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Langhans cell

The BCSC covers three types of giant cells, one being the Langhans cell. What are the other two?



# The names of these two cell types are easily confused—what are they?

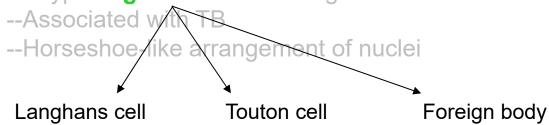
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Langerhans cell

#### This one is:

--A type of **giant cell** found in granulomas



Langhans cell

The BCSC covers three types of giant cells, one being the Langhans cell. What are the other two? **Touton** giant cells, and **foreign-body** giant cells

# thev?

#### The names of these two cell types are easily confused—what are they?

This one is:

- --A type of dendritic cell residing in the ocular surface epithelium
- --An antigen-presenting cell (APC)
- --Described as the 'immune sentinels of the ocular surface'

Langerhans cell

#### This one is:

--A type of **giant cell** found in granulomas

--Associated with TB
--Horseshoe like arrangement of nuclei

Langhans cell Touton cell Foreign

Langhans cell

Foreign body For each type, the way the nuclei are distributed forms a characteristic pattern.

(No question—proceed when ready)

#### The names of these two cell types are easily confused—what are they?

This one is:

- --A type of dendritic cell residing in the ocular surface epithelium
- --An antigen-presenting cell (APC)
- --Described as the 'immune sentinels of the ocular surface'

Langerhans cell

#### This one is:

- --A type of **giant cell** found in granulomas
- -- Associated with TB

--Horseshoe-like arrangement of nuclei

Langhans cell 'Horseshoe'

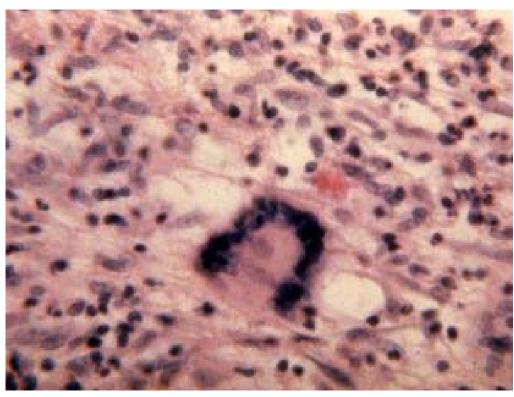
Touton cell

Langhans cell

Foreign body For each type, the way the nuclei are distributed forms a characteristic pattern. For Langhans giant cells, it's a horseshoe.

(No question—proceed when ready)





Note the horseshoe-shaped arrangement of nuclei

Langhans giant cells

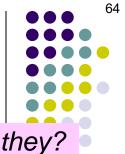


Mnemonic time-out: If you're familiar with the story of 'Clever Hans'—the horse that was famous for seeming to be able to count—you have a ready-made way to remember that Langhans cells have nuclei aggregates that look like horseshoes. (If you're not familiar with the story, I got nothing for ya.)



Note the horseshoe-shaped arrangement of nuclei

Langhans giant cells



# The names of these two cell types are easily confused—what are they?

#### This one is:

- --A type of dendritic cell residing in the ocular surface epithelium
- --An antigen-presenting cell (APC)
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Langerhans cell

# This one is: ---A type of **giant cell** found in granulomas ---Associated where ---Horseshoe like arrangement of nuclei Langhans cell ---Horseshoe' For each type, the way the nuclei are distributed forms a characteristic pattern. For Langhans giant cells, it's a horseshoe. What is the pattern for: --Touton giant cells?



# 65

The names of these two cell types are easily confused—what are they?

#### This one is:

- --A type of dendritic cell residing in the ocular surface epithelium
- --An antigen-presenting cell (APC)
- --Described as the 'immune sentinels of the ocular surface'

'Peripheral ring'

Langerhans cell

#### This one is:

'Horseshoe'

--A type of **giant cell** found in granulomas

-- Associated with --Horseshoe-like arrangement of nuclei Langhans cell Touton cell

Foreign body

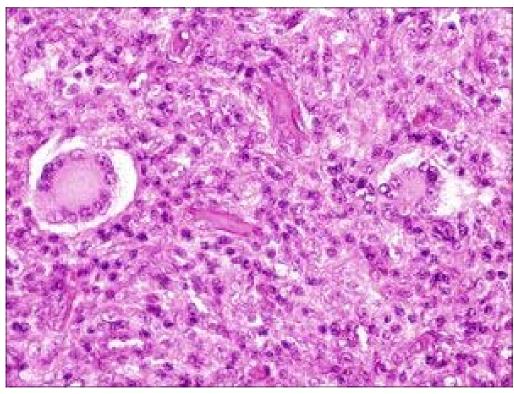
Langhans cell

For each type, the way the nuclei are distributed forms a characteristic pattern. For Langhans giant cells, it's a horseshoe.

