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Clinical Study

Inferior oblique muscle paresis as a sign of myasthenia gravis

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ABSTRACT

Myasthenia gravis may affect any of the six extra-ocular muscles, masquerading as any type of ocular motor pathology. The frequency of involvement of each muscle is not well established in the medical literature. This study was designed to determine whether a specific muscle or combination of muscles tends to be predominantly affected. This retrospective review included 30 patients with a clinical diagnosis of myasthenia gravis who had extra-ocular muscle involvement with diplopia at presentation. The diagnosis was confirmed by at least one of the following tests: Tensilon test, acetylcholine receptor antibodies, thymoma on chest CT scan, or suggestive electromyography. Frequency of involvement of each muscle in this cohort was inferior oblique 19 (63.3%), lateral rectus nine (30%), superior rectus four (13.3%), inferior rectus six (20%), medial rectus four (13.3%), and superior oblique three (10%). The inferior oblique was involved more often than any other muscle ($p < 0.01$). Eighteen (60%) patients had ptosis, six (20%) of whom had bilateral ptosis. Diagnosing myasthenia gravis can be difficult, because the disease may mimic every pupil-sparing pattern of ocular misalignment. In addition diplopia caused by paresis of the inferior oblique muscle is rarely encountered (other than as a part of oculomotor nerve palsy). Hence, when a patient presents with vertical diplopia resulting from an isolated inferior oblique palsy, myasthenic etiology should be highly suspected.

1. Introduction

Myasthenia gravis (MG) is an autoimmune disease of neuromuscular transmission. Involvement of extra-ocular muscles (EOM), causing ptosis and diplopia are frequent presenting and persistent features. Up to 75% of patients with MG complain of diplopia at presentation, and this rises to 90% throughout the course of the disease [1–3].

Bilateral involvement and weakness of horizontal and vertical eye movements are typical [4]. However, despite the importance of early diagnosis, it is often delayed because EOM involvement does not follow a certain pattern and may mimic other conditions [3]. Since the spectrum of EOM conditions is wide, any acquired ocular motility disturbance, with or without ptosis but with normally reacting pupils, should raise a clinical suspicion of MG [4]. We report our observation of the pattern of EOM involvement among patients with MG.

2. Material and methods

A retrospective chart review of all consecutive MG patients with EOM involvement seen by a single neuro-ophthalmologist (Y.A.) was conducted. Only patients in whom the neuro-ophthalmological examination documented involvement of a specific EOM with diplopia at presentation were included.

The diagnosis of MG was made on the basis of immunological, neurophysiological and clinical findings. Diagnostic tests included anti-acetylcholine receptor antibodies (anti-AChRAb), Tensilon test and single-fiber electromyography of the frontalis muscle, although not all patients underwent every investigation.

The pre-surgical evaluation included a full slit lamp examination, visual acuity and a full neuro-ophthalmic examination, including ptosis workup. Measurements of the ocular movements were based the horizontal and vertical deviation in all positions of gaze using Maddox rod and prism diopter measurements. The ptosis workup included measurements of the marginal reflex distance (MRD), levator function, the palpebral fissure, and the presence of Bell's phenomena. Ptosis was considered in cases of MRD less than 2 mm, or differences of more than 1 mm between eyes. The study was approved by the Meir Medical Center Institutional Review Board.

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2.1. Statistical analysis

Fisher's exact test was used to evaluate differences between groups. Results were considered significant when the p value was < 0.05. Analysis was done using the Statistical Package for the Social Sciences version 21 software (IBM, Armonk, NY, USA).

3. Results

A total of 30 patients (24 males, 80%), with a mean age of 64.8 (range 38–82) years with a clinical diagnosis of MG were included. MG was confirmed by at least one additional positive test: Tensilon test (24%), anti-AChRAb (43.3%), thymoma on chest CT scan (10%) or suggestive electromyography (20%).

Frequency of involvement of each muscle with diplopia at presentation in this cohort was as follows: inferior oblique (IO), 19 patients (63.3%); lateral rectus, nine patients (30%); superior rectus, four patients (13.3%); inferior rectus, six patients (20%); medial rectus, four patients (13.3%); and the superior oblique three patients (10%). The IO was involved more often than any other muscle ($p < 0.01$). Ptosis was observed in 18 (60%) patients with EOM involvement (Table 1).

