Cysticercosis of midbrain presenting with fluctuating ptosis

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Abstract

Fluctuating ptosis is usually caused by Myasthenia gravis. There are a few case reports of central causes of fluctuating ptosis. A 58-year-old man presented with fluctuating ptosis of one year duration. He was diagnosed as having ocular myasthenia and investigated. On evaluation, his electrophysiological tests revealed negative decremental response and results of neostigmine test was negative. During follow-up, patient developed headache. Imaging of the brain revealed midbrain cysticercosis granuloma. The focal encephalitis and edema was responsible for fluctuating ptosis. It is therefore essential to be aware of conditions that cause such pseudomyasthenic features. Patients with ptosis need to be evaluated for other rare central cause especially when neostigmine test is negative.

Key Words

Midbrain, myasthenia, neurocysticercosis, pseudomyasthenia, ptosis

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Introduction

Ocular weakness presenting as fatiguable ptosis especially when it is asymmetrical is the hallmark of Myasthenia gravis (MG), seen in 85% patients.[1] The initial ocular symptom usually becomes generalized within 2 years in MG. Several case reports have documented some rare conditions, which cause ptosis, and it has been suggested that when myasthenia is limited to the ocular or cranial musculature then imaging of the brain needs to be done to rule out other rare central causes of ptosis. We report a patient with fluctuating ptosis who was initially suspected to have MG and on evaluation central cause was found.

Case Report

A 58-year-old male patient with diabetes of twenty five years duration, presented with fluctuating ptosis of one year duration. There was neither associated diplopia nor diurnal variation. On examination, he had bilateral ptosis at rest, which was fatiguable in nature. Extraocular movements were full in all directions with normal fundus examination. Rest of the appendicular examination was within normal limits. Due to its fatiguability, first possibility of ocular myasthenia was considered. Hence, initial evaluation was carried out to look for myasthenia. He had absent decremental response on repetitive nerve stimulation in nasalis, orbicularis oculi, trapezius, and abductor digiti minimi muscles. His acetyl choline receptor antibody (ACH-R Ab) was negative and had normal thyroid functions. There was no improvement in ptosis with 1.5 mg of neostigmine. One month later during follow-up patient developed headache, which was mainly bifrontal in location, not associated with nausea, vomiting, or visual aura. He then underwent magnetic resonance imaging (MRI) of the brain due to the presence of headache to rule out any intracranial pathology. A cystic lesion was documented in the midbrain (3 tesla strength with CISS-3D sequence and gadolinium contrast) with scolex [Figure 1] suggestive of neurocysticercosis (NCC), thus fulfilling the Del Brutto criteria for NCC.[2] Intraocular cysticercosis was ruled out, and then he was treated with short course of oral corticosteroids (40 mg of Wysolone for ten days followed by gradual tapering over 4 weeks). Seven days of Albendazole was also given during the second week of corticosteroids. The ptosis and headache improved completely over ten days. But within two weeks after stopping the steroids, the ptosis recurred and he was again started on low dose corticosteroids (20 mg of Wysolone) following which his ptosis improved.

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Discussion

The fatiguable ptosis in this patient obscured the diagnosis. The first and foremost diagnosis in fatiguable ptosis is MG. Hence, patient was evaluated for the same. MG is an autoimmune disorder presenting with fatiguable ptosis, diplopia with diurnal variation. Around 85% of patients present with ocular