What is the pattern for:

--Touton giant cells? 'Peripheral ring'

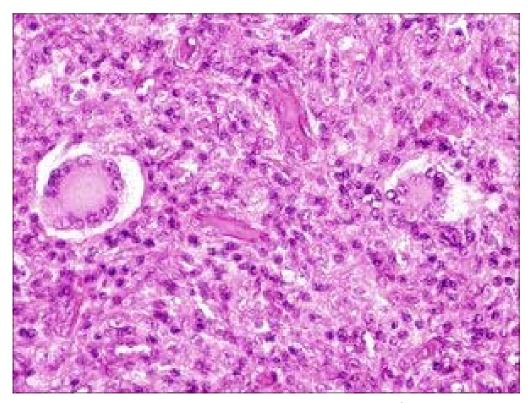




Note the complete peripheral ring of nuclei

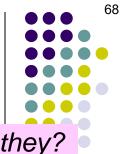
Touton giant cells





Note the complete peripheral ring of nuclei (Note also the clear lipid 'halo,' another feature of this type)

Touton giant cells



#### The names of these two cell types are easily confused—what are they?

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- --A type of dendritic cell residing in the ocular surface epithelium
- --An antigen-presenting cell (APC)
- --Described as the 'immune sentinels of the ocular surface'

Langerhans cell

#### This one is: --A type of **giant cell** found in granulomas -- Associated with Langhans cell --Horseshoe-like arrangement of nuclei Foreign body Langhans cell Touton cell For each type, the way the nuclei are distributed forms a characteristic pattern. L 2 'Horseshoe' 'Peripheral ring' For Langhans giant cells, it's a horseshoe. What is the pattern for: -- Touton giant cells? 'Peripheral ring' --Foreign-body giant cells? Next Q



The names of these two cell types are easily confused—what are they?

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- --Described as the 'immune sentinels of the ocular surface'

'Peripheral ring'

Langerhans cell

#### This one is:

'Horseshoe'

--A type of **giant cell** found in granulomas

-- Associated with --Horseshoe like arrangement of nuclei Foreign body Langhans cell Touton cell ┗ 'Haphazard'

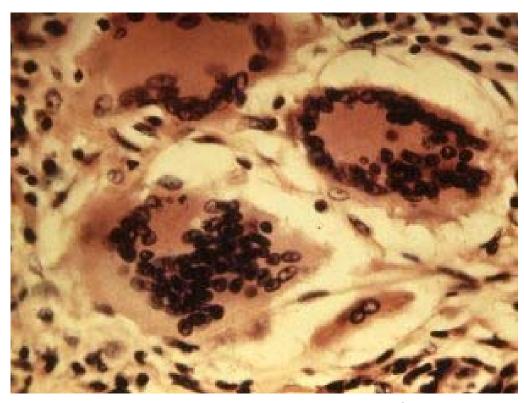
Langhans cell

For each type, the way the nuclei are distributed forms a characteristic pattern. For Langhans giant cells, it's a horseshoe.

What is the pattern for:

- -- Touton giant cells? 'Peripheral ring'
- --Foreign-body giant cells? 'Haphazard'





Note the haphazard arrangement of nuclei

Foreign-body giant cells

# The names of these two cell types are easily confused—what are they?

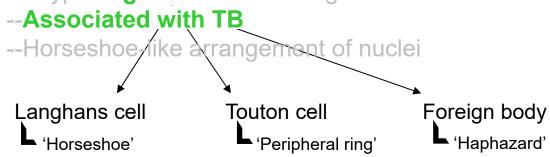
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- --An antigen-presenting cell (APC)
- --Described as the 'immune sentinels of the ocular surface'

Langerhans cell

#### This one is:

--A type of **giant cell** found in granulomas



Langhans cell

Each type has a classic condition with which it's associated.

(No question—proceed when ready)

This part changed—re-read it!

# The names of these two cell types are easily confused—what are they?

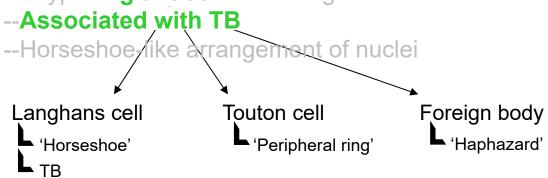
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Langerhans cell

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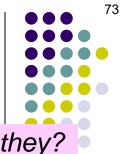


Langhans cell

Each type has a classic condition with which it's associated. For Langhans giant cells, it's infections (eg, TB).

(No question—proceed when ready)

This part changed—re-read it!



## The names of these two cell types are easily confused—what are they?

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Langerhans cell

#### This one is:

--A type of **giant cell** found in granulomas

--Associated with TB
--Horseshoe like arrangement of nuclei

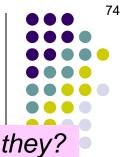
Langhans cell Touton cell Foreign body
'Horseshoe' 'Peripheral ring' 'Haphazard'

Langhans cell

Each type has a classic condition with which it's associated. For Langhans giant cells, it's infections (eg, TB).

What is the condition for:

--Touton giant cells?



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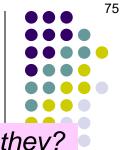
Langhans cell Touton cell Foreign body
'Horseshoe' 'Peripheral ring' 'Haphazard'
TB

Langhans cell

Each type has a classic condition with which it's associated. For Langhans giant cells, it's infections (eg, TB).

What is the condition for:

--Touton giant cells? JXG



## The names of these two cell types are easily confused—what are they?

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Langerhans cell

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-- Associated with TB

--Horseshoe-like arrangement of nuclei

Langhans cell

'Horseshoe'

Touton cell 'Peripheral ring'

Foreign body 'Haphazard'

Each type has a classic condition with which it's associated. For Langhans giant cells, it's infections (eq. TB).

What does JXG stand for?

e condition for:



# The names of these two cell types are easily confused—what are they?

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Langhans cell

Langhans cell 'Horseshoe'

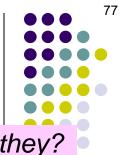
Touton cell 'Peripheral ring' JXG

'Haphazard'

Foreign body Each type has a classic condition with which it's associated. For Langhans giant cells, it's infections (eq. TB).

What does JXG stand for? Juvenile xanthogranuloma

e condition for:



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Langerhans cell

--Foreign-body giant cells?

