



Hyperglobus and Pseudoptosis in Type 1 Lipogenic Thyroid Eye Disease

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ABSTRACT

Purpose: We present a case of Type 1 (lipogenic) Thyroid Eye Disease (TED) and our aim is to describe an atypical presentation of a rare orbital process.

Observations: A man in his 50s presented with left-sided eyelid drooping. His exam showed no evidence of active inflammation but did show left hyperglobus and ipsilateral upper eyelid pseudoptosis. He had no prior history or symptoms of Graves' Disease and imaging did not show evidence of extraocular muscle enlargement, bony asymmetries, or masses in the orbit. Subsequent lab work showed a low TSH (thyroid-stimulating hormone), elevated free T4 (thyroxine) and T3 (triiodothyronine), and elevated TSI (thyroid-stimulating immunoglobulin) index.

Conclusions and importance: This is a unique and atypical presentation of a patient diagnosed with Type 1 (lipogenic) TED causing hyperglobus and pseudoptosis secondary to fat expansion in the absence of other classic TED findings such as contralateral eyelid retraction or extraocular muscle enlargement. Thyroid eye disease can have a heterogenous disease presentation, as evidenced by this case, and should always be considered in the differential diagnosis of pseudoptosis.

1. Introduction

Thyroid eye disease (TED) is a rare, immune-mediated, inflammatory condition commonly associated with thyroid dysfunction with an incidence of about 2.67–16 per 100,000 females per year and 0.54–2.9 per 100,000 males per year.^{1–4} TED patients most commonly present with ocular surface irritation, conjunctival injection, eyelid swelling, lid retraction, proptosis, pain, or diplopia.^{5,6} Inflammation and expansion of the extraocular muscles and orbital fat drive these clinical findings and are best characterized by computed tomography (CT) or magnetic resonance imaging (MRI) of the orbits. Based on these clinical and imaging findings, TED can be classified as Type 1 (lipogenic), in which orbital fat expansion is the dominant feature, or Type 2 (myogenic), in which extraocular muscle enlargement predominates.^{7–9} Some patients have features of both types of TED. In a prior study of TED in Caucasian patients, 60% demonstrated myogenic predominance, in line with Type 2, 5% demonstrated lipogenic predominance, or Type 1, less than 10% had mixed presentation of both fat and muscle volume increase, and the remaining 25% had normal fat and muscle volume.^{2,10} TED can have heterogenous clinical manifestations with devastating systemic and visual consequences if not diagnosed and treated in a timely manner.

2. Case report

A man in his 50s presented to the oculoplastics service with eight months of left-sided eyelid “drooping” that was most notable during video conference calls. He endorsed intermittent obstruction of his visual axis without diplopia or pain. The patient denied recent trauma, prior eye or eyelid surgery, contact lens use, or periocular neurotoxin injections. His medical history was significant for psoriasis, idiopathic thrombocytopenia, and migraines. On examination, his visual acuity was 20/30 in both eyes with full Ishihara color plate testing and no relative afferent pupillary defect was noted in either eye. Extraocular motility was full bilaterally and exophthalmometer measurements were normal on both sides with no evidence of proptosis (17 mm on the right and 19 mm on the left at a base of 98 mm on Hertel exophthalmometry). External examination revealed a margin-reflex distance 1 of +4mm on the right and +2.5 mm on the left. A left hyperglobus was noted on exam with no palpable masses in the periorbita (Fig. 1). Levator function was 16 mm bilaterally and there was no superior scleral show on the right. There was no evidence of a fatigability component to his history or on examination. The anterior and posterior segment examination were otherwise unremarkable. Due to the external examination findings, an

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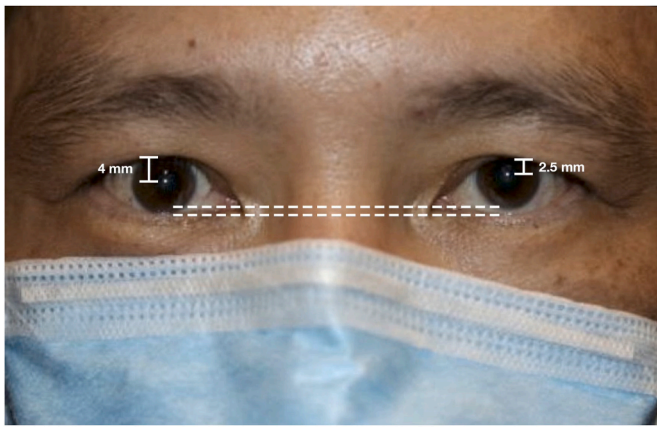


Fig. 1. Margin to Reflex Distance 1 was noted to be 4 mm on the right and 2.5 mm on the left.

