

The Singular Case of the OD With Double Vision

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Kim Lee* spends her working week performing eye exams and answering her patients' queries. Recently, however, the 49-year-old Asian optometrist started questioning her own vision—she had noticed progressive vertical diplopia and right-sided proptosis. She started keeping meticulous records of her vertical deviation and became increasingly concerned as it progressed from 6 prism diopters to 35 prism diopters in the three months since she first noticed the problem. At this point in time, she came to us.

We Get a Look

Ms. Lee denied any pain, loss of vision, fever, fatigue or weight loss. Her ocular history was significant for persistent dry eye treated with punctal plugs and cyclosporine (Restasis). She also had a history of dry mouth.

On exam at presentation, Ms. Lee's best-corrected visual acuity was 20/20 in both eyes with reactive pupils and no relative afferent pupillary defect. Her intraocular pressures were within normal limits. Hertel measurements

were 23 mm on the right and 15 mm on the left. In addition, hypoglobus and complete restriction of supraduction were noted in the right eye (Figs. 1 and 2). Dilated fundus exam revealed a normal disc, macula, vessels and periphery in both eyes.

Differential Diagnosis

The differential diagnosis for her unilateral proptosis with significant muscle restriction included thyroid-associated orbitopathy, nonspecific

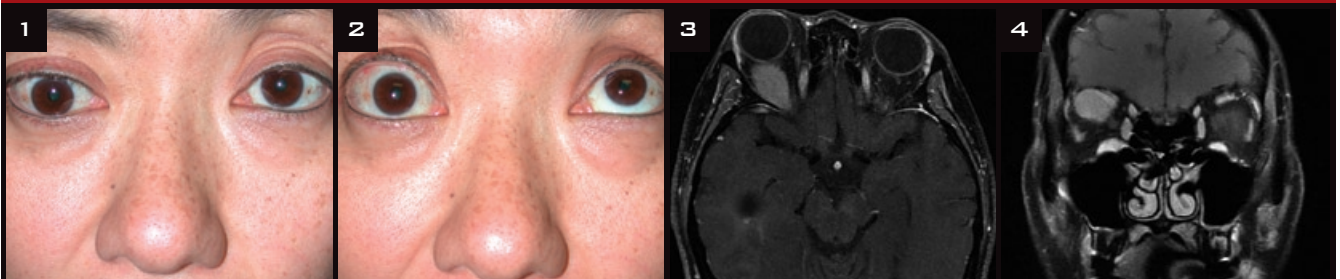
inflammatory disease (pseudotumor), or neoplasm, including metastasis and lymphoma.

Further Workup and Treatment

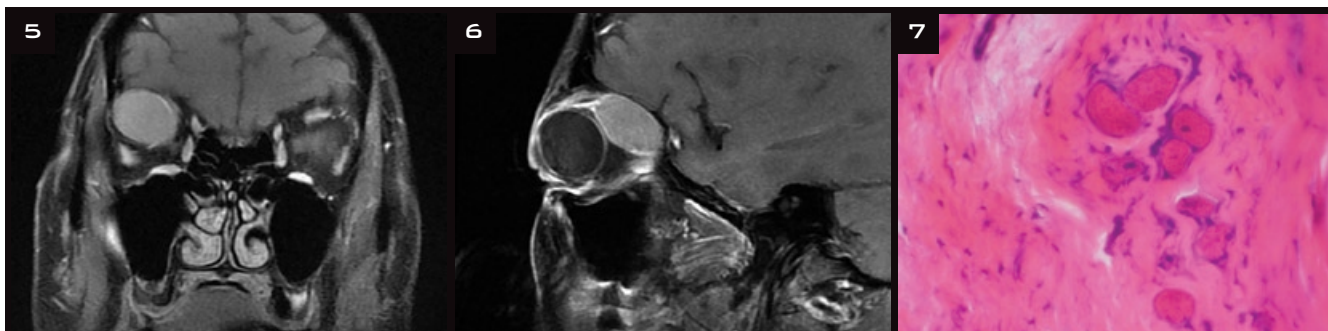
A CT scan revealed an impressive enlargement of the right superior rectus muscle (Figs. 3 and 4). In this initial scan, the inferior rectus muscle also seemed somewhat enlarged. Thyroid function tests and thyroid uptake scan were normal. Antithyroglobulin antibody levels were elevated. CBC, ANA, anti-SS-A and anti-SS-B were all normal.