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Next Q



## The names of these two cell types are easily confused—what are they?

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Langerhans cell

#### This one is:

-- Associated with

--A type of **giant cell** found in granulomas

--Horseshoe like arrangement of nuclei

Langhans cell Touton cell Foreign body

'Horseshoe' 'Peripheral ring' 'Haphazard'

TB

TOUTON CELL FOREIGN BODY

'Haphazard'

FBs

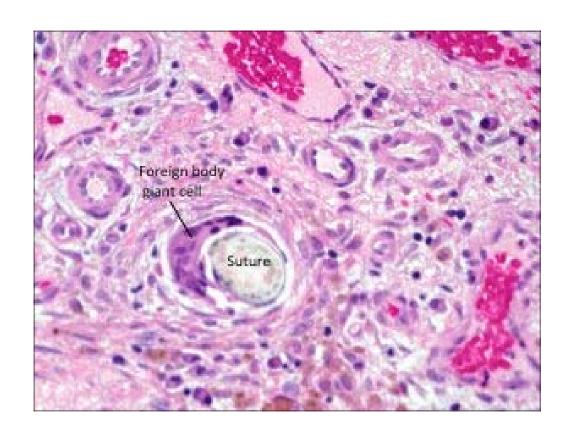
Langhans cell

Each type has a classic condition with which it's associated. For Langhans giant cells, it's infections (eg, TB).

What is the condition for:

- --Touton giant cells? JXG
- --Foreign-body giant cells? Hurr durr, is it foreign bodies?





Foreign-body giant cell in response to a suture



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Langerhans cell

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Langhans cell

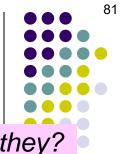
Langhans cell

'Horseshoe'

Touton cell

Foreign body

'Periphe In addition to Touton giant cells, JXG has another classic histologic finding—what is it?



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Langhans cell

Langhans cell 'Horseshoe'

Touton cell

JXG

Foreign body

Periphe In addition to Touton giant cells, JXG has another classic histologic finding—what is it? The presence of macrophages



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Langhans cell

Langhans cell 'Horseshoe'

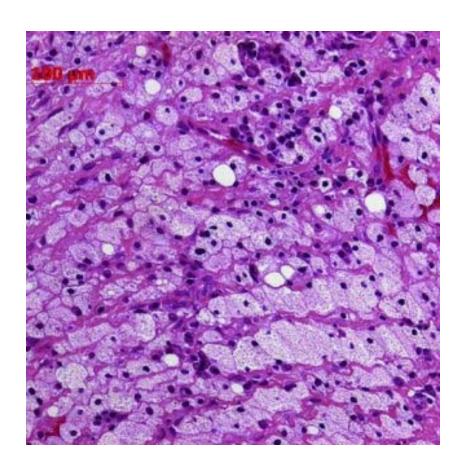
Touton cell

JXG

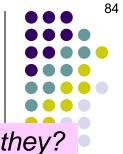
Foreign body

Periphe In addition to Touton giant cells, JXG has another classic histologic finding—what is it? The presence of foamy macrophages





Foamy macrophages



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Touton cell

JXG

Foreign body

Periphe In addition to Touton giant cells, JXG has another classic histologic finding—what is it? The presence of **foamy** macrophages

What gives these cells their foamy appearance?



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Langhans cell

Langhans cell 'Horseshoe'

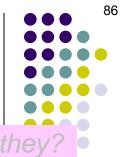
Touton cell

JXG

Foreign body

Periphe In addition to Touton giant cells, JXG has another classic histologic finding—what is it? The presence of **foamy** macrophages

> What gives these cells their foamy appearance? They are full of fat (the term of art is 'lipid laden')



The names of these two cell types are easily confused—what are they?

Switching gears, but still on topic: To what does the term Langerhans cell histiocytosis refer?

**Langerhans** cell **histiocy**tosis



The names of these two cell types are easily confused—what are they?

Switching gears, but still on topic: To what does the term Langerhans cell histiocytosis refer? To a set of proliferative disorders characterized by the accumulation of Langerhans cells

**Langerhans** cell **histiocytosis** 



The names of these two cell types are easily confused—what are they?

Switching gears, but still on topic: To what does the term Langerhans cell histiocytosis refer? To a set of proliferative disorders characterized by the accumulation of Langerhans cells

By what name were these conditions formerly known as? (TBH, this name is not really 'former'—it's still used a lot.)

**Langerhans** cell **histiocytosis** 

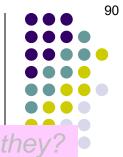


The names of these two cell types are easily confused—what are they?

Switching gears, but still on topic: To what does the term Langerhans cell histiocytosis refer? To a set of proliferative disorders characterized by the accumulation of Langerhans cells

By what name were these conditions formerly known as? (TBH, this name is not really 'former'—it's still used a lot.) 'Histiocytosis X'

**Langerhans** cell **histiocytosis** 



The names of these two cell types are easily confused—what are they?

Switching gears, but still on topic: To what does the term Langerhans cell histiocytosis refer? To a set of proliferative disorders characterized by the accumulation of Langerhans cells

Are these disorders common, or rare?

