Orbital imaging in thyroid-related orbitopathy

Christopher Lo, MD, Shoaib Ugradar, MD, and Daniel Rootman, MD, MS

SUMMARY A broad understanding of the different imaging modalities used to assess the physiologic changes seen in Graves' orbitopathy complement clinical examination. Subtle applications of radiographic imaging techniques allow for a better understanding of the overall physiology of the orbit, quantify progression of disease, and differentiate it from orbital diseases with overlapping features. A nuanced approach to interpreting imaging features may allow us to delineate inactive from active thyroid eye disease, and advances within this field may arm clinicians with the ability to better predict and prevent dysthyroid optic neuropathy. (J AAPOS 2018;22:256.e1-256.e9)

Orbital imaging plays a central role in the diagnosis and management of thyroid-related orbitopathy (TRO). Diagnostically, it is used to compliment a careful ophthalmic examination, laboratory values, and ancillary studies to confirm the presence of TRO and/or dysthyroid optic neuropathy (DON). It can also be helpful in surgical planning and understanding the progression of thyroid myopathy. Computed tomography (CT), magnetic resonance imaging (MRI), ultrasound, and nuclear medicine all have applications in the field. In this review we present the imaging features of TRO and discuss the current utility of imaging to assess disease activity. Differential diagnoses and indications for biopsy are also described. A range of imaging features and their practical applications are presented.

Imaging Findings

Muscles

Extraocular muscle enlargement is a hallmark of Graves' orbitopathy that has been observed intraoperatively, radiographically, and in postmortem studies.[1-5] In many cases the changes are obvious; however, the determination of whether a muscle is enlarged volumetrically can pose a challenge, particularly if there is symmetric mild disease (Figure 1). In the clinical setting, it is practical to determine muscle width in the midbelly region (approximately 1 cm posterior to the globe), which can be used as a proxy for muscle volume.[6,7] Normative data has been developed: mean inferior rectus width, 4.8 mm; medial rectus width, 4.2 mm; superior rectus width, 4.6 mm; and lateral rectus width, 3.3 mm.[8,9] These numbers can be used as a guide; however, they represent population averages, each with significant variation. Overlap in populations exist, and both diseased and nondiseased muscles can have widths close to these values. In the end, there are no strict rules.

In terms of muscle involvement, clinical myopathy is thought to most often involve the inferior rectus muscle, followed by the medial, superior, and lateral rectus muscles.[10] However, any of the extraocular muscles can be involved. Multiple muscles are typically involved simultaneously, and bilateral disease is found in 76%-90% of patients, although it is often asymmetric (Figure 2).[11,12] Isolated muscle disease (often the superior muscle complex) is rare but seen in 5% of cases.[10] It is also important to consider the image to be a snapshot in time and view myopathy as a dynamic process. Serial MRI can demonstrate fluctuations and change (Figure 3).

Classically, enlargement is described to spare the anterior tendon insertion (Figure 4A). This feature may help differentiate TRO disease from other forms of myositis. However, the rectus insertion can be enlarged in some cases of TRO (Figure 4B)[3,13,14] and, conversely, can be spared in non-TRO-related myositis.[15] The precise sensitivity and specificity of this sign is not well established and therefore represents supportive evidence of disease in the context of other characteristic biochemical and clinical findings, rather than a pathognomonic sign.

Fat

Fat expansion is additionally a central characteristic of TRO. A spectrum of disease exists, with fat predominant at one end and muscle predominant at the other. Lipogenic, or type 1, patients tend not to exhibit significant muscle enlargement or dysfunction (Figure 5), and in these circumstances clinical and biochemical correlates are critical for diagnosis.[16] Further, type 1 patients are less likely to develop strabismus after decompression surgery, making

Author affiliation: Division of Orbital and Ophthalmic Plastic Surgery, Stein and Doheny Eye institutes, University of California, Los Angeles

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Correspondence: Daniel Rootman, MD, MS, 100 Stein Plaza #2-267, Los Angeles, CA 90095 (email: Rootman@jsei.ucla.edu).

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FIG 1. Axial computed tomography (CT) of the orbits with symmetric enlargement of bilateral medial and inferior rectus muscle groups.

