Typical Optic Neuritis

Optic Neuropathy

Lots of ways to divvy these up…
Optic Neuropathy

Typical Optic Neuritis

- Inflamatory
- Noninflammatory

Lots of ways to divvy these up... this is just one.
Optic Neuropathy

Inflammatory

Noninflammatory

Lots of ways to divvy these up…

Typical Optic Neuritis
Optic Neuropathy

- Inflammatory
  - Typical (demyelinating)
  - Atypical

- Noninflammatory

*Typical Optic Neuritis*

Lots of ways to divvy these up... this is just one.
Typical Optic Neuritis

Optic Neuropathy

Inflammatory

Typical (demyelinating)

Atypical

Noninflammatory
Optic Neuritis

Optic Neuropathy

Inflammatory

Typical (demyelinating)

Atypical

Infectious

Immune

Noninflammatory
Typical Optic Neuritis

Optic Neuropathy

Inflammatory

Typical (demyelinating)

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Infectious

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Noninflammatory
Optic Neuropathy

Inflammatory
- Typical (demyelinating)
  - Infectious
  - Immune

Atypical

Noninflammatory
- Ischemic
- Compressive
- Toxic/nutritional
- Congenital/hereditary
- Traumatic

Typical Optic Neuritis
What is far-and-away the most common type of optic neuropathy?

*Hint*…
What is far-and-away the most common type of optic neuropathy?

Hint...It’s not listed on this slide!

Hint...
**What is far-and-away the most common type of optic neuropathy?**

*Hint…It’s not listed on this slide!

*Hint…It’s so common, it gets its own ophthalmic subspecialty!

It’s…
Typical Optic Neuritis

Optic Neuropathy

Inflammatory
- Typical (demyelinating)
  - Infectious
- Atypical
  - Immune

Noninflammatory
- Ischemic
- Compressive
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- Traumatic

What is far-and-away the most common type of optic neuropathy? Hint…It’s not listed on this slide! Hint…It’s so common, it gets its own ophthalmic subspecialty! It’s…Glaucoma (don’t forget—glaucoma is an optic neuropathy!)
What exam finding is the sine qua non of unilateral or asymmetric bilateral optic neuropathy?
What exam finding is the sine qua non of unilateral or asymmetric bilateral optic neuropathy?
A relative afferent pupillary defect (RAPD)
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A relative afferent pupillary defect (RAPD)

What should you do if a presumptive unilateral/asymmetric bilateral ON pt doesn’t have an RAPD?
What exam finding is the sine qua non of unilateral or asymmetric bilateral optic neuropathy?
A relative afferent pupillary defect (RAPD)

What should you do if a presumptive unilateral/asymmetric bilateral ON pt doesn’t have an RAPD?
You should question the diagnosis
What functional abnormalities are likely to be found in a pt with an optic neuropathy?

-- Decreased
-- Abnormal
-- Impaired
What functional abnormalities are likely to be found in a pt with an optic neuropathy?
- Decreased central acuity
- Abnormal visual fields
- Impaired color vision
Optic Neuropathy

Inflammatory

Typical (demyelinating)

Atypical

Infectious

Immune

Noninflammatory

- Ischemic
- Compressive
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- Congenital/hereditary
- Traumatic

Name 3 infectious causes of atypical ON:
1)
2)
3)

(There are many others, of course)
Optic Neuropathy

**Inflammatory**
- Typical (demyelinating)
- **Infectious**
- Atypical

**Noninflammatory**
- Ischemic
- Compressive
- Toxic/nutritional
- Congenital/hereditary
- Traumatic

*Name 3 infectious causes of atypical ON:*
1) Lues (syphilis)
2) TB
3) Lyme

(There are many others, of course)
Optic Neuropathy

Inflammatory

Typical (demyelinating)

Atypical

Infectious

Immune

Noninflammatory

Ischemic

Compressive

Toxic/nutritional

Congenital/hereditary

Traumatic

Name 3 immune-related causes of atypical ON:
1)
2)
3)

(There are many others, of course)
Optic Neuropathy

Inflammatory
- Typical (demyelinating)
- Infectious

Atypical
- Immune

Noninflammatory
- Ischemic
- Compressive
- Toxic/nutritional
- Congenital/hereditary
- Traumatic

Typical Optic Neuritis

Name 3 immune-related causes of atypical ON:
1) Sarcoid
2) SLE
3) Wegener’s

(There are many others, of course)
Typical Optic Neuritis

Optic Neuropathy

Inflammatory

Typical (demyelinating)

Who is the typical ‘typical optic neuropathy’ patient?