**Langerhans** cell **histiocytosis** 



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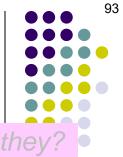
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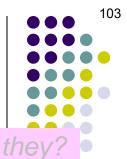
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Langerhans cell histiocytosis

two words



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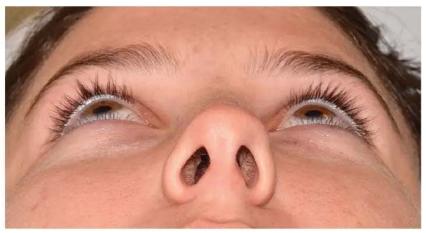
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**Langerhans** cell **histiocytosis** 

<u>ns</u> cell



18-year-old woman with ptosis of the right upper eyelid, fullness (arrow), and inferior globe displacement



Worm's-eye view demonstrates 2 mm of proptosis of the right eye

Langerhans cell histiocytosis



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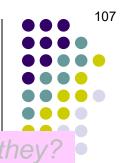
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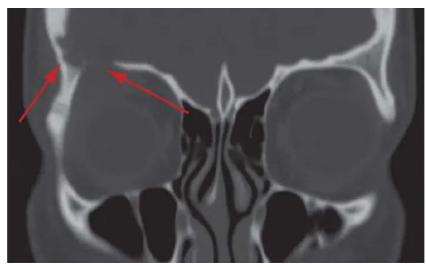
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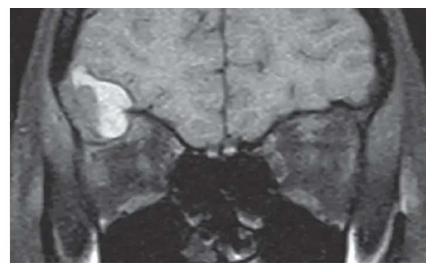
A soft-tissue mass (usually superotemporal) with associated lytic bone lesions

Langerhans cell histiocytosis



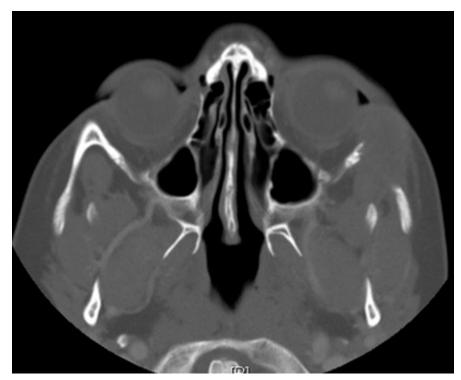


Coronal CT scan shows lytic erosion of the superotemporal orbital bone (arrows)



Coronal MRI scan shows the soft tissue with an intracranial and intraorbital component

Langerhans cell histiocytosis



CT with soft tissue mass-like lesion adjacent to lateral wall of the left orbit. Note the erosion of portions of the lateral orbital wall and zygoma.



110

Lytic skull lesion and large mass in maxillary sinus and orbit in a 4 yo with Langerhans cell histiocytosis

Langerhans cell histiocytosis



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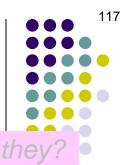
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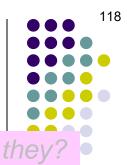
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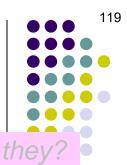
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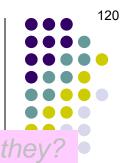
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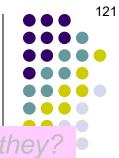
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- --A type of dendritic cell residing in the ocular surface epithelium
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**Langerhans** cell histiocytosis

#### This one is:

- --A type of giant cell found in granulomas
- --Associated with TB
  --Horseshoe-like arrangement of nuclei

Langhans cell

'Horseshoe'

Touton cell

'Peripheral ring'

'Haphazard'

FRe

Langhans cell



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Langhans cell (Horseshoe)

Switching gears part deaux: Is non-Langerhans cell histiocytosis a thing?

Peripheral ring

'Haphazard'



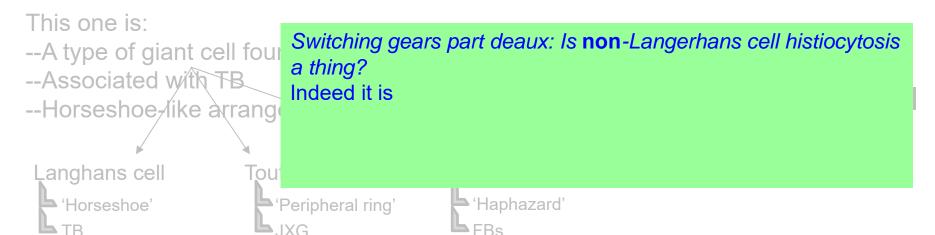
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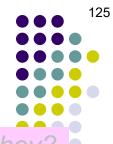
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Indeed it is

What is the classic non-LCH condition? (Hint: It's on this page.)

Peripheral ring'

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•'Peripheral ring'

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JXG: Skin papules



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What is the classic ophthalmic presentation?

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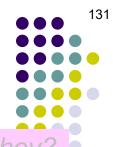
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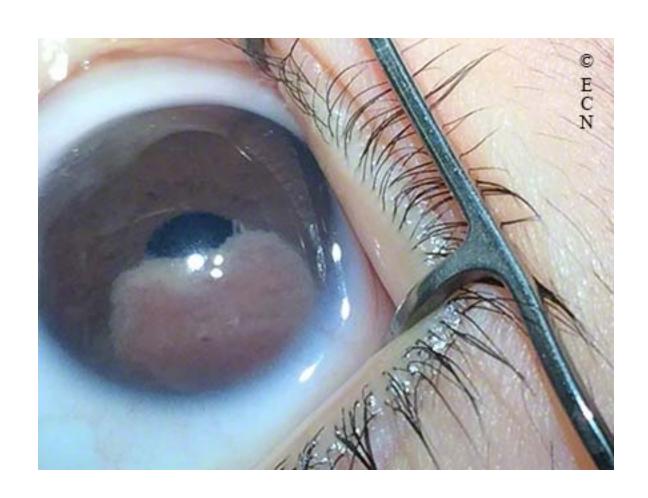
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How does JXG usually present? (It's not ophthalmic.)

