Thyroid Associated Ophthalmopathy with Ocular Myasthenia in Primary Hypothyroidism

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A 62-year-old female presented with bilateral proptosis and 1 year episodic eye pain, grittiness, redness, watering and intermittent diplopia for 6 months, and drooping of right eye lid for 2 months (Figure 1). She had a firm WHO grade-1b goiter, exophthalmos (26 mm and 23 mm in left and right eye respectively), Hertel exophthalmometer, clinical activity score of 1/6, without any evidence of bulbar, neck muscles and limb weakness.

Figure 1. Photo of patient in primary gaze, showing bilateral proptosis (left > right) with right eye ptosis

Computerized tomography (CT) of orbits revealed extraocular muscles thickening and expansion of retro-orbital fibro-fatty tissue confirming the diagnosis of thyroid associated ophthalmopathy (TAO) (Figures 2, 3). Investigations revealed low free T₄ (0.8 ng/dl; normal: 0.9-1.8 ng/dl), elevated TSH (38 µU/ml; normal: 0.4-4.2 µU/ml), elevated anti-thyroid peroxidase antibody (189 IU/ml; normal <35 IU/ml), anti-acetylcholine receptor antibody (2.32 nmol/L; normal <0.5 nmol/L); a repetitive nerve stimulation test of bilateral nasalis muscle was suggestive of ocular myasthenia with normal electrodiagnostic studies of limbs and normal CT thorax. Anti-TSH receptor antibody testing was not done, and is a limitation of this report. When she was last evaluated 3 months after diagnosis, ptosis had improved with pyridostigmine (60 mg 4 times daily) and levothyroxine (75 µg/d) initiated at dose of 1.25 µg/kg/day, which resulted in normalization of TSH (3.4 µU/ml).

Figure 2. Computerized tomography (CT) orbits showing bilateral thickening of intraocular muscles, predominantly involving inferior and medial rectus followed by superior rectus and lateral rectus due to thyroid associated ophthalmopathy.

Figure 3. Transverse section of computerized tomography orbits showing bilateral proptosis (left > right).
Primary hypothyroidism in TAO is rare, observed in only 5% cases, and is associated with milder and more asymmetrical ophthalmopathy. Hypothyroidism is extremely rare with myasthenia and is more associated with hyperthyroidism, occurring in 3-8% patients of patients. However, the occurrence of isolated ocular myasthenia with TAO is extremely uncommon with isolated reports in Graves’ disease, but never with primary hypothyroidism. Any patient with TAO and ptosis should be evaluated for ocular myasthenia. This case intends to highlight that TAO and ocular myasthenia can co-exist, even in primary hypothyroidism, is a diagnostic challenge with important consequences, as inability to achieve euthyroid status can worsen both TAO and myasthenia.

References