Noninflammatory

Infectious

Immune

Ischemic

Compressive

Toxic/nutritional

Congenital/hereditary

Traumatic
Typical Optic Neuritis

Optic Neuropathy

Inflammatory

Typical (demyelinating)

Typical optic neuropathy patient?
A female age 15 - 45

Noninflammatory

Infectious

Immune

Ischemic

Compressive

Toxic/nutritional

Congenital/hereditary

Traumatic
Optic Neuropathy

Inflammatory

Typical (demyelinating)

Who is the typical ‘typical optic neuropathy’ patient?
A female age 15 - 45

What should you do if a presumptive demyelinating ON pt doesn’t fit this profile?

Typical Optic Neuritis

Noninflammatory

Ischemic

Traumatic

Immune

Compressive

Toxic/nutritional

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Typical Optic Neuritis

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Who is the typical ‘typical optic neuropathy’ patient?
A female age 15 - 45

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Optic Neuropathy

Inflammatory

- Typical (demyelinating)

Noninflammatory

- Ischemic
- Traumatic
- Congenital/hereditary
- Toxic/nutritional
- Immune
- Infectious

Who is the typical ‘typical optic neuropathy’ patient?
A female age 15 - 45

What percentage of typical/demyelinating ON pts are female?
About 75%
Typical Optic Neuropathy

Inflammatory

Typical (demyelinating)

Who is the typical ‘typical optic neuropathy’ patient?
A female age 15 - 45

What percentage of typical/demyelinating ON pts are female? About 75%
Optic Neuropathy

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With what disease is demyelinating optic neuropathy associated?

Multiple sclerosis (MS)

No, but the lifetime risk of developing MS is high.
With what disease is demyelinating optic neuropathy associated? Multiple sclerosis (MS)
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With what disease is demyelinating optic neuropathy associated?
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Is it always accompanied or followed by MS?
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What is the pattern of vision loss in typical optic neuritis?

Unilateral vision loss developing over a period of a few days, with spontaneous recovery beginning a week or two later

How profound is the vision loss?

The vision can be anywhere from 20/20 to NLP; however, most are in the 20/40 – 20/200 range

What is the long-term visual prognosis?

Very good—about 95% will be 20/40 or better at one year

What should you do if spontaneous recovery fails to occur?

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What percent of typical optic neuritis pts present with bilateral vision loss?

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What percent of typical optic neuritis pts present with bilateral vision loss?

Less than 1% of adult optic neuritis pts present bilaterally

There is a subpopulation of optic neuritis pts for whom bilateral presentation is somewhat more common—what is that population?

Children
What is the pattern of vision loss in typical optic neuritis?

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**Typical Optic Neuritis**

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What is the pattern of vision loss in typical optic neuritis?

**Bilateral** vision loss developing over a period of a few days, with spontaneous recovery beginning a week or two later

What diagnosis should be considered if optic neuritis presents bilaterally and/or fails to recover spontaneously?

What should you do if spontaneous recovery fails to occur?

You should question the diagnosis

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**Typical Optic Neuritis**

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**What diagnosis should be considered if optic neuritis presents bilaterally and/or fails to recover spontaneously?**

- Neuromyelitis optica (NMO), aka Devic’s disease.
Optic Neuropathy

NMO involves two specific disease processes occurring together. One is optic neuritis; what is the other?

Typical Optic Neuritis

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Neuromyelitis optica (NMO), aka Devic’s disease

What diagnosis should be considered if optic neuritis presents bilaterally and/or fails to recover spontaneously?