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What is the classic ophthalmic presentation? An iris mass/nodule





JXG: Iris mass





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At what age does it present?



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- -- Associated with TB
- --Horseshoe-like arrangement of

Langhans cell

'Horseshoe'

Touton cell

'Peripheral

How does JXG usually present? (It's not ophthalmic.)

As orangish skin papules

What is the classic ophthalmic presentation?

An iris mass/nodule

At what age does it present?

The majority before age and almost all by





The names of these two cell types are easily confused—what are they?

# **Histiocytoses**

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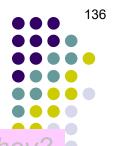
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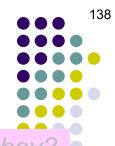
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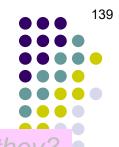
At what age does it present?

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When JXG iris nodules are present, are they uni-, or bilateral? Unilateral

In what three ways are the iris nodules clinically significant?





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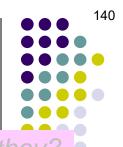
Unilateral

In what three ways are the iris nodules clinically significant?

1) They are prone to spontaneous bleeding, with subsequent hyphema and secondary glaucoma

2) They are in the DDx as a 'masquerade syndrome' in peds uveitis
3) If enough nodules are present, two words will result





The names of these two cell types are easily confused—what are they?

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Duton cell
Peripheral
(JXG)

At what age does it present? The majority before age 1 year, and almost all by 2

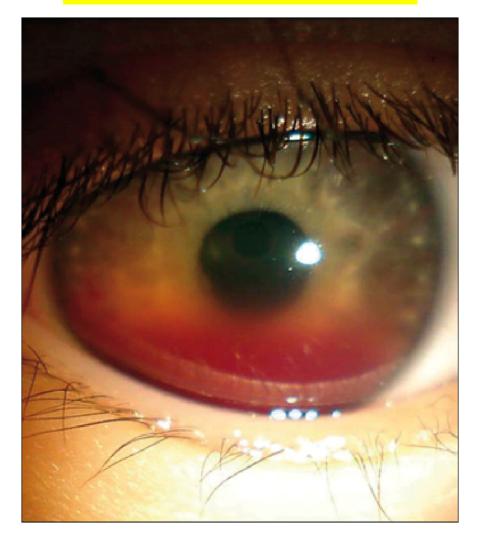
When JXG iris nodules are present, are they uni-, or bilateral?

Unilateral

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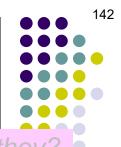
2) They are in the DDx as a 'masquerade syndrome' in peds uveitis 3) If enough nodules are present, heterochromia iridis will result











The names of these two cell types are easily confused—what are they?

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(JXG)

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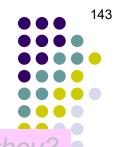
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What is the natural history of JXG?





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Guidentell
Supplementation
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What is the natural history of JXG? It is self-limited, usually resolving by age





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Duton cell
Peripheral
(JXG)

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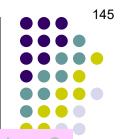
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Langerhans cell histiocytosis

#### This one is:

- --A type of giant cell found in granulomas
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- --Horseshoe-like arrangement of nuclei

Langhans cell

Langhane call

The Plastics book mentions four other non-Langerhans histiocytoses. See if you can name them from a brief description...

buton cell

JXG

Peripheral ring'

Foreign body 'Haphazard'

(No question—proceed when ready)



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## histiocytosis

Langerhans cell histiocytosis

#### This one is:

- --A type of giant cell four
- -- Associated with TB
- --Horseshoe-like arrai

Langhane coll

The Plastics book mentions four other non-Langerhans histiocytoses. See if you can name them from a brief description...

1) While all four present with skin lesions, this one is notorious for those lesions ulcerating:

- 2)
- 3)
- 4)

**L** ?

G **L**FBs



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# Langerhans cell histiocytosis

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2)

3)

4)

► JXG

► Necrobiotic xanthogranuloma





Necrobiotic xanthogranuloma



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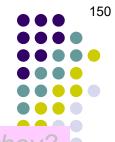
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- 2) This one presents with adult-onset asthma and periocular lesions:
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**►** ?

► JXG ► FBs

• Necrobiotic xanthogranuloma



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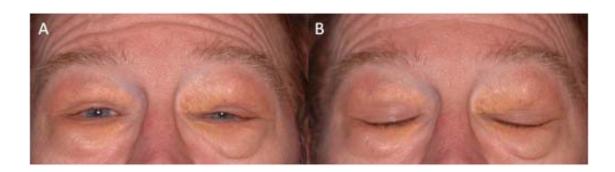
Adult-onset asthma with periocular xanthogranuloma

4)

**L** FBs

Necrobiotic xanthogranuloma

Adult-onset asthma with periocular xanthogranuloma





151



It can look like xanthelasma on steroids (well, not on steroids; you know what I mean)

Adult-onset asthma with periocular xanthogranuloma



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#### Adult-onset asthma with periocular xanthogranuloma

3) This one presents with isolated xanthogranulomas—no systemic findings whatsoever:

Langhane coll

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XG FBs

► Necrobiotic xanthogranuloma

Adult-onset asthma with periocular xanthogranuloma



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Langhane coll

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JXG FBs

► Necrobiotic xanthogranuloma

Adult-onset asthma with periocular xanthogranuloma

■ Adult-onset xanthogranuloma





Adult-onset xanthogranuloma



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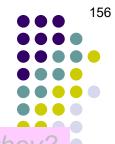
JXG FBs

Necrobiotic xanthogranuloma

■ Adult-onset asthma with periocular xanthogranuloma
■ Adult-onset xanthogranuloma

(No question—proceed when ready)

These two look alike—
it's the asthma that
differentiates them!



