

Case Report

A Case of Unilateral Euthyroid Graves' Disease

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Abstract: Unilateral proptosis and ptosis in euthyroid Graves's disease is very rare with 5 -10% cases reported. We present a clinically Euthyroid male with features of thyroid ophthalmopathy who presented with unilateral proptosis and ptosis, restriction of extra ocular movements and compressive optic neuropathy. Thyroid function tests were normal and anti-thyroid peroxidase antibody was positive. CT scan revealed extraocular muscle thickening sparing the tendons. He was treated with systemic steroids for 8 weeks. Subsequently, symptoms gradually recurred and there was no improvement in vision. He was referred for orbital decompression. It is to be emphasized that any patient with unilateral proptosis and ptosis should be evaluated for thyroid eye disease and should be screened for anti-thyroid antibodies if thyroid function tests are normal. Early diagnosis and prompt treatment of ophthalmopathy with systemic corticosteroids can prevent vision threatening complications. This case intends to highlight that unilateral proptosis and ptosis may be the presenting features of euthyroid Graves' disease, and is a diagnostic challenge, with important consequences, as lack of timely treatment of Graves' disease can aggravate visual morbidity.

Keywords: Euthyroid Graves', proptosis, ophthalmopathy, thyroid antibodies, orbital decompression.

INTRODUCTION

Graves's disease is the most common form of thyrotoxicosis and may occur at any age, and is mostly seen in females. Graves's ophthalmopathy reflects the underlying autoimmune process and it is not a direct consequence of altered thyroid function [1]. The rarity of co-occurrence of euthyroid Graves's disease and ophthalmoplegia is not surprising because of rarity of euthyroid Graves's ophthalmopathy itself. Graves's disease should be considered as an important differential diagnosis of unilateral proptosis. TED is usually bilateral, although clinical signs may be asymmetrical [3, 4, 5]. Diagnostic difficulties in this patient were due to lack of characteristic systemic features of Graves' disease. Thyroid function tests were normal and anti TPO antibodies were positive. High levels of anti-TPO antibodies are demonstrable in the majority of patients and provide an important lead to the diagnosis when thyroid function tests are normal [7]. Euthyroid Graves' can present with unilateral proptosis, ptosis, ophthalmoplegia and optic neuropathy. Compressive optic neuropathy is to be treated with a trial of systemic steroids and if response is inadequate, should be promptly considered for orbital decompression [9, 10].

no pain, fever or systemic upset. There were no systemic features of thyroid disease or myasthenia gravis. There was no history suggestive of Wegener's granulomatosis or sarcoidosis. There was no history suggestive of primary malignancy in the nasopharynx, lung or gastrointestinal tract. General and systemic examination was normal. There was no thyroid mass or lymphadenopathy.

Visual acuity in right eye was counting fingers. Axial proptosis was 20 mm in the right eye and 10 mm in left eye as recorded by Hertel's exophthalmometer at 104 mm. There was periorbital oedema, ptosis, conjunctival congestion and chemosis (Fig 1, 2).



Fig 1: Proptosis with ptosis and chemosis

CASE REPORT

50 year old male presented with gradual onset of prominence of right eye and progressive visual loss for 1 month and drooping of upper lid for 1 week. There was



Fig 2: Proptosis - lateral view

Extra ocular movements were restricted in all gazes. Restriction of elevation is shown in fig. 3. Pupil was semidilated with sluggish direct light reflex in right eye and sluggish consensual reaction in left eye. Corneal sensations and facial sensations were normal.



Fig 3: Ocular motility restriction on elevation

Slit lamp examination revealed congestion and corkscrew vessels. Cornea was clear and lens showed nuclear sclerosis. Intraocular pressure was normal. Fundus examination showed normal optic disc in both eyes. Complete Haemogram, blood sugar and ESR were normal. Thyroid function tests (TFT) were normal (T3 - 134; T4 - 8.8; TSH- 3.7). Anti-thyroid peroxidase (TPO) antibody level was 52.8 IU/ml (normal 0-30) and anti thymoglobulin was 80 (normal < 225). VDRL & HIV were non reactive. CXR was normal.

Imaging (USG/ CT/ MRI) showed increased muscle bulk of all extra ocular muscles sparing tendon predominantly affecting inferior rectus and medial rectus muscles. There was an increase in the volume of orbital fat. Left orbit and contents were normal.