Transverse myelitis

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Typical (demyelinating) optic neuritis?
Typical Optic Neuritis

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How does transverse myelitis present clinically?
As a symmetric para- or quadriparesis, often with sensory loss (note that symmetric deficits are distinctly uncommon in MS).
Typical Optic Neuritis

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Transverse Myelitis

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Optic Neuropathy

OPTIC NEUROPATHY

Inflammatory
Noninflammatory

Typical (demyelinating)
Atypical

Infectious
Immune
Ischemic
Compressive
Toxic/nutritional
Congenital/hereditary
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Neuromyelitis optica (NMO) involves two specific disease processes occurring together.

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How does transverse myelitis present clinically?
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How does transverse myelitis present radiologically?
As longitudinally extensive spinal cord lesions.

How long are we talking about?
2-3 vertebral segments or so.
**Typical Optic Neuritis**

**Optic Neuropathy**

*Inflammatory Noninflammatory*

Typical (demyelinating)

Atypical

Infectious Immune

Ischemic

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Toxic/nutritional

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**Neuromyelitis optica (NMO), aka Devic’s disease**

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Indeed it is! Further, optic nerve lesions in NMO tend to be more posterior than those found in typical optic neuritis.

Anterior vs posterior.
Typical Optic Neuritis

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Typical Optic Neuritis

How does transverse myelitis present radiologically?
As longitudinally extensive spinal cord lesions

How long are we talking about?
2-3 vertebral segments or so

How does transverse myelitis present clinically?
As a symmetric para- or quadripareisis, often with sensory loss (note that symmetric deficits are distinctly uncommon in MS)

What is the long-term visual prognosis?
Very good—about 95% will be 20/40 or better at one year

What should you do if spontaneous recovery fails to occur?
You should question the diagnosis
Optic Neuropathy

**Typical Optic Neuritis**

What is the pattern of vision loss in typical optic neuritis?

Bilateral vision loss developing over a period of a few days, with spontaneous recovery beginning a week or two later.

How profound is the vision loss?

The vision can be anywhere from 20/20 to NLP; however, most are in the 20/40 – 20/200 range.

What is the long-term visual prognosis?

Very good—about 95% will be 20/40 or better at one year.

What should you do if spontaneous recovery fails to occur?

You should question the diagnosis.

Transverse Myelitis

NMO involves two specific disease processes occurring together. One is optic neuritis; what is the other?

Transverse myelitis.

How does transverse myelitis present radiologically?

As longitudinally extensive spinal cord lesions.

How long are we talking about?

2-3 vertebral segments or so.

How does transverse myelitis present clinically?

As a symmetric para- or quadriplegia, often with sensory loss (note that symmetric deficits are distinctly uncommon in MS).

So, the spinal cord lesions in NMO are longitudinally extensive... Perchance, is the same true of the optic nerve lesions in NMO?

Indeed it is! Further, optic nerve lesions in NMO tend to be more posterior than those found in typical optic neuritis.

How posterior are we talking about here?

They can extend to the optic chiasm, which is almost unheard of in typical optic neuritis.

What is the long-term visual prognosis?

Very good—about 95% will be 20/40 or better at one year.

What should you do if spontaneous recovery fails to occur?

You should question the diagnosis.
Optic Neuropathy

Typical Optic Neuritis

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Neuromyelitis optica (NMO), aka Devic’s disease

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What diagnosis should be considered if optic neuritis presents bilaterally and/or fails to recover spontaneously?
Neuromyelitis optica (NMO), aka Devic’s disease.

NMO involves two specific disease processes occurring together.
One is optic neuritis; what is the other?
Transverse myelitis. NMO is primarily a disease of ‘the optic nerves and spinal cord’ (although importantly, other aspects of the CNS can be affected—more shortly).
Optic Neuropathy

Typical Optic Neuritis

**NMO involves two specific disease processes occurring together.**

One is optic neuritis; what is the other?

**Transverse myelitis.** NMO is primarily a disease of ‘the optic nerves and spinal cord’ (although importantly, **other aspects of the CNS can be affected—more shortly**)

Time for the ‘more shortly.’ When NMO affects ‘other aspects of the CNS,’ one of three classic (but not pathognomonic!) ‘intractable’ presentations may arise. What are they?

--Intractable...
--Intractable...
--Intractable...

---

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What should you do if spontaneous recovery fails to occur?