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#### Adult-onset asthma with periocular xanthogranuloma

- 3) This one presents with isolated xanthogranulomas—no systemic findings whatsoever: Adult-onset xanthogranuloma
- 4) This one is **awful**—progressive fibrosclerosis of the internal organs and but body cavities; often fatal. If that's not bad enough, intraconal involvement is common, and can result in blindness:

► Necrobiotic xanthogranuloma

Adult-onset asthma with periocular xanthogranuloma

■Adult-onset xanthogranuloma

. ?



cell types are easily confused—what are they?

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- 4) This one is **awful**—progressive fibrosclerosis of the internal organs and but body cavities; often fatal. If that's not bad enough, intraconal involvement is common, and can result in blindness: **Erdheim-Chester dz**

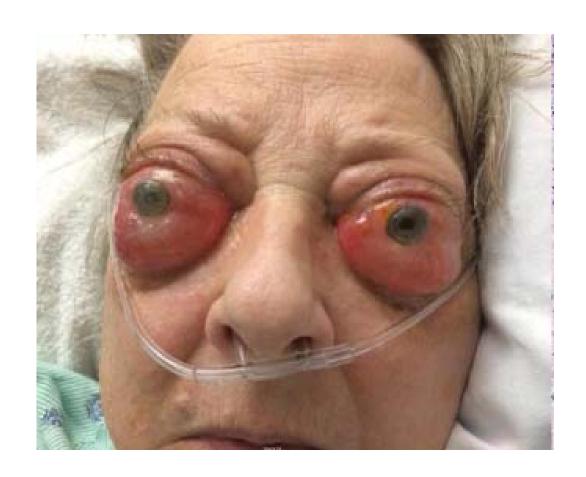
► Necrobiotic xanthogranuloma

Adult-onset asthma with periocular xanthogranuloma

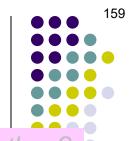
Adult-onset xanthogranuloma

Erdheim-Chester dz





Erdheim-Chester disease



The names of these two cell types are easily confused—what are they

# Histiocytoses

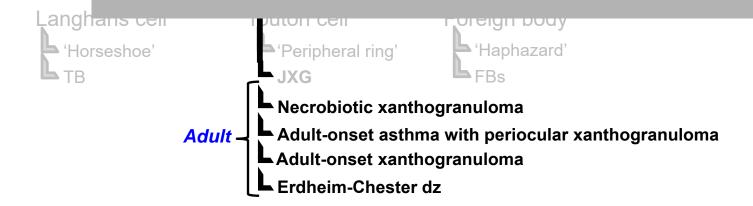
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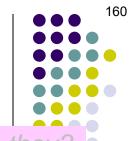
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histiocytosis

#### This one is:

- --A type of giant cell found in granulomas
- Note that all of the non-Langerhans cell histiocytoses present in adulthood





The names of these two cell types are easily confused—what are they

# Histiocytoses

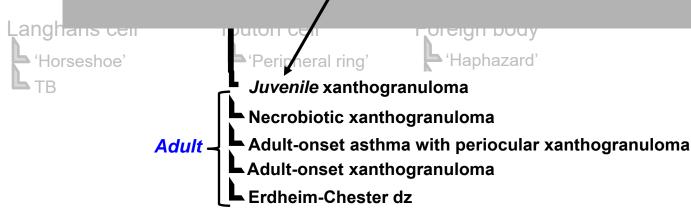
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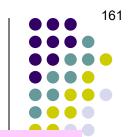
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# histiocytosis

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Note that all of the non-Langerhans cell histiocytoses present in adulthood (except the one with juvenile in the name, duh).





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Langerhans cell histiocytosis

#### This one is:

--A type of giant cell found in granulomas

Note that all of the non-Langerhans cell histiocytoses present in adulthood (except the one with *juvenile* in the name, duh). For this reason, together they are called the *adult xanthogranulomatous diseases*—AXGs for short.

Horseshoe'

'Horseshoe'

