

Case Report


A rare case of acute thyroid eye disease in a young male

Ruchira Nomula^{1*}, M. Vijaya Rama Raju², M. Ravinath³

¹Post Graduate, ²Professor and HOD, ³Assistant Professor

Department of Ophthalmology, SVS Medical College and Hospital, Mahabubnagar, Telangana, India

*Corresponding author email: ruchiranomula.95@gmail.com

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Abstract

Thyroid Eye Disease also known as Thyroid Associated Orbitopathy, Grave's Disease is typically a self-limiting autoimmune process associated with dysthyroid states usually manifesting in middle age. The clinical presentation may vary from very mild disease to severe irreversible sight threatening complications. Primary risk factors for TED are environmental influences especially smoking, previous pathogen exposures, stress, previous use of radio iodine and complex genetic components. Here we report a case of 25-year-old male who presented to our OPD with complaints of swelling of eyes, photophobia, lacrimation since 3days. Bilateral mild proptosis, severe congestion and conjunctival injection with 20/20 vision in both eyes were noted on examination. Blood investigations showed Eu-clinical Thyroid state whereas MRI Brain and Orbits showed all four Recti muscle belly enlargement. This case emphasizes one of the rare presentations of TED in accordance to Age, Sex and Clinical status of the thyroid.

Key words

Thyroid eye disease, Grave's Ophthalmopathy, Euthyroidism, Acute phase, Young male.

Introduction

TED (Thyroid Eye Disease) is a complex orbital inflammatory disease, which can be sight threatening, debilitating and disfiguring. TED is also known as Graves' ophthalmopathy, named after Robert J. Graves, an Irish physician who first described thyrotoxicosis in a woman

presenting with goitre, rapid heartbeat and exophthalmos [1]. The overwhelming majority (90%) of TED cases are associated with hyperthyroidism, while the rest are euthyroid or hypothyroid. Cross reactivity against antigen(s) in thyroid and orbital tissue is most likely responsible for the autoimmune ophthalmologic reaction [2]. The incidence of TED is 16 per

100,000 females and 2.9 per 100,000 males with an approximate prevalence of 0.25% with no significant ethnic predisposition. The higher preponderance in females relates to the higher incidence of hyperthyroidism in females. However, for severe TED, the ratio of females to males reverses to approximately 1:4 [3].

Risk factors for the disease include female gender, Middle age. Smoking is the strongest modifiable risk factor for developing TAO. Recent studies have shown to have impact on specific gene expression involved in several disease-related pathways, which seems to be reversible with smoking cessation. Selenium deficiency and high serum cholesterol have been described to be potential independent risk factors for TED [4].

TED is caused by retro-orbital inflammation to which orbital fibroblast activation is a key contributor. Fibroblast activation is presumed to occur secondary to stimulatory auto-antibodies like anti-TSHR, anti-IGF-1. These fibroblasts express the TSH receptor and produce extracellular matrix components and pro-inflammatory molecules, they also cause infiltration of immunocompetent T-helper cells (type-1), B lymphocytes, macrophages and mast cells. Inflammation of extraocular muscles lead to restricted eye movements and proptosis. A unique feature of TED is it is self-limiting, the reason being the absence of lymphoid tissue within the orbit. TED typically has an active inflammatory phase subsides over 6months to 5years followed by a fibrotic, inactive phase [5, 6].

The signs and symptoms of TAO are as varied as its epidemiology and pathophysiology. Symptoms range from tearing, eye pain to double vision, and signs extend from conjunctival injection and chemosis to lid retraction, EOM restriction, strabismus, to sight threatening exposure keratopathy and compressive optic neuropathy [7]. Diagnosis is based on series of investigations including blood levels of Thyroid hormones, MRI Brain including orbits, enhanced

CT Brain that shows extra ocular muscle enlargement. Treatment is symptomatic in the acute phase and other modalities like orbital decompression as when required [8].