You should question the diagnosis

Neuromyelitis optica (NMO), aka Devic’s disease

---

Infectious Immune

Ischemic

Compressive

Toxic/nutritional

Congenital/hereditary

Traumatic
Typical Optic Neuritis

Optic Neuropathy

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Time for the 'more shortly.' When NMO affects 'other aspects of the CNS,' one of three classic (but not pathognomonic!) 'intractable' presentations may arise. What are they?

--Intractable…nausea and vomiting
--Intractable…hiccups
--Intractable…daytime somnolence

Typical optic neuritis?

- Bilateral vision loss developing over a period of a few days, with spontaneous recovery beginning a week or two later

How profound is the vision loss?

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Typical Optic Neuritis

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What should you do if spontaneous recovery fails to occur? You should question the diagnosis.

Make a mental note of these—they’re very important! They will clue you in to the fact you’re dealing with NMO rather than typical optic neuritis.


**Optic Neuropathy**

---

**Typical Optic Neuritis**

Does it really matter that much whether a pt has NMO vs typical optic neuritis?

Time for the "more shortly." When NMO affects "other aspects of the CNS," one of three classic (but not pathognomonic!) "intractable" presentations may arise. What are they?

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Optic Neuropathy

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- Intractable…hiccups
- Intractable…daytime somnolence

Make a mental note of these–they’re very important! They will clue you in to the fact you’re dealing with NMO rather than typical optic neuritis.

Does it really matter that much whether a pt has NMO vs typical optic neuritis? Very much so, for two reasons:

1)  
2)

What should you do if spontaneous recovery fails to occur?

- You should question the diagnosis

Very good—about 95% will be 20/40 or better at one year

Very much so, for two reasons:

1) The treatment regimens for the two diseases are different
2) By correctly identifying and treating NMO, you might preclude the dreaded sequelae of the disease, those being paralysis or even death owing to the transverse myelitis (untreated NMO carries a very high risk of both)
Typical Optic Neuritis

Optic Neuropathy

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NMO rather than typical optic neuritis.

What should you do if spontaneous recovery fails to occur?
You should question the diagnosis.
Typical Optic Neuritis

In general terms, what is the key difference in the treatment regimens?

1) The treatment regimens for the two diseases are different
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What should you do if spontaneous recovery fails to occur?
You should question the diagnosis

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Very much so, for two reasons:
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2) By correctly identifying and treating NMO, you might preclude the dreaded sequelae of the disease, those being paralysis or even death owing to the transverse myelitis (untreated NMO carries a very high risk of both)
**Typical Optic Neuritis**

*In general terms, what is the key difference in the treatment regimens?*
Typical optic neuritis is treated with **immuno modulatory** meds, whereas NMO is treated with **immuno suppressive** regimens.

**The treatment regimens for the two diseases are different**
1) By correctly identifying and treating NMO, you might preclude the dreaded sequelae of the disease, those being *paralysis* or even *death* owing to the transverse myelitis (untreated NMO carries a very high risk of both).

**Time for the ‘more shortly.’** When NMO affects ‘other aspects’ of the CNS, ‘one of three classic (but not pathognomonic!) ‘intractable’ presentations may arise. What are they?
--Intractable…nausea and vomiting
--Intractable…hiccups
--Intractable…daytime somnolence

They will clue you in to the fact you’re dealing with NMO rather than typical optic neuritis.

**What should you do if spontaneous recovery fails to occur?**
You should question the diagnosis.
**Typical Optic Neuritis**

In general terms, what is the key difference in the treatment regimens? Typical optic neuritis is treated with immunomodulatory meds, whereas NMO is treated with immunosuppressive regimens.

The treatment regimens for the two diseases are different:

1) By correctly identifying and treating NMO, you might preclude the dreaded sequelae of the disease, those being paralysis or even death owing to the transverse myelitis (untreated NMO carries a very high risk of both).

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You should question the diagnosis if spontaneous recovery fails to occur.

NMO is a variant of MS?
**Typical Optic Neuritis**

*In general terms, what is the key difference in the treatment regimens?*
Typical optic neuritis is treated with *immunomodulatory* meds, whereas NMO is treated with *immunosuppressive* regimens.