Juvenile xanthogranuloma

Adult xanthogranulomatous diseases

Adult-onset asthma with periocular xanthogranuloma
Adult-onset xanthogranuloma

**L** Erdheim-Chester dz

Necrobiotic xanthogranuloma



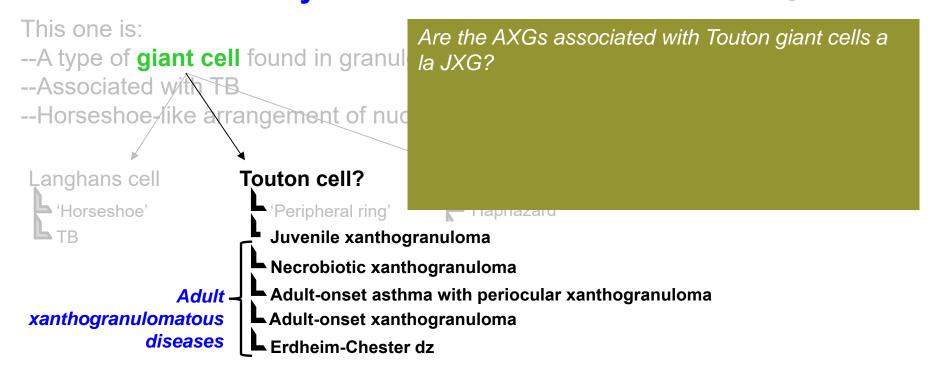
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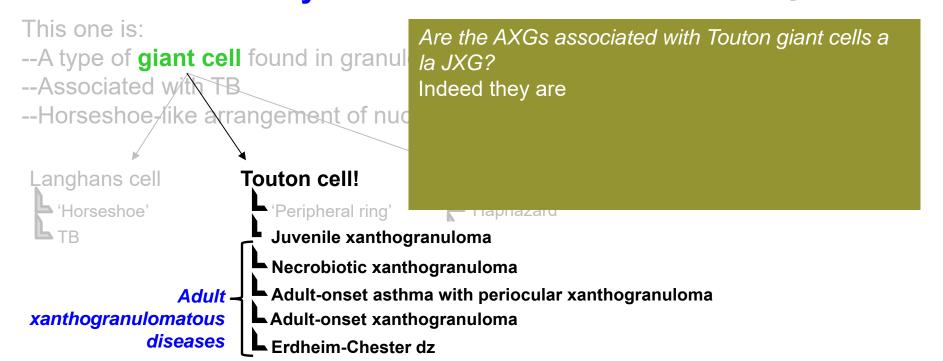
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## **Histiocytoses**

#### This one is:

xanthogranulomatous

diseases

-- A type of dendritic cell residing in the ocular surface epithelium

▲ Adult-onset xanthogranuloma

Erdheim-Chester dz

- --An antigen-presenting cell (APC)
- --Described as thon-Langerhans cell ular surface' histiocytosis

# Langerhans cell histiocytosis

#### This one is: Are the AXGs associated with Touton giant cells a --A type of **giant cell** found in granul la JXG? -- Associated with TB Indeed they are --Horseshoe-like arrangement of nuc Are the AXGs associated with 'foamy macrophages' a la JXG? Ļanghans cell Touton cell (Horseshoe) 'Peripheral ring' Juvenile xanthogranuloma ■ Necrobiotic xanthogranuloma Adult-onset asthma with periocular xanthogranuloma



The names of these two cell types are easily confused—what are they?

## **Histiocytoses**

#### This one is:

xanthogranulomatous

diseases

-- A type of dendritic cell residing in the ocular surface epithelium

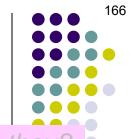
▲ Adult-onset xanthogranuloma

Erdheim-Chester dz

- --An antigen-presenting cell (APC)
- --Described as thon-Langerhans cell ular surface' histiocytosis

# **Langerhans** cell histiocytosis

#### This one is: Are the AXGs associated with Touton giant cells a --A type of **giant cell** found in granul la JXG? -- Associated with TB Indeed they are --Horseshoe-like arrangement of nuc Are the AXGs associated with 'foamy macrophages' a la JXG? Ļanghans cell Touton cell Them too, yes (Horseshoe) 'Peripheral ring' Juvenile xanthogranuloma ■ Necrobiotic xanthogranuloma Adult-onset asthma with periocular xanthogranuloma



The names of these two cell types are easily confused—what are they

# Histiocytoses

#### This one is:

xanthogranulomatous

diseases

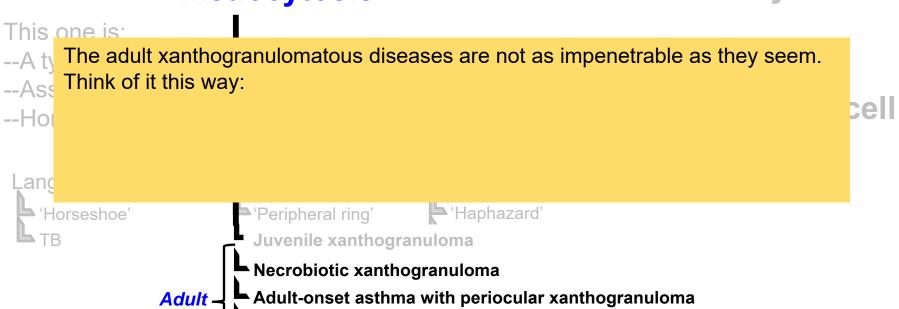
-- A type of dendritic cell residing in the ocular surface epithelium

▲ Adult-onset xanthogranuloma

Erdheim-Chester dz

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- --Described as thon-Langerhans cell ular surface' Langerhans cell histiocytosis

histiocytosis





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-- A type of dendritic of

--An antigen-presenting centar C

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Langerhans cell histiocytosis

what are they

This one is: The adult xanthogranulomatous diseases are not as impenetrable as they seem. Think of it this way: On one of our tests, I would anticipate they will present as --Ass 'super-xanthelasmas.' If that's the end of the story, it's just adult-onset cell xanthogranuloma. \_and Haphazard' (Horseshoe) •'Peripheral ring' Juvenile xanthogranuloma Necrobiotic xanthogranuloma

xanthogranulomatous diseases Adult-onset asthma with periocular xanthogranuloma

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Langerhans cell histiocytosis

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Think of it this way: On one of our tests, I would anticipate they will present as --Ass 'super-xanthelasmas.' If that's the end of the story, it's just adult-onset

xanthogranuloma. If there's a history of adult-onset asthma, it's that one.

