Ocular Myasthenia Gravis and Auto-Immune Thyroiditis in Children

Kitthisak Kitthaweesin MD*, Narong Auvichayapat MD**, Ouyporn Panamonta MD**

* Department of Ophthalmology, Faculty of Medicine, Khon Kaen University, Khon Kaen
** Department of Pediatrics, Faculty of Medicine, Khon Kaen University, Khon Kaen

Case Report

A 7-year-old girl presented at a university hospital with ptosis of the left eye. This resolved spontaneously within 4 weeks but then the right eye became similarly affected but responded to prostigmine. Left hypertropia with restriction of the right inferior rectus, mild exophthalmos, non-tender diffuse enlargement of the thyroid, normal thyroid function tests, anti-thyroglobulin, and anti-microsomal antibodies indicated an association of autoimmune thyroiditis and ocular myasthenia. The ptosis was remedied with pyridostigmine and short-course oral prednisolone, but the hypertropia persisted.

Keywords: Pediatric myasthenia gravis, Autoimmune, Thyroiditis, Prostigmine

Myasthenia gravis in children, as in adults, is caused from an idiopathic production of acetylcholine receptor antibodies(1). Eyelid abnormalities, including ptosis, fatigability and weakness of eyelid closure, are the most prominent neuro-ophthalmic signs. Pupil-sparing, ophthalmoparesis in any pattern, may also occur(2). An association of myasthenia gravis and autoimmune thyroiditis has been reported in adults(3-10), but rarely in children(11).

Correspondence to: Kitthisak K, Department of Ophthalmology, Faculty of Medicine, Khon Kaen University, Khon Kaen 40002, Thailand. Phone: 0-4334-8383, Fax: 0-4334-8383, E-mail: Kitthisak@hotmail.com

J Med Assoc Thai Vol. 88 Suppl. 9 2005  S131
Thyroid function tests were normal but 1:1,600 serum titers of anti-thyroglobulin, and anti-microsomal antibodies were detected.

This patient was initially treated with oral pyridostigmine 60 mg tid. Since a residual ptosis persisted, the authors prescribed 60 mg pyridostigmine tid and 1 mg of prednisolone/kg/day. The ptosis resolved without any adverse reaction and the prednisolone was tapered off within 2 weeks. At the 12-month follow-up, the child was still on 60 mg of pyridostigmine tid without recurrence. The abnormal head position to compensate for the vertical diplopia caused by the restricted right inferior rectus persisted. Orthophoria and good binocular vision were achieved with this head position and the parents declined strabismus surgery.

Discussion
The presented patient had ptosis, which fluctuated with time and improved after the prostigmine test, signifying ocular myasthenia gravis. However, the incomitant hypertropia did not improve with the test and showed restriction during the forced duction test. The patient had mild exophthalmos and non-tender diffuse enlargement of the thyroid. The thyroid function tests were normal, though positive for anti-thyroglobulin and anti-microsomal antibodies, suggesting an association with autoimmune thyroiditis.

Myasthenia gravis has been associated with other autoimmune disorders such as: scleroderma (3), primary biliary cirrhosis (4), pernicious anemia (7,9), insulin dependent diabetes mellitus (9), polymyositis (11), Graves' disease (10), and autoimmune or Hashimoto's thyroiditis (3-11). A common cell-mediated immune disorder has been postulated (4).

Autoimmune thyroiditis, the most common cause of thyroid disease in children and adolescents, is diagnosed by diffuse thyroid enlargement and the presence of anti-thyroglobulin and anti-microsomal antibodies (7). This disorder is more common in girls, particularly after 6 years of age. The most common clinical manifestations are growth retardation and non-tender diffusely enlarged goiter with attendant hypothyroidism. Most affected children are clinically euthyroid and asymptomatic, but a small number may show hyperthyroidism. Some of the clinically euthyroid children may have laboratory evidence of hypothyroidism. A long-term follow-up of the thyroid function test was necessary for the presented patient, even though she was clinically and laboratory euthyroid.

Autoimmune thyroiditis and myasthenia gravis can be considered pathogenetically related because both cause abnormal T-lymphocyte function, so the autoimmune response is directed against cell membrane receptors (10). Cross-reactivity seems to be a property of pathological autoantibodies (12). Occasionally, auto-immune thyroiditis may coexist with Graves' disease (13-14); however, ophthalmopathy may occur without evidence of Graves'.

Diplopia without obvious exophthalmos, caused by infiltrative endocrine ophthalmopathy, has been reported in patients with clinically and laboratory confirmed autoimmune thyroid disease. Vertical diplopia is the most common manifestation of the disorder: this is caused by hypotropia secondary to restrictive tightening of the inferior rectus muscle (15). The presented patient stared with the right eye, and this secondary deviation made the left hypertropia more prominent.

The association of ocular myasthenia gravis and autoimmune thyroiditis in children was reported by Tsao et al (11). They reported that their pediatric patient, like an adult, developed generalized myasthenia gravis within two years. The authors, therefore, intend to follow-up the patient for two years.

Pyridostigmine is the first-line therapy in pediatric ocular myasthenia, but just as in adults, the results are often unsatisfactory. Oral corticosteroids have been suggested as an additional control, but the long-term effects of corticosteroids on bone growth must be kept in mind.

The presented patient represented a good candidate for various associated autoimmune diseases, including autoimmune thyroiditis in children with myasthenia gravis. The forced duction test was the best tool to differentiate between a restrictive vs. a paralytic cause of the incomitant strabismus.

Acknowledgements
The authors wish to thank Mr. Bryan Roderick Hamman for assistance with the English-language presentation of the manuscript.

References