Coexisting Graves disease and ocular myasthenia gravis in an 18 year old female: a case report

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ABSTRACT

Myasthenia gravis is an autoimmune disorder which affects the neuromuscular junction which has a predilection for muscles innervated by the cranial nerves. Graves’ disease is a similar autoimmune disorder due to circulating thyroid auto-antibodies. The coexistence of Grave’s disease and ocular Myasthenia Gravis has been reported occasionally even though it is rare. But reports of its occurrence in African patients were very few. This is a case report of an eighteen year old Ethiopian female patient presenting with features of both diseases. Since these features could easily be overlooked, the discussion emphasizes on the importance of high index of suspicion in similar cases.

Key words: Graves’ disease, Graves’ ophthalmopathy, Myasthenia Gravis, Autoimmune thyroid disease

INTRODUCTION

Autoimmune Thyroid Diseases (AITD) are autoimmune disorders due to circulating thyroid autoantibodies. They include Graves’ disease and Hashimoto thyroiditis. Graves’ disease is characterized by hyperthyroidism having one or more of the following features: Thyrotoxicosis, Goiter, Ophthalmopathy and Dermopathy (pretibial myxedema). Graves’ Ophthalmopathy (thyroid associated orbitopathy) is part of the autoimmune process that affects the orbital and periorbital tissues. Its ocular manifestations include eyelid retraction, proptosis, chemosis, periorbital edema and altered ocular motility.

Myasthenia Gravis (MG) is a relatively rare autoimmune disorder in which antibodies form against acetylcholine nicotinic post synaptic receptors at the neuromuscular junction of skeletal muscles. It is usually identified as having an ocular and generalized form with predilection for muscles innervated by the cranial nerves. The usual systemic symptoms are generalized muscular weakness including limbs and truncal muscles with later involvement of respiratory muscles, while the common ocular features are extraocular muscle weakness and/or ptosis.

There is a common incidence of association between Graves’ disease and other autoimmune diseases including MG, insulin dependent diabetes mellitus, pernicious anaemia, Sjögren’s syndrome, SLE and rheumatoid arthritis. Especially there were frequent reports of coexistence of MG and Graves’ disease even though reports from African countries were very few. It is said that MG occurs in approximately 5-10% of patients with MG, while MG is reported in 0.2% of patients with AITD, the most common of which is thyrotoxicosis.

Garlepp et al described that antithyroid antibodies and a history of thyroid disease were much more prevalent in ocular MG than in generalized MG. The clinical presentation of MG associated with AITD is frequently restricted to eye muscles evidenced by a greater frequency of thyroid antibodies in patients with ocular MG (51.8%) than generalized MG (16.6%), and the presence of thyroid antigens in ocular tissues of patients with ocular MG. Generalized MG was found to be more frequent in patients without thyroid disease. Thymic abnormalities were less frequent in MG associated with AITD than in MG without AITD. Here is a case report of a young female patient presenting with ocular MG after having symptoms suggestive of Graves’ disease.

CASE REPORT

An eighteen year old Ethiopian female patient presented to Jimma University, Department of Ophthalmology with anterior neck swelling of 2 years duration and protrusion of both eyes of one year duration. She had associated blurring of vision and drooping of eyelids but no pain, discharge or redness of the eyes. She had history of hot intolerance, palpitation, easy fatigability, weight loss, and amenorrhea. She had no history of smoking.

On general examination, she had tachycardia (Pulse rate 140/min) but other vital signs were within the normal range. She had diffuse, 12x8cm, non tender anterior neck mass which moved with swallowing (Figure 1). There was no evidence of other skeletal muscle involvement.
On ocular examination, she had bilateral symmetrical mild proptosis measuring 23mm from lateral orbital margin by Hertel exophthalmometer in both eyes. She also had bilateral symmetrical ocular motility restriction in all fields of gaze and ptosis (Table 1, Figures 2 and 3). There was no evidence of inflammation of conjunctiva or eyelids. Intraocular pressure in both eyes was 16mmHg. The pupils were equal and symmetrically reactive to light. Fundus examination was normal. Ice pack test was positive - ice was put in small plastic bags and placed over both upper lids for two minutes, then ptosis assessment repeated (Figures 4 and 5, Table 1). Edrophonium chloride was not available for tensilon test.