MRI of the orbits was performed. Initial imaging for this patient was unremarkable with no evidence of extraocular muscle enlargement, orbital fat stranding, or orbital masses (Fig. 2A and B). CT of the orbits showed normal bony symmetry (Fig. 3). Labs drawn shortly after initial consultation revealed a TSH (thyroid-stimulating hormone) <0.01 (0.40–5.00 uIU/mL), free T4 (thyroxine) 6.4 (0.9–1.8 ng/dL), T3 (triiodothyronine) 345 (60–181 ng/dL), and a TSI (thyroid-stimulating immunoglobulin) 3.4 (≤ 1.3 TSI index). With a diagnosis of Graves' Disease and Type 1 (lipogenic) TED, the patient was subsequently referred to endocrinology and started on methimazole. Six months after treatment initiation for Graves' Disease, the patient's hyperglobus and pseudoptosis persisted. An inferior floor-only or fat-only orbital decompression was offered to aid in the symmetry of the eye position, but was ultimately not pursued.

3. Discussion

With these clinical and radiographic findings, we present a patient diagnosed with Type 1 TED with significant fat expansion causing hyperglobus and pseudoptosis of the left eye. This patient did not have a classic presentation of TED. He had no known history of thyroid



Fig. 3. Computed tomography of the orbits demonstrating normal bony symmetry.

dysfunction or symptoms of hyperthyroidism at the time of presentation or prior to presentation. Imaging did not show the characteristic extraocular muscle enlargement of TED, bony asymmetry, or masses to otherwise explain the left hyperglobus. Clinically, he did not show any active signs of inflammation. Given this constellation of findings, the left hyperglobus was the most dramatic finding resulting in left upper eyelid pseudoptosis. This could only otherwise be explained by fat expansion in the orbit. Pseudoptosis is caused by the appearance of a decreased palpebral fissure height, but without pathology of the eyelid muscles or aponeurosis. This most commonly presents in the setting of dermatochalasis and redundant upper eyelid skin. Pseudoptosis is an important consideration when evaluating for ptosis and can also be caused by blepharospasm, brow ptosis, enophthalmos, microphthalmos, or phthisis bulbi. Contralateral proptosis or contralateral lid retraction can also be causes of pseudoptosis and are more suggestive of TED. Due to the various etiologies that can drive pseudoptosis, it is important to effectively evaluate with a thorough work-up including imaging and bloodwork when indicated. Surgical intervention should be deferred until the etiology is determined. This is because hasty intervention can negatively impact the patient's ultimate outcome or mask an underlying pathology.

4. Conclusions

Thyroid eye disease can have varied presentations, but rarely presents with hyperglobus and pseudoptosis attributable to fat expansion. This case illustrates a unique example of Type 1 (lipogenic) TED and supports the inclusion of TED in the differential for pseudoptosis. More commonly in TED patients, pseudoptosis may present with contralateral eyelid retraction which can be indicative of TED. However, as evidenced by this case, even this finding may not be present. This case supports approaching patients with atypical presentations with a broad differential to avoid misdiagnosis, delayed diagnosis, or adverse outcomes, especially in the setting of TED.

Consent

The patient consented to publication of the case in writing.

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Author's contributions

All authors attest that they meet the current ICMJE criteria for

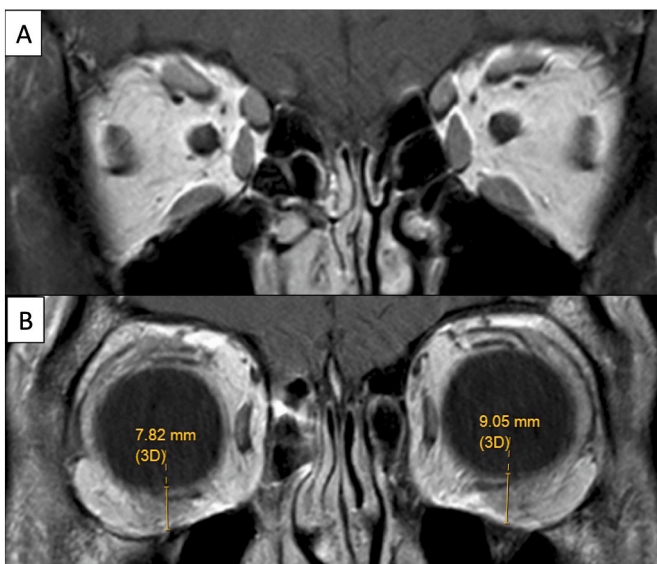


Fig. 2. A-B. A, Magnetic resonance imaging of the orbits demonstrating normal extraocular muscle size without enlargement and, B, hyperglobus of the left globe.

Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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