*So, NMO doesn’t respond to immunomodulatory meds?*
No, it responds all right—*it gets worse*.

*The treatment regimens for the two diseases are different*
1) By correctly identifying and treating NMO, you might preclude the dreaded sequelae of the disease, those being *paralysis* or even *death* owing to the transverse myelitis (untreated NMO carries a very high risk of both).

*Should be considered if optic neuritis presents bilaterally and/or fails to recover spontaneously?*
Neuromyelitis optica (NMO), aka Devic’s disease.

*What should you do if spontaneous recovery fails to occur?*
You should question the diagnosis.

*Time for the ‘more shortly.’ When NMO affects ‘other aspects of the CNS,’ one of three classic (but not pathognomonic!) ‘intractable’ presentations may arise. What are they?*
--- Intractable…*nausea and vomiting*  
--- Intractable…*hiccups*  
--- Intractable…*daytime somnolence*

*Make a mental note of these—they’re very important!*  
They will clue you in to the fact you’re dealing with NMO rather than typical optic neuritis.

*Very good point. What are the sequelae?*
NMO can lead to paralysis or even death owing to the transverse myelitis (untreated NMO carries a very high risk of both).
**Typical Optic Neuritis**

In general terms, what is the key difference in the treatment regimens? Typical optic neuritis is treated with immunomodulatory meds, whereas NMO is treated with immunosuppressive regimens.

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Oh it responds all right--it gets worse.

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Does it really matter that much whether a pt has NMO vs typical optic neuritis?
Very much so, for two reasons: 1) The treatment regimens for the two diseases are different. 2) By correctly identifying and treating NMO, you might preclude the dreaded sequelae of the disease, those being paralysis or even death owing to the transverse myelitis (untreated NMO carries a very high risk of both).
Optic Neuropathy

Typical (demyelinating)

In a few words, how would you characterize the pathological process in NMO?

What is the pattern of vision loss in typical optic neuritis?
Bilateral vision loss developing over a period of a few days, with spontaneous recovery beginning a week or two later

How profound is the vision loss?
The vision can be anywhere from 20/20 to NLP; however, most are in the 20/40 – 20/200 range

What is the long-term visual prognosis?
Very good—about 95% will be 20/40 or better at one year

What should you do if spontaneous recovery fails to occur?
You should question the diagnosis

What diagnosis should be considered if optic neuritis presents bilaterally and/or fails to recover spontaneously?
Neuromyelitis optica (NMO), aka Devic’s disease

In typical optic neuritis,

Typical Optic Neuritis
**Typical Optic Neuritis**

In a few words, how would you characterize the pathological process in NMO? As an antibody-mediated autoimmune condition.

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What is the target of the antibodies?

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The protein

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What does this protein do? It is the main water channel protein in astroglial cells. Damage to this protein interferes with CNS fluid homeostasis.

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In-Text Reference

^no
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Is lab testing available to detect these antibodies?
Yes, and they form part of the diagnostic criteria for NMO

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**Neuromyelitis optica (NMO), aka Devic’s disease**
Optic Neuropathy

- Inflammatory
  - Typical (demyelinating)
  - Atypical
- Noninflammatory

**Typical Optic Neuritis**

- What is the pattern of vision loss in typical optic neuritis?
  - Bilateral vision loss developing over a period of a few days, with spontaneous recovery beginning a week or two later

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(next question)
Typical Optic Neuritis

Optic Neuropathy

Inflammatory

Typical (demyelinating)

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What is the long-term visual prognosis in NMO? Very good—about 95% will be 20/40 or better at one year. Poor—vision <20/200 in at least one eye is the rule!

What diagnosis should be considered if optic neuritis presents bilaterally and/or fails to recover spontaneously? Neuromyelitis optica (NMO), aka Devic’s disease.

What should you do if spontaneous recovery fails to occur? You should question the diagnosis.
Optic Neuropathy

Inflammatory

Noninflammatory

Typical (demyelinating)

Atypical

Infectious

Immune

Ischemic

Compressive

Toxic/nutritional

Congenital/hereditary

Traumatic

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Neuromyelitis optica (NMO), aka Devic’s disease.