Our presumptive diagnosis was Graves' disease. The patient was started on 60 mg of oral prednisone as a therapeutic challenge. This dose was tapered over several months. Upon taper, however, the proptosis worsened. In addition, the patient demonstrated signs of optic nerve compression, including decreased color vision and

What's Your Diagnosis?



WE GET A LOOK. We noted hypoglobus and proptosis of the right eye in primary gaze (1) and on attempted upgaze (2). CT imaging reveals enlargement of the right superior rectus (3 AND 4).



THREE MONTHS ON. When we repeated CT imaging, we noted progression of the superior rectus enlargement (5 AND 6). This prompted a biopsy. H & E stains of the superior rectus biopsy show extensive fibrosis and sparse muscle fibers (7).

visual field compromise. Repeat imaging demonstrated progression of the superior rectus enlargement (Figs. 5 and 6).

Because the patient's symptoms had increased, we decided that a biopsy of the superior rectus muscle should be performed. (To see the surgical video, together with scans from the 10-month follow-up, go to the online version of this article at www.eyenetmagazine.org.)

The muscle biopsy confirmed the diagnosis of Graves' disease. H & E staining demonstrated extensive fibrosis (Fig. 7). Trichome staining also demonstrated an overall increase in fibrosis, including endomysial fibrosis. This extensive fibrosis was consistent with the chronic phase of extraocular involvement in Graves' disease, but other typical findings—mononuclear inflammatory infiltrate with edema and myxoid material—were not found to be present.

The patient was continued on prednisone with a very slow taper. We were ready to offer surgical decompression, but her vision responded to the prednisone.

Ten months after initial presentation, an MRI of the orbits was repeated. This demonstrated a marked decrease in the right-sided proptosis and in the size of the previously enlarged superior rectus. Dr. Lee is now completely off steroids and continues to do well. Her vision is 20/20 in both eyes without a visual field defect or color vision compromise. She will continue to be monitored with repeat ophthalmologic examination and imaging.

Discussion

Thyroid-associated orbitopathy (TAO) is the most common cause of proptosis in adults and occurs in 50 percent of patients with Graves' disease. TAO is an autoimmune disorder in which it is believed orbital fibroblasts play an important role. The disease generally affects women in the fourth or fifth decade of life.

The clinical signs of TAO include eyelid retraction (the most common feature), lid lag, proptosis, restrictive extraocular myopathy and compressive optic neuropathy. In addition to the orbital signs, the diagnosis can be made in the presence of immune-related thyroid dysfunction and radiographic evidence of enlargement of one or more of the extraocular muscles (in particular, the inferior recti, medial recti and superior recti/levator complex). The enlargement of the muscles contributes to the proptosis and strabismus characteristics of this disease. Ninety percent of patients with TAO have hyperthyroidism, 6 percent have euthyroidism, 1 percent have primary hypothyroidism and 3 percent have Hashimoto's thyroiditis. In general, TAO is self-limiting. The active phase lasts approximately one year, but the disease runs its course over 18 to 24 months. It has a longer course in patients who smoke. Treatment options include observation, lubricants, decompression surgery, strabismus surgery, eyelid surgery, radiation and anti-inflammatory medications, such as steroids, montelukast sodium (Singulair), rituximab (Rituxan) and methotrexate.

An unusual case. Our patient had significant unilateral, painless proptosis associated with marked enlargement of the superior rectus muscle in the face of normal thyroid function tests. It is our belief that this represents a unique presentation of thyroid eye disease.

1 Bartley, G. B. et al. *Am J Ophthalmol* 1996; 121:284–290.

2 Kazim, M. et al. *Arch Ophthalmol* 2002; 120:380–386.

3 Mourits, M. P. et al. *Clin Endocrinol* 1997; 47:9–14.

* Patient name is fictitious.

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