Commentary: Ocular myasthenia gravis: Interpreting the investigations

In this paper, the authors have assessed a very important aspect in the clinical diagnosis of ocular myasthenia.[1] Typically, when evaluating a case of ocular myasthenia, clinicians are tempted to order a battery of tests at one go with the hope that at least one of them would show up a positive finding, thus confirming the diagnosis.[2] This is because the different tests for myasthenia (clinical and laboratory) have varying sensitivity and specificity. In their study, the authors have assessed the diagnostic accuracy of forced eyelid closure test (FECT), ice pack test (IPT), repetitive nerve stimulation (RNS) test, and acetyl choline receptor (AChR) antibody test in patients with suspected ocular myasthenia. The FECT is a relatively unknown test which the authors have investigated. This test assesses the fatigability of the orbicularis muscle compared to the traditional "fatigability test" which induces fatigue in the levator muscle on prolonged upgaze.

One of the important takeaways of this paper is the results of the clinical tests, namely, the FECT and the IPT. In patients in whom AChR antibodies and RNS tests were negative, the specificity of combined FECT and IPT (both positive) was 100% and the sensitivity and specificity of FECT alone was 100% and 80%, respectively. This gives us strong evidence that in spite of negative laboratory investigations, solely based on the history and clinical findings, a therapeutic trial is recommended in such patients.

The strengths of this paper include the follow-up period, which is 36 months. In this study, 19% (13 patients) of the patients who presented with ocular myasthenia progressed to generalized myasthenia despite early therapy that included corticosteroids. Of these 13 patients, 11 patients were positive for AChR antibodies at the time of diagnosis. This indicates that presence of AChR antibodies is a strong prognostic factor in predicting progression to generalized myasthenia. It has been suggested that in these patients, chest imaging and RNS should be routinely performed to assess the risk of generalization.[3] However, this progression of ocular myasthenia to generalized myasthenia does not represent the true natural history of the disease since all patients received therapy including corticosteroids, which are known to decrease progression of ocular to generalized myasthenia gravis.[4] The authors are encouraged to report the findings after longer follow-up period as it is known that while some patients attain maximal improvement on corticosteroids in the first 6 months, others may take as long as 2 years or more.[5]

While performing the IPT, bilateral simultaneous orbital cooling using thin surgical gloves filled with ice cubes is recommended as opposed to commercially available ice packs, as icepacks are associated with lower sensitivity.[6,7] Furthermore, the authors reported that patients with isolated diplopia showed no response to IPT after 2 min. While the 2-min cut-off is acceptable for patients with ptosis alone, increasing the duration of the ice application for 5 min has a reported sensitivity of 76.9% while detecting myasthenia in patients presenting with diplopia.[7] In addition, it would have been useful to know how many patients had episodes of myasthenic crisis during the course of the 36-month follow-up period.

In summary, the diagnosis, therapy, and eventual prognosis of ocular myasthenia are largely based on the findings of clinical and laboratory investigations, which need to be performed in standardized ways and interpreted appropriately.

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