Typical Optic Neuritis

(Warning: Soapbox speech ahead)

Can typical optic neuritis present bilaterally? Yes. Can it be chronic? Yes. But you (speaking to errbody who isn’t a fellowship-trained neuro-oph) shouldn’t make that call, because such cases are zebras, if not unicorns (or even zebracorns). So, don’t select ‘bilateral typical optic neuritis’ or ‘chronic typical optic neuritis’ as answers on the OKAP or WQEs, don’t utter those words when taking the Boards, and most importantly, don’t write them on a pt’s chart!
What is the usual appearance of the ONH in typical optic neuritis?

Benign—no edema is the rule.
Optic Neuropathy

- Inflammatory
  - Typical (demyelinating)
  - Atypical
- Noninflammatory
  - Ischemic
  - Compressive
  - Toxic/nutritional
  - Congenital/hereditary
  - Traumatic

What is the usual appearance of the ONH in typical optic neuritis? Benign—no edema is the rule. Or if present, mild edema w/o heme.
What is the usual appearance of the ONH in typical optic neuritis? Benign—no edema is the rule. Or if present, mild edema w/o heme.

What should you do if a typical optic neuritis pt has florid disc edema?
Typical Optic Neuritis

Optic Neuropathy

Inflammatory

- Typical (demyelinating)

- Atypical

Noninflammatory

- Ischemic
- Compressive
- Toxic/nutritional
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- Traumatic

What is the usual appearance of the ONH in typical optic neuritis? Benign—no edema is the rule. Or if present, mild edema w/o heme.

What should you do if a typical optic neuritis pt has florid disc edema? You should question the diagnosis.
Is typical optic neuritis associated with ocular pain?

Yes. >90% will complain of pain, especially during ocular rotations.

What should you do if a typical optic neuritis pt does not c/o pain?

You should question the diagnosis.
Optic Neuropathy

Inflammatory

- Typical (demyelinating)
- Atypical

Noninflammatory

- Ischemic
- Compressive
- Toxic/nutritional
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Is typical optic neuritis associated with ocular pain? Yes. >90% will complain of pain, especially during ocular rotations.

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- Typical (demyelinating)

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- Traumatic

Is NMO optic neuritis associated with ocular pain?

What about optic neuritis due to NMO—is it associated with pain?

What should you do if a typical optic neuritis pt does not c/o pain?

You should question the diagnosis
Optic Neuropathy

Inflammatory
- Typical (demyelinating)
- Atypical

Noninflammatory
- Ischemic
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- Traumatic

Is NMO optic neuritis associated with ocular pain?

What about optic neuritis due to NMO--is it associated with pain?
Yes, but much less so--only 1/3 will complain of ocular pain.

You should question the diagnosis.
Optic Neuropathy

Inflammatory

Typical (demyelinating)

Atypical

Infectious

Immune

**Typical Optic Neuritis**

With respect to treatment, what is the key difference between typical and atypical ON?

In atypical ON, treatment can influence final visual outcome; whereas in typical ON, treatment has no effect on final visual outcome.

Huh? What about the Optic Neuritis Treatment Trial (ONTT)? Didn’t it show a benefit of IV steroids (and a deleterious effect of PO steroids)?

True and true. But the effect of IV steroids was to 1) hasten visual recovery and 2) delay onset of MS; the effect of PO steroids was to increase the risk of recurrent optic neuritis.

Neither treatment had any effect on final visual outcome, or on the likelihood of developing MS.
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On the other hand, most infectious and immune processes can be either cured or controlled via proper treatment!
If a pt presents with typical optic neuritis, what sort of workup should be done?

MRI brain and orbits, with contrast. That's it.

What is the purpose of the MRI?

To look for white-matter changes, the presence of which increases the likelihood of developing MS.

Noninflammatory

- Ischemic
- Compressive
- Toxic/nutritional
- Congenital/hereditary

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...no white matter changes on MRI:
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With regard to the 25% of ON patients who went on to develop MS despite having no white-matter changes, which of the following characteristics were associated with an increased risk of developing MS?

- Severe ONH edema?
- Lack of pain?
- VA = NLP?
- Macular exudates?

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In general terms, what sort of drug is Avonex?

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