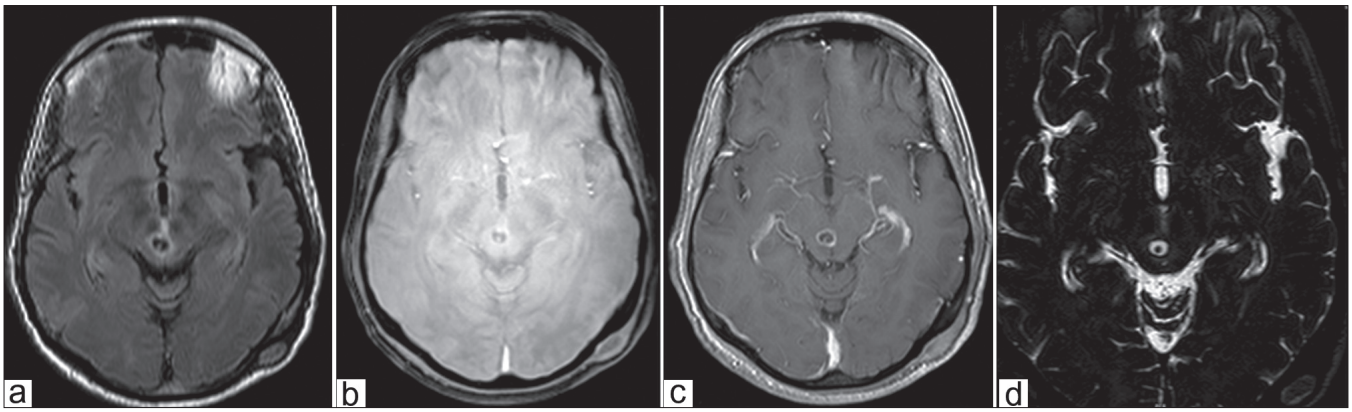

Figure 1: MRI brain (a) T1-weighted sequence showing hyperintense rim with scolex showing, (b) Diffusion-weighted sequence showing scolex within the ring lesion, (c) T1-weighted image with gadolinum enhancement showing contrast enhancing ring and scolex and (d) Constructive Interference Steady state (CISS) 3D sequence showing the cystic lesion

features and nearly 30% of them progress to generalized MG within two years.[1] The initial evaluation of a fatiguable ptosis includes electrophysiological testing; repetitive nerve stimulation to rule out any neuromuscular junction defects, and the clinical response to neostigmine. The ACH-R Ab is positive in 55% cases of ocular myasthenia.[3] All these tests were negative in this patient warranting further evaluation of ptosis. Many case reports suggest several rare causes of ptosis with Pseudomyasthenia-like picture observed in conditions like carotid artery aneurysm,[4] basilar artery aneurysm,[5] NCC,[6-9] midbrain glioma,[10-12] hematoma, metastases,[13] and Wernicke's encephalopathy.[14] Similarly our patient had NCC in the brain, which was confirmed by neuroimaging with MRI that documented the scolex within the cystic lesion, thus fulfilling the Del Brutto criteria for NCC.[2]

NCC is the most common parasitic infestation of the nervous system.[5] It is caused by the larval stage of the tapeworm taenia solium.[15] The various clinical manifestations of NCC are protean depending on the size, location, and meningeal involvement. It usually presents with seizures. Other various CNS clinical manifestations include headache, hydrocephalus, chronic meningitis, focal neurological deficits, dementia and psychiatric manifestatons.[16] Involvement of third nerve is rare especially due to a parenchymal lesion.[6] The treatment of NCC is debatable. The treatment guidelines depend on the location, size, and number of lesions. There are very few case reports of such midbrain NCC. Focal encephalitis and edema was responsible for fluctuating ptosis in our patient. As reported in few reports earlier, our patient was also treated with corticosteroids and antihelminthic therapy. Recurrence of symptoms warranted re-introduction of steroids, which the patient tolerated. Treatment with corticosteroids especially in elderly diabetics is not accepted by many physicians due to its side effects profile. But as seen in other cases it has been the only proven drug to reduce inflammation and edema. These people may need long term treatment with steroids as documented by Kim *et al*.[6]

Ptosis is due to the dysfunction of one of the upper eyelid elevator muscles. These elevator muscles are levator palpebrae superioris with its aponeurosis (supplied by third cranial nerve) and the muller muscle (sympathetic innervation). The lesion in such midbrain myasthenia is due to the involvement of the central caudal nucleus (CCN), which is a small subgroup of oculomotor nucleus (in the caudal portion) containing subnucleus for levator palpebrae superioris.[17] Involvement of this nucleus results in complete, bilateral ptosis. There are few reports that suggest that bilateral ptosis may also be caused by a single lesion of the supraoculomotor area located immediately dorsal to the oculomotor complex.[17]

We propose a more detailed evaluation of patients with fatiguable or fluctuating ptosis. In those patients where the electrophysiological results are negative and/or ptosis is restricted to ocular muscle without any other neuromuscular involvement, cranial imaging needs to be done, preferably MRI of the brain with contrast studies that may show lesions in the midbrain.

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