Case report

A 25-year-old male patient presented with complaint of swelling of eyes, pain, photophobia and watering since 3 days (**Figure - 1A and 1B**). No complaints of headache, diminution of vision. No complaint of double vision. Patient didn't give any history of fever or any trauma. There were no similar complaints in the past. No similar complaints in the family as well. He is a chronic smoker since 8 years. On examination, his visual acuity was 20/20 in both eyes, bilateral mild proptosis with upper lid retraction was noted. Extra ocular muscles showed movement restriction in all the gazes, more in infra-duction followed by supra-duction and adduction. On slit lamp bio-microscopy, anterior segment showed severe congestion and chemosis of conjunctiva in both eyes. Rest of the ocular and systemic examination was within normal limits.

Figure - 1A and 1B: Patient showing severe chemosis, conjunctival injection, upper lid retraction and mild proptosis of both eyes.



Figure - 2: MRI Brain and Orbits- Axial view: showing Lateral and Medial Rectus Muscle belly enlargement of both the eyes (pointed by the red arrows).

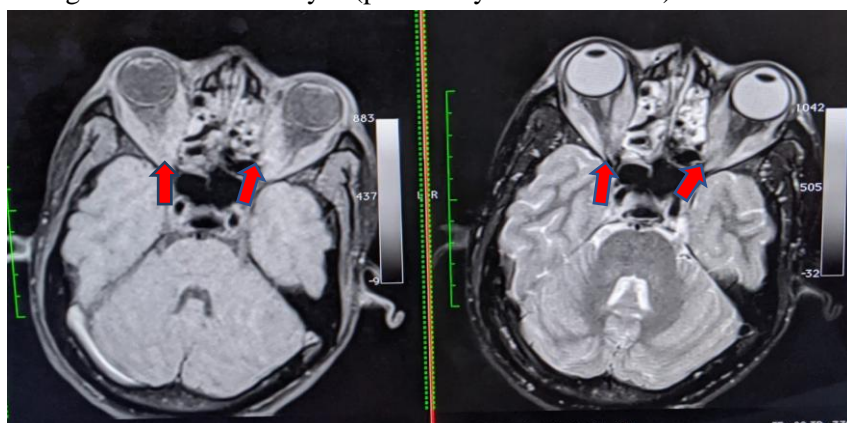


Figure - 3: MRI Brain and Orbits- Coronal view: showing the enlargement of all the four Recti muscles (superior, medial, inferior, lateral rectus muscles in clockwise direction) of both the eyes indicating the classic feature of Thyroid Orbitopathy.

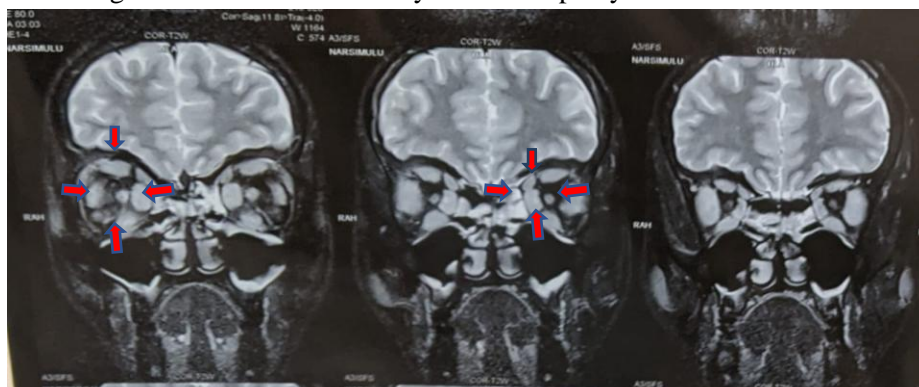


Figure - 4: Patient post treatment with systemic and oral steroids.



Investigations

Hb - 12.2 g/dL

ESR - 40 mm/Hr

Viral Serology - Negative

Free T3 - 3.37pg/mL

Free T4 - 1.04 ng/dL

Thyroid Stimulating Hormone (TSH) - 0.78 μ IU/mL

MRI Brain and Orbits showed Enlargement of all four Rectii muscle belly enlargement as seen in **Figure - 2 and 3**. Patient post-treatment with systemic and oral steroids is as seen in **Figure - 4**.