**Table 1:** Comparison of ptosis before and after icepack test

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Before ice pack</th>
<th>After ice pack</th>
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<tbody>
<tr>
<td></td>
<td>Right eye</td>
<td>Left eye</td>
</tr>
<tr>
<td>Margin reflex distance</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Palpebral fissure height</td>
<td>7mm</td>
<td>7mm</td>
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<tr>
<td>Levator excursion</td>
<td>7mm</td>
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**Figure 2:** Front view of the patient showing bilateral symmetrical proptosis with ptosis

**Figure 3:** Lateral view showing the mild proptosis

Based on the above findings, investigations were done. All thyroid function tests were found to be in the normal range: TSH = 4.33 mIU/l, total serum T3 = 0.83 nmol/l, and total serum T4 = 71.61 nmol/l. Her ESR was raised (60mm/1st hour) but all other haematological and biochemical tests were within the normal limit. Chest X-ray was normal with no significant thymus enlargement. Ultrasound of the orbit was also normal with no significant extra ocular muscle thickening.

**Table 2:** Comparison parameters before and after treatment

<table>
<thead>
<tr>
<th>Parameters</th>
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<tr>
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<td>Levator excursion</td>
<td>7mm</td>
<td>7mm</td>
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<tr>
<td>Proptosis</td>
<td>23mm</td>
<td>23mm</td>
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**Figure 4:** Showing the improvement of ptosis after 2 minutes of applying ice pack

**Figure 5:** Shows more prominent lid retraction after few more minutes of ice pack test

**Figure 6:** Improvement of ptosis and proptosis after one month of initiation of treatment

The patient was started on oral steroids then referred to expert internist and surgeons for further management and she was started on antithyroid drugs.
The patient was followed up one month later and so far she had managed to use only the oral steroids due to unavailability of acetylcholine esterase inhibitors locally. Ptosis, proptosis and ocular motility had improved markedly at the one month follow up review (Table 2 and Figure 6).

**DISCUSSION**

This case report shows the association of Graves disease and ocular MG in an Ethiopian patient and supports the previous data on the two diseases in other parts of the world. The reason for the association between the two disease entities is not clearly known, but several hypotheses have been postulated. The most investigated explanation was the finding of positive relationship between circulating auto-antibodies against both TSH receptors and acetylcholine receptor as well as their immunological and clinical activities. There were evidences for the presence of immunological cross-reactivity against epitopes or auto-antigens shared by the thyroid and the eye muscles. The other hypothesis suggests that there might be common genetic background. Human Leukocyte Antigen (HLA) specificity (B8, DR3, and BW46) between MG and thyroid disease has been reported. However, further studies are needed to clarify the precise association between thyroid disease and MG.

Our patient initially had symptoms of thyrotoxicosis, which includes anterior neck swelling, hot intolerance, weight loss and amenorrhoea, one year prior to dropping of both eyelids and blurring of vision. This is consistent with the studies done before which stated that in 75% of patients with both conditions, thyrotoxic symptoms occur before or concurrently with those of myasthenic symptoms. In one case report, the patient had thyrotoxicosis symptoms 2 months before his ptosis developed.

We couldn’t compare the response of treatment with those studies which used standard treatment modalities because of unavailability of the drugs like anticholinesterase and plasmapharesis in our setup. Surgical intervention like thymectomy was not considered due to lack of strong evidence of thymic abnormality in this patient. But the patient showed significant improvement in ptosis, proptosis and ocular motility after initiation of antithyroid therapy and oral steroid. This is in line with the study by Schneider-Gold et al. who described two double blind trials comparing prednisolone with placebo and revealed significantly greater improvement at two weeks in patients taking prednisolone for generalized MG. In another study, two-thirds of the patients with both disorders showed improvement in MG after treatment of thyroid disease only. Some authors state that thymectomy may have positive effects on both myasthenia and AITD in patients with evidence of the presence of an enlarged thymus and thymic abnormalities. On the other hand, a trial by Schneider-Gold et al. on the use of adrenocorticotropic hormone compared with placebo for the treatment of ocular MG did not show any advantage.

In conclusion, this case report emphasizes on the need of high index of suspicion of coexisting ocular MG in patients with both signs of thyroid eye disease and ptosis. The report also shows that it is still possible to diagnose the presence of Myasthenia Gravis indirectly using ice pack test in limited resource centers (even though it may not be confirmatory), with improvement of both conditions after antithyroid drugs and steroids. Generally, appropriate diagnosis of MG at an early stage is recommended as it may have prognostic significance.

**REFERENCES**