FIG 2. Axial CT of the orbits with prominent asymmetric muscle enlargement of the inferior and medial rectus on the right side compared with the left.

the phenotype important in prognosticating. Although each end can manifest at any age, muscle predominant disease is more frequently observed in older patients and fat predominant disease in younger.[17]

Though hypertrophy and hyperplasia of the fatty tissue may account for expansion of this compartment, an increase in orbital fat volume as a result of venous congestion from compression of the superior ophthalmic vein has been postulated as an alternative etiology for this phenotype.[3,18]

Clinically, expansion of the brow, galeal, premalar region, and cheek fat have also been described.[19-23] Complimentary changes to the retro-orbicularis oculi fat and the premalar suborbicularis oculi fat can also be detected using multiple imaging modalities.[24,25] This fat expansion is often underappreciated; however, it is pathologic in terms of the histology, and is widely derided by patients with the disease (Figure 6).[20]

Exophthalmos

While extraocular muscle enlargement is a principle radiographic abnormality of TRO, exophthalmos is a common clinical measure that can be confirmed radiographically.

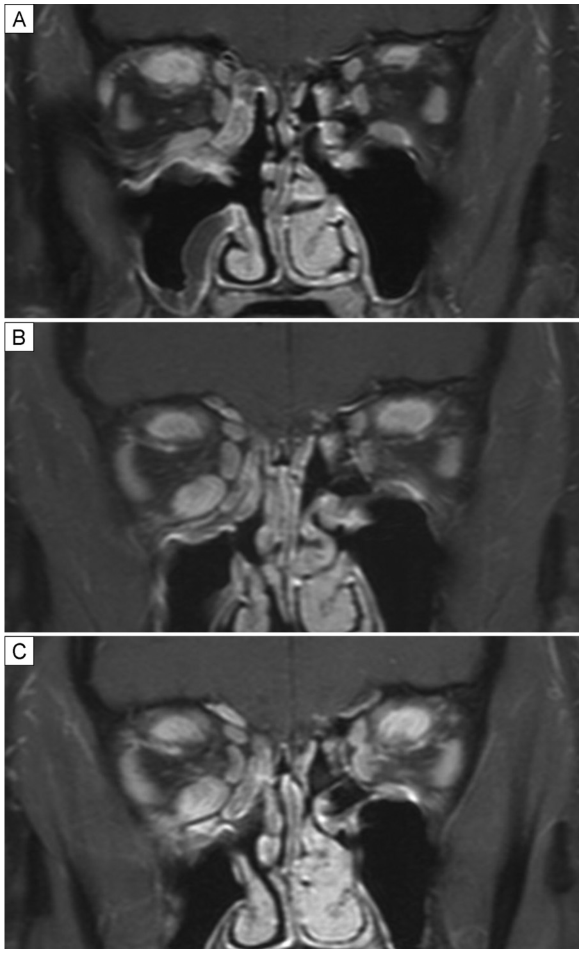

FIG 3. Serial coronal magnetic resonance imaging (MRI) T1 fat saturation sequence with contrast demonstrating progressive enlargement of the right inferior rectus and the left superior rectus muscle during the active stage of thyroid-related orbitopathy (TRO). A, Baseline exam. B, 6 months. C, Examination 5 months later.

Multiple methods have been proposed for this purpose. One involves tracing a reference line between the anterior extents of each zygomatic bone in a midorbital axial section. The distance from this line to the posterior aspect of the globe can then be measured. A normal distance is defined as being >10 mm, while smaller (or negative) distances indicate exophthalmos (Figure 7).[26]

Bony Changes

It is well known that the bony orbit undergoes remodeling in TRO. Bowing of the lamina papyracea of the ethmoid (Figure 8A) may represent an adaptive response to the enlargement of the adjacent medial rectus muscle and/or elevation of the intraorbital pressure.[3,27,28] This type of remodeling is less well described in the lateral wall because of differences in bone compliance, but it can occur (Figure 8B).[29] Medial wall bowing on CT or MRI is associated with diplopia and DON, both indicators of disease severity.[28,30,31]