Discussion

The term Thyroid Eye Disease (TED) denotes typical ocular changes which include lid retraction, lid lag, and proptosis. These changes have also been labelled as: Endocrine Exophthalmos, Malignant Exophthalmos, Dysthyroid ophthalmopathy, Ocular Graves' Disease, Graves' Ophthalmopathy and Thyroid Associated Ophthalmopathy. There are many classifications that elaborates the course of the

disease in regard to symptoms, signs and prognosis. They are NOSPECS classification by American Thyroid Association, VISA classification for severity of the disease, EUGOGO classification for severity grading, CAS (clinical activity scoring) for grading TED [9, 10].

Diagnostic criteria for TED include combinations of the classical signs, as only 5% of the patients present with entire constellation of the findings.

The criteria include the following:

- 1) Eyelid retraction to the level of superior limbus or above along with any one of these: Thyroid abnormalities, preferably including anti-thyroid antibodies; Exophthalmos; Optic neuropathy; Extraocular muscle involvement. OR
- 2) Thyroid function abnormality with one of the following: Exophthalmos; Optic neuropathy; Extraocular muscle involvement [10, 11].

The treatment for TED should be a multidisciplinary approach involving an ophthalmologist, an endocrinologist, a radiologist and a general physician. The principal aim should be thyroid function control as this is associated with reduction in the severity of the disease. General supportive measures should be implemented immediately that includes ocular lubricants, head elevation (as gravity supports lid closure), taping lids at nights, tinted spectacles and prisms in spectacles to control diplopia, and counseling should be given to support the morale of the patient. Thereby treatment is given according to the phase of the disease [1]. In acute active phase Systemic Corticosteroids are considered and are most effective in this phase. In our case we administered intravenous Methyl Prednisolone 1mg/kg/day for three days followed by tapering doses of Oral Prednisolone for a period of 3weeks. Following the treatment patient was recovered from the symptoms. Other agents like cyclosporin, azathioprine, rituximab are gaining popularity and some are under clinical trials [12, 13].

Cessation of smoking, selenium supplementation can be used as it showed significant improvement in the quality of life, reduces ocular involvement and slows down the progression of TAO. Orbital radiotherapy is used as an adjunctive therapy and is particularly effective at improving ocular motility during active phase due to the sensitivity of orbital lymphocytes to radiotherapy [14, 15]. Surgical intervention for cosmetic appearance and symptoms should be avoided till the inactive or quiescent phase. Surgical options include orbital decompression, extraocular muscle surgery and lid correction in that order. Decompression of either fat or bony part of orbit aims to improve proptosis and lid position whereas motility surgeries involving muscle repositioning to reduce/ eliminate diplopia and abnormal head postures. Finally lid surgeries are done to reposition the lids(s) [16].

Additional therapies have been proposed and are in varying stages of study for efficacy in TAO. A retrospective review demonstrated an improvement in both CAS score and motility restriction with methotrexate therapy, prospective data have not been presented. TNF- α is known to be up-regulated in TAO, several studies have evaluated the use of TNF- α antagonists. Cyclic peptides maybe another promising avenue for future therapy. Further, IL-15, IL-17 are up-regulated in patients' tears in active disease compared with inactive disease. A targeted therapy to these may have a role in the management of TAO [1].

Conclusion

Thyroid associated orbitopathy is a debilitating and vision-threatening disease. Pathophysiology of the disease is yet to be completely elucidated; however, TAO is understood to be an inflammatory process. Activation of proinflammatory cytokines results in increased hyaluronan production by orbital fibroblasts, leading to the classical manifestations of TAO. As we already discussed the age, sex and thyroid status preponderance of TED/TAO, this case throws light on the rare presentation as in a

young male (25 years) with eu-clinical status of the thyroid. Patients with thyroid abnormalities should be screened and educated about the signs and symptoms of TAO. A multidisciplinary group including oculoplastic surgery, strabismus surgery, and neuro-ophthalmology can closely monitor patients for disease progression and implement appropriate medical and surgical therapy.

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