•'Peripheral ring'

Haphazard'

Juvenile xanthogranuloma

Necrobiotic xanthogranuloma

Adult-onset asthma with periocular xanthogranuloma

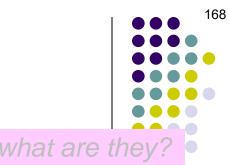
Adult-onset xanthogranuloma

Erdheim-Chester dz

\_and

(Horseshoe)

xanthogranulomatous diseases



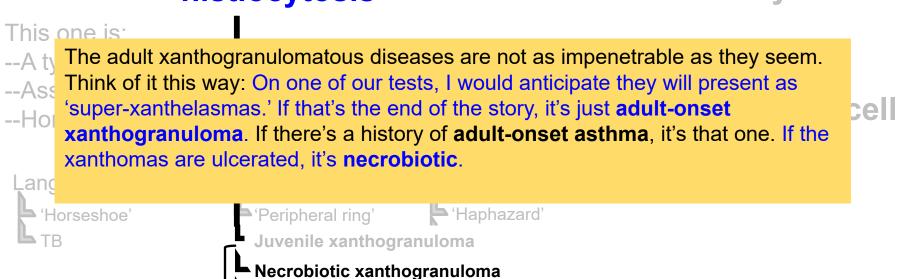
cell



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Langerhans cell histiocytosis

169



xanthogranulomatous diseases

The names of the

-- A type of dendritic of

This one is:

Adult-onset asthma with periocular xanthogranuloma **▲** Adult-onset xanthogranuloma Erdheim-Chester dz



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Langerhans cell histiocytosis

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This one is: The adult xanthogranulomatous diseases are not as impenetrable as they seem. --Ass Think of it this way: On one of our tests, I would anticipate they will present as 'super-xanthelasmas.' If that's the end of the story, it's just adult-onset xanthogranuloma. If there's a history of adult-onset asthma, it's that one. If the xanthomas are ulcerated, it's **necrobiotic**. Finally, if there is significant intraorbital Lang involvement and/or the pt is very ill, it's Erdheim-Chester.

Haphazard'

Juvenile xanthogranuloma Necrobiotic xanthogranuloma Adult-onset asthma with periocular xanthogranuloma xanthogranulomatous **▲** Adult-onset xanthogranuloma diseases **L** Erdheim-Chester dz

'Peripheral ring'



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# **Histiocytoses**

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#### This one is:

- --A type of giant cell found in granulomas
- -- Associated with TB
- --Horseshoe-like arrangement of nuclei

#### Remind me—what are the three subtypes of LCH?

'Horseshoe'

'Peripheral ring'

FBs

'Haphazard

Necrobiotic xanthogranuloma

Adult-onset asthma with periocular xanthogranuloma

Adult-onset xanthogranuloma

**L** Erdheim-Chester dz

Langerhans cell histiocytosis

Langhans c

?\_



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# Langerhans cell histiocytosis

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## Langhans cell

Unifocal eosinophilic granuloma of bone

### Remind me—what are the three subtypes of LCH?

'Horseshoe'

'Peripheral ring'

FBs

Multifocal eosinophilic granuloma of bone (Hand-Schüller-Christian disease)

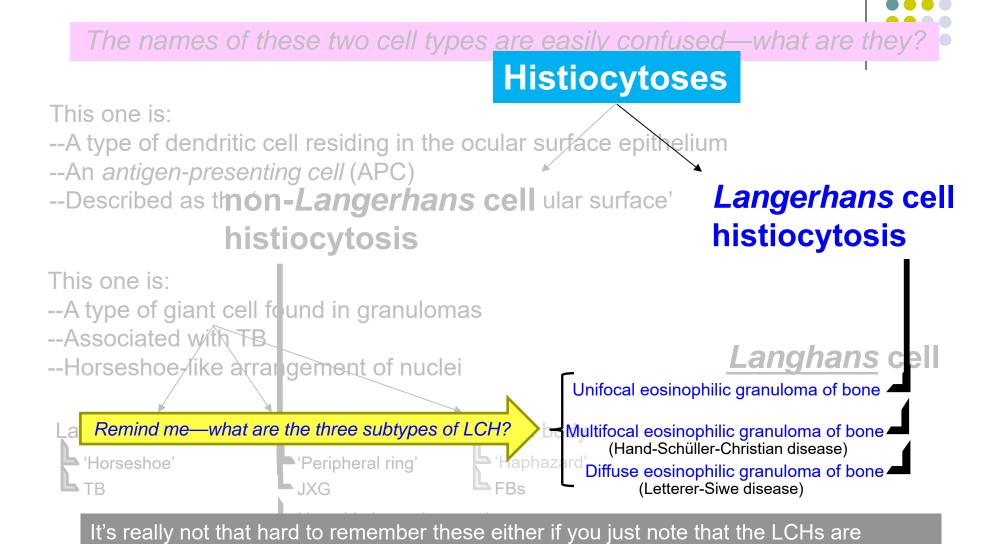
Diffuse eosinophilic granuloma of bone (Letterer-Siwe disease)

Necrobiotic xanthogranuloma

Adult-onset asthma with periocular xanthogranuloma

Adult-onset xanthogranuloma

Erdheim-Chester dz



'eosinophilic granulomas of bone' that present in unifocal, multifocal, or diffuse fashion.

It's the eponyms that are tough.



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# Langhans cell

Unifocal eosinophilic granuloma of bone

Multifocal eosinophilic granuloma of bone

Diffuse eosinophilic granuloma of bone

Remind me—what are the three subtypes of LCH?

'Horseshoe'

Peripheral ring

JXG

It's really not that hard to remember these either if you just note that the LCHs are 'eosinophilic granulomas of bone' that present in unifocal, multifocal, or diffuse fashion. It's the eponyms that are tough. But as the BCSC downplays these, they can be ignored.



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