3.1. Representative case report

A 73-year-old male was seen in the neuro-ophthalmology unit due to complaints of vertical diplopia for the past 2 weeks. MG had been diagnosed 4 years previously when he presented with left eyelid ptosis. Myasthenia was confirmed clinically as well as on laboratory tests, at that time anti-AChRAb level was 14.1 pmol/ml (normal range 0–0.5 pmol/ml). He had no other signs of muscular weakness and his ptosis was successfully treated with pyridostigmine tablets. Soon after thymoma was detected and excised from his chest. For 3 years he was free of myasthenic symptoms though untreated, until he presented to our neuro-ophthalmology clinic with double vision.

On examination his visual acuity in the right eye was 20/25 and 20/30 in the left. There was 2 mm ptosis of the left lid with good levator function and clear fatigue on up gaze. There was a full range of eye movement in all directions of gaze but the cover test revealed left hypotropia, increasing on right and up gaze. Measurements of the vertical deviation with Maddox rod and prism bar revealed two prism-diopter of left hypotropia on primary position, 10 on right gaze, two on left gaze, 16 on upgaze, one on downgaze, five on right head tilt and two on left head tilt. This pattern of incomitance clearly suggests weakness of the left IO muscle.

4. Discussion

IO muscle palsy is rarely encountered, other than as a part of oculomotor nerve palsy. It is the least common isolated EOM palsy [5]. The diagnosis of isolated IO muscle palsy is controversial, because the anatomic design of the third nerve nucleus makes it unlikely that only the IO muscle could be affected [6]. Some have suggested that it is caused by either brain stem infarction, involving the oculomotor fascicular fibers [7,8] or from early embryologic damage that could partially spare the Edinger–Westphal nucleus [9]. Other authors have interpreted the clinical picture compatible with isolated IO muscle palsy as superior oblique muscle overaction, skew deviation [6] or pulley heterotopy [10].

However, several authors have reported patients with findings suggestive of isolated IO muscle palsy [6,11–13]. Ela-Dalman et al. used MRI to demonstrate reduced IO muscle size in patients with clinically diagnosed IO muscle palsy, supporting the concept of isolated IO muscle weakness [14].

Table 1
Demographic and clinical details of patients with myasthenia gravis

No	Age	Sex	EMG	AChRAb	Tensilon	Thymoma	Ocul/Syst	IO	IR	SR	LR	MR	SO	Ptosis
1	63	M		3.9			Syst				L			
2	64	M		44			Syst						R	R
3	62	M	+	1.3			Syst	L						R
4	76	M		21				L						BE
5	44	F		11.6		+	Syst	L						
6	77	M		10.9								BE	L	
7	44	F			+		Syst	BE	BE	BE	BE	BE	BE	BE
8	70	M	+				Syst		L					
9	63	M	+				Syst		R	R		R		
10	28	M		14.4		+	Syst	L	L		L			R
11	75	F		49.1	+			L						BE
12	57	M	+	1.2	+		Syst			R				R
13	78	M	+				Syst	L			L			
14	67	M		10.9						R				R
15	80	M			+		Syst	R	R					BE
16	56	M			+			L						L
17	61	M	+		+		Syst	L						
18	65	M		2.6	+			R						L
19	80	M		14.1		+	Syst	L			L			L
20	38	F		248			Syst				R			
21	80	M			+				L			L		BE
22	69	F						L						BE
23	47	F						BE			L			
24	55	M									L			R
25	68	M		5.3							BE			L
26	74	M					Syst	R						
27	71	M					Syst	L						L
28	82	M						L						
29	75	M						R						
30	76	M											L	R

AChRAb = acetylcholine receptor antibodies, BE = both eyes, EMG = electromyography, F = female, IO = inferior oblique paresis, IR = inferior rectus paresis, L = left, LR = lateral rectus paresis, M = male, MR = medial rectus paresis, Ocul = ocular, R = right, SO = superior oblique paresis, SR = superior rectus paresis, Syst = systemic, + = present.