



ENDGAMES

CASE REVIEW

Ptosis that resolves with application of an ice pack

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A man in his 50s presented with a one week history of left sided ptosis and difficulty swallowing. The ptosis was absent on waking, worsened during the day, and improved after rest. His medical history included hypertension. Apart from bilateral ptosis that was worse on the left (fig 1), clinical examination was unremarkable. Application of an ice pack to the left eye for five minutes led to improvement of the ptosis on that side. The ptosis recurred 20-30 minutes after the ice pack was removed.



Patient on presentation and after application of ice pack

Within two weeks of the initial presentation, the patient started to experience difficulty chewing, dysphagia for food and saliva, dysarthria, fatigue, and generalised proximal weakness.

Anti-acetylcholine receptor (anti AChR) autoantibody titre was noticeably increased, at 581×10^{-10} mol/L (normal range 0.5×10^{-10} mol/L).

Single fibre electromyography showed increased jitter of the left orbicularis oculi muscle, and repetitive nerve stimulation showed reduced compound muscle action potential of the nasalis muscle.

Question

What is the most likely underlying diagnosis?

Answer

Generalised myasthenia gravis.

Most patients have unilateral eye involvement, but the condition can present bilaterally.

Diagnosis is confirmed by raised anti-AChR autoantibody titres, reduced compound muscle action potential on repetitive nerve stimulation, and increased jitter on single fibre electromyography.

Myasthenia gravis has localised and generalised forms:

- Ocular myasthenia gravis: ptosis and diplopia only.
- Oculo-bulbar myasthenia gravis: episodes of respiratory involvement with ptosis and diplopia.
- Generalised myasthenia gravis: generalised proximal weakness; in 85% of cases it is associated with anti-AChR antibodies.

Ptosis that is absent on waking, worsens as the day progresses, and improves with rest corresponds with the typical pattern of fatigability of all forms of myasthenia gravis. The ice pack test is positive in most patients who have ptosis caused by myasthenia gravis—sensitivity is 77-100% and specificity is 88-100%, implying a 0-12% chance that the result will also be positive in patients with non-myasthenic ptosis.¹⁻⁸

In myasthenia gravis with ocular manifestations, cooling is believed to reduce acetylcholinesterase activity by making more acetylcholine available at the neuromuscular junction thereby reversing the ptosis (in non-myasthenic ptosis there is no acetylcholine deficiency at the neuromuscular junction).

Ten per cent of patients who test positive for anti-AChR antibodies will have thymoma.¹

Causes of unilateral non-myasthenic ptosis include Horner's syndrome, oculomotor nerve palsy, and levator aponeurosis

dehiscence (unilateral ptosis). Causes of bilateral non-myasthenic ptosis include myotonic dystrophy and oculopharyngeal muscular dystrophy.

Patient outcome

The patient underwent computed tomography of the chest, which did not show thymoma. He was successfully treated with pyridostigmine and prednisolone.

Learning points

- Marked fatigability is characteristic of all forms of myasthenia gravis.
- At presentation, most patients with ocular or generalised myasthenia gravis or both have asymmetrical involvement of the eyes
- If a patient presents with ptosis, consider the ice pack test.

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Patient consent obtained.

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