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RARE ASSOCIATION OF MYASTHENIA GRAVIS AND POLYMYOSITIS: A JUVENILE CASE REPORT

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Abstract:

Polymyositis is the chronic inflammation of the muscles, mostly involving the proximal musculature. Myasthenia Gravis is characterized by fluctuating muscle weakness, with improvement of strength after rest. Juvenile Myasthenia Gravis presents before the age of 18. Polymyositis and Myasthenia gravis rarely exist together. Only few cases of this association

have been reported so far in adults, while only one such case has been reported in adolescents. We present a 13-year-old patient with 2-year history of proximal muscle weakness and easy fatigability with bilateral ptosis since childhood. Clinical features, examination, laboratory findings, Electromyography (EMG), muscle biopsy and acetylcholine receptor antibodies of patient were in favor of the overlap. The patient improved clinically with pyridostigmine, prednisolone and methotrexate.

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INTRODUCTION:

Myositis include polymyositis (PM), dermatomyositis (DM) and, inclusion body myositis. It causes chronic inflammation in symmetric proximal musculature resulting in weakness [1]. An approximated prevalence is reported to be around 5 to 22 per 100,000 persons [2]. Patients with myositis may also have other autoimmune diseases such as Systemic Sclerosis, Sjogren's syndrome, or Systemic lupus erythematosus. It is very important to recognize the overlapping conditions, as change in management strategies might be needed [3].

Although rare, myositis and myasthenia gravis (MG) may present as an overlap. This overlap is reported in less than 3% of myasthenic patients [4]. In MG, autoantibodies against acetylcholine receptor (AChR) at the post-synaptic membrane, reduce the number of receptors available, disrupting neuromuscular transmission and causing muscