Original Research Article

Severe bilateral orbitopathy in a middle-aged male patient with Hashimoto's thyroiditis

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Abstract

Background: Thyroid-associated orbitopathy (TAO), also known as Graves' orbitopathy, is typically a self-limiting autoimmune process associated with dysthyroid state. It is the most common orbital disorder and most common cause of proptosis in adults[9]. TAO is most prevalent in hyperthyroid patients with Graves' disease (GD); however, severe cases of orbitopathy associated with Hashimoto's thyroiditis (HT) have rarely been described. A 46-year-old male presented to our institute with bilateral proptosis since 4 months. The patient was referred to the radiology department for further investigational work up with computed tomography and magnetic resonance imaging of the orbit. The findings noted were marked bilateral exophthalmos secundary to an increase in orbital fat volume and bilateral symmetric enlargement of extraocular muscle(EOMs) belly with sparing of the tendinous Insertions which suggest the diagnosis of Thyroid-associated orbitopathy (TAO).

Keywords: Orbitopathy, Hashimoto's thyroiditis, Multidetector Computed Tomography, Magnetic Resonance Imaging, Ultrasonography, Color Doppler Ultrasonography

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INTRODUCTION

Thyroid-associated orbitopathy (TAO), is an organ specific autoimmune inflammation, centered on the extraocular muscles and orbital connective tissue. It is associated with Graves' disease in 90% of cases, less frequently with Hashimoto's thyroiditis[4].TAO may occur during or after the onset of hyperthyroidism and less frequently, in euthyroid or hypothyroid patients[10]. There are two distinct stages of thyroid orbitopathy – the active inflammatory phase, which usually encompasses an average period of 18-36 months, characterised by lymphocytic infiltration, interstitial oedema and glycosaminoglycans deposition in enlarged extraocular muscles and retro orbital fat, and the inactive disease with fibrosis and fat infiltration of the orbital tissues (fibroblast and preadipocytes' induction towards lipogenesis)[4,10]. CT,MRI scanning are accurate imaging procedures for the diagnosis of thyroid associated orbitopathy . Beside the visualization of the enlarged extraocular muscles and the proptosis extent, CT scans provide the surgeon with depictions of the bony architecture of the orbit when orbital decompression is required[4]. Apical crowding seems to be the most sensitive indicator for the presence of optic neuropathy. Herewith we report an unusual case of a middle-aged clinically and biochemically hypothyroid man with a Hashimoto's thyroiditis who developed a severe bilateral TAO.

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CASE REPORT

A 46-year-old male was admitted to hospital because of bilateral eye proptosis progressively getting worse for about four-months and pedal edema. He had suffered from hypothyroidism and hyperertension for the past eight months and had been treated with levothyroxine 50µg daily and Telma 40mg daily respectively. He was a non-smoker and did not present any other significant comorbidities. His previous personal and family history was negative for thyroid disorders. On physical examination, body temperature was 37 C, pulse rate was 102 beats/ min, blood pressure was 120/70 mmHg. The skin was warm and sweaty. Eyes had bilateral extremely exopthalmic appearance and also swelling and congestion of the conjunctivas were present. Laboratory results indicated hypothyroidism - TSH levelwas 47.0 µIU/ml (reference range 0.4–4.20 µIU/ml), the free T3 and free T4 concentrations were 2.63pg/ml (reference range 2.3–4.2 pg/ml) and 0.55 ng/dl (reference range 0.89–1.76 ng/dl), respectively. Anti-thyroid peroxidase antibodies (TPOAb) serum levels and anti-thyroglobulin autoantibodies (TgAb) were significantly elevated: 482 U/ml (reference range <60 U/ml) and 486.3 IU/ml (reference range <34 IU/ml), respectively. Ultrasound examination of thyroid revealed heterogeneously decreased echogenicity of bilateral lobes of thyroid with micro cystic spaces ,septations and normal flow on colour doppler, suggestive of Hashimoto's thyroiditis(Figure 1).USG guided FNAC was done and report came as Bethesda-II ,Benign, Consistent with lymphocytic(Hashimoto) thyroiditis CT scan of the orbits showed bilateral symmetric enlargement of extraocular muscle belly (EOMs)(Figure 3) with sparing of the tendinous Insertions and the globe has been displaced anterior to the inter-zygomatic line (IZL) secundary to an increase in orbital fat volume and enlargement. The distance between the inter-zygomatic line and the anterior margin of the right globe was 28.8mm and the left globe was 29.4mm(Figure 2). MRI of the orbits confirmed EOM enlargement with T2 hyperintensity with in it; largest in bilateral inferior, medial and superior rectus. Closer review of MRI images also showed lacrimal gland enlargement, The presence of TPO antibody, a picture of Hashimoto's thyroiditis on USG scan, FNAC as Bethesda-II Benign, Consistent with lymphocytic thyroiditis and changes in the orbital muscles confirmed the diagnosis of thyroid associated orbitopathy with Hashimoto's thyroiditis.

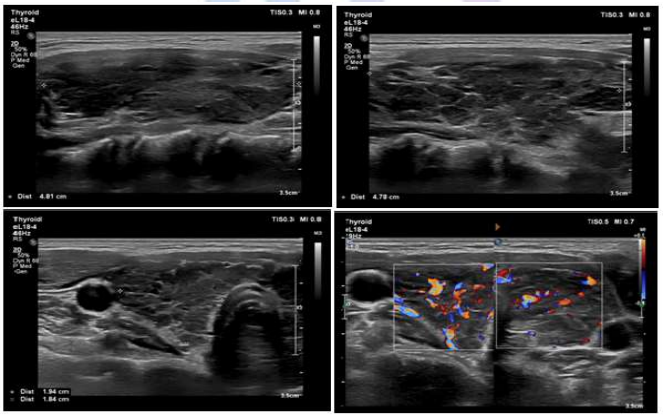


Figure 1

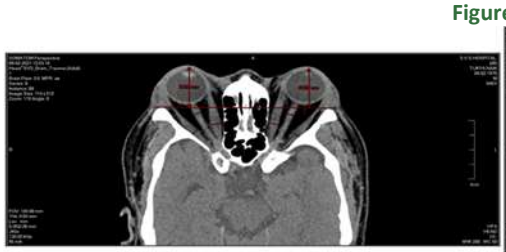
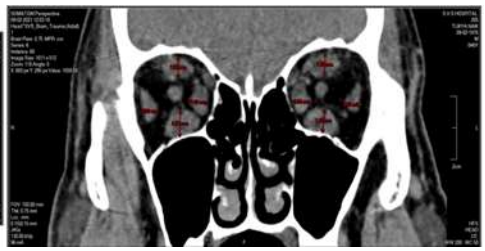


Figure 2



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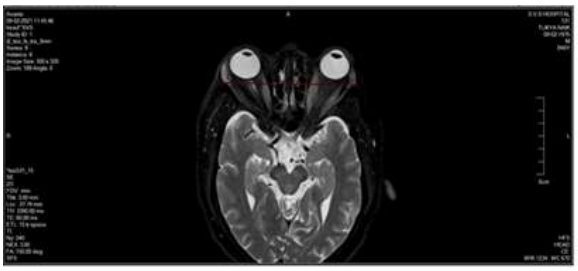

Figure 4

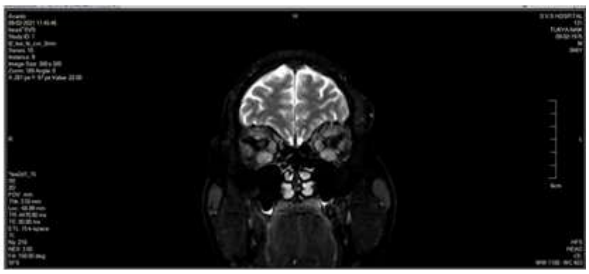

Figure 5

Figure 1: Transverse and longitudinal gray-scale usg neck demonstrates ill-defined heterogeneous hypoechoic areas with microcystic spaces, septations in bilateral lobes of thyroid and transverse scan of bilateral lobes of thyroid shows normal vascularity on color doppler sonogram; **Figure 2:** Axial non contrast CT scan at mid globe demonstrates the globe has been displaced anterior to the inter-zygomatic line (IZL) secundary to an increase in orbital fat volume and enlargement of extraocular muscle bellies. The distance between the inter-zygomatic line and the anterior margin of the globe, width of the optic nerve-sheath complex, and muscle diameter measurement for the medial and lateral rectus; **Figure 3:** Coronal CT reconstruction of the orbit demonstrates enlargement of the inferior, medial, superior and lateral rectus muscles, the measurement of the extraocular muscle; **Figure 4.** Axial T2 WI fat suppressed MR image shows marked bilateral exophthalmos as indicated by inter-zygomatic line and Bilateral enlargement of medial and lateral rectus, which have increased signal intensity. The anterior tendon is typically spared with the enlargement largely confined to the muscle belly. Enlargement of the bilateral lacrimal glands; **Figure 5:** Coronal T2WI fat suppressed MR image shows enlargement of the inferior, medial, superior and lateral rectus muscles, which shown T2WI increased signal intensity.

DISCUSSION

TAO is the most common disease affecting the orbit, mostly affects people between 30 and 50 years of age, females are five times more likely to present with it[5,6].The incidence of TAO is approximately 16/100,000 in females and 3/100,000 in males (TAO is more severe in males)[5].Although TAO is more common in women, the proportion of men increases as severity increases. The female to male ratio was 9.3:1 in patients with mild ophthalmopathy, 3.2:1 in those with moderate ophthalmopathy, and 1.4:1 with severe ophthalmopathy. At the time of diagnosis 90% are hyperthyroid, 1% are hypothyroid, 6% are euthyroid, and 3% are Hashimoto thyroiditis[1].There is strong evidence for a causal association between smoking and development of TAO[5]. To date only a few studies regarding the issue of TAO accompanying Hashimoto thyroiditis have been conducted. The first study on the prevalence of ophthalmic symptoms in patients with HT was published by Tijang *et al.* who evaluated a group of 20 patients with HT, and found that eye signs were prevalent in about 34% of patients[2]. TAO is the most frequent extra thyroid symptom of Graves' disease, as well as one of the most difficult and deceptive in clinical presentation. The disease can significantly deteriorate the life quality of the patient and may lead to vision loss, resulting permanent disability. Therefore, diagnosis of the dysthyroid orbitopathy and assessment of the effects of its treatment should rely on relevant measurements, i.e. diameters of extraocular muscles, volume of retrobulbar fibro-adipose tissue, proptosis, lid fissure width, range of extraocular motion on perimeter, and quantitation of diplopia fields[4]. Sensitive spatial radiological diagnostic modalities (CT, MRI) can evaluate not only clinically noticeable infiltrative stage of the disease, but also muscle involvement in the latent phase. According to The European Group on Graves' Orbitopathy (EUGOGO) imaging of the orbits by CT or MRI scanning is recommended when diagnosis based on the clinical or serologic findings is uncertain, in cases of apparently unilateral disease and suspected dysthyroid optic neuropathy[4,8]. It is also used to indicate the degree of muscle involvement and its bilaterality, to monitor patients for progression of the disease, to exclude coexisting pathology and to aid surgical planning and evaluating the result of orbital decompression surgery . Computed tomography scanning provides a number of objective quantitative measures which identify patients at increased risk of orbital complications[4,7]. It surpasses ultrasonography with its imaging capability to evaluate extraocular muscles and optic nerve involvement at the orbitalapex, and is complementary to MRI – with poorer soft tissue resolution but better bone architecture resolution. Extraocular muscles are measured in the planes parallel to their course: in axial scans(medial rectus, lateral rectus) or in coronal scans (inferior rectus, superior group, superior oblique) and perpendicularly to the orbital wall, which provides accuracy, comparability and repeatability of the results. Post processed planar reconstructions enable the consultant to perform a survey of the orbits while the scanning is performed in single plane acquisition. Studies should be assessed both in soft tissue and bone algorithm. Enlargement of the extraocular muscle diameter as well as density decrease was pertinent to all muscle groups, implying that fat and inflammation in dysthyroid orbitopathy usually do not incorporate individual orbital elements, but the entire orbit compartment, regardless of

the fact that their reaction manifestation is not accurately similar. Computed tomography imaging enables highly sensitive and specific diagnostic of the development of optic neuropathy with apical crowding induced by the muscles and/or fat tissue that seems to be the most appropriate indicator of intraorbital pathology. MRI is preferred for studies assessing disease activity because of its better performancein the evaluation in soft tissues, provides better imaging of the optic nerve than CT and better assessment of the effacement of the perineural fat secundary to apical crowding. MRI is the modality of choice to identify active inflammatory changes and to assess any immuno suppressive treatment response. MRI is a valuable tool to distinguish the acute inflammatory active disease from fibrotic, inactive end stage disease based on the signal intensity of enlarged muscles[9].

CONCLUSION

TAO is a autoimmune disease associated mainly with hyperthyroidism, but also with hypothyroid and euthyroid states. Graves' disease is the most common cause of orbitopathy. Hashimoto's thyroiditis results in orbitopathy in a small percentage of patients. Diagnosis of TAO is usually given by careful ophthalmological examination when clinical manifestations occur. The main symptoms of the orbitopathy derive from the discrepancy between limited space of the orbit and expansion of pathologically affected orbital tissues. Therefore, in most patients, CT and MRI of the orbit confirm diagnosis by showing enlarged extraocular muscles (without involvement of the tendon) and/or increased orbital fibroadipose tissue.

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