He was clinically diagnosed as a case of Euthyroid Graves' with unilateral proptosis, ptosis, and ophthalmoplegia with compressive optic neuropathy. IV pulse steroid therapy with methyl prednisolone 1 g IV was given for 3 days followed by oral prednisolone with an initial dose of 1.5 mg/kg tapered gradually over 6 weeks. Patient was symptomatically better at 4 weeks.

There was regression of proptosis (12mm on Exophthalmometry), reduction of periorbital edema and chemosis (Fig. 4, 5).



Fig 4: Regression of proptosis on follow-up



Fig 5: Regression of proptosis – lateral view

CT scan showed reduction in muscle bulk. There was minimal recovery of extra ocular movements and pupillary reaction but vision remained unchanged. After 6 weeks, pain and periorbital swelling reappeared and proptosis was measured as 16 mm. Increase in extra ocular muscle thickening was documented on CT scan. Extraocular movements and pupillary reactions were static. Vision remained unchanged at 8 weeks and he was referred for orbital decompression.

DISCUSSION

Proptosis in Thyroid Eye Disease (TED) is axial, non- pulsatile and is secondary to orbital venous congestion, accumulation of glycosaminoglycans and adipogenesis [1]. The swollen extra ocular muscles may compress optic nerve at the orbital apex causing visual compromise [2]. It is imperative that the ophthalmologist must not attribute visual loss to exposure keratopathy and miss concurrent optic nerve compression. Hence every patient with TED should be evaluated by visual acuity, colour vision, visual field testing to exclude optic nerve compression.

Graves's disease cannot be dismissed as a differential diagnosis of unilateral proptosis. TED usually involves both orbits, although clinical signs tend to manifest asymmetrically. When TED presents with unilateral proptosis, clinical features such as inflammation, impaired motility and eyelid retraction, may be present in the fellow eye, and proptosis may be obvious later in the course of the disease [3]. The prevalence of unilateral TED reported to range from 9% to 15% [4, 5].

Ptosis in a patient with Graves' disease suggests the co-existence of myasthenia gravis[6]. Restriction of extra ocular movements is observed in patients with thyroid associated ophthalmoplegia and in ocular myasthenia. Diagnostic difficulties in this patient were due to lack of characteristic clinical features of myasthenia gravis and Graves' disease. The presence of ptosis in thyroid eye disease can also be explained by levatoris insertion.

TFT was normal and anti TPO antibodies were positive. 80% of patients have high levels of anti-TPO antibodies[7].Ophthalmopathy dominant patients had significantly lower anti- TPO and anti-thyroglobulin antibodies [8].Regarding the correlation between presence, type or levels of antibodies and Graves' ophthalmopathy, studies have reported both thyroid stimulating antibody and thyroid binding immunoglobulin levels to be closely correlated with Graves' ophthalmopathy clinical activity score [8].

In patients with mild TED, progression to more severe disease is reflected by a change in the clinical activity score (CAS). The definition recommended by EUGOGO for moderate to severe TED is that sight-threatening disease is absent but that symptoms are sufficiently disabling to warrant immunosuppression during active disease, or surgery if inactive. Treatment is most effective when given early in the course of active disease, with high-dose intravenous glucocorticoids pulses for moderate to severe TED, with response rates of about 80% for parenteral treatment, as compared to about 60% for the oral route[9].High-dose systemic steroids (intravenous and oral) are sufficient to relieve tissue pressure at the orbital apex, and a gradual taper, without recurrence of the optic neuropathy, can be achieved[9]. However, where an adequate response is not seen with medical treatment, surgical decompression of the poster medial orbit should be considered, with studies reporting a rapid beneficial effect on vision. The objective is to relieve the hydrostatic pressure at the orbital apex and thereby, reduce orbital congestion and improve vascular perfusion and axonal flow within the optic nerve [10].

CONCLUSION

Euthyroid Graves' can present with unilateral proptosis, ptosis, ophthalmoplegia and optic neuropathy. Compressive optic neuropathy is to be

given a trial of systemic steroids and if response is unsatisfactory, should be promptly considered for orbital decompression. This case intends to highlight that unilateral proptosis with ptosis can coexist with euthyroid Graves' disease and is a diagnostic challenge. Anti thyroid antibodies may provide an important clue leading to the diagnosis when thyroid function tests are normal. Timely diagnosis is crucial as lack of treatment of clinically silent Graves' ophthalmopathy can aggravate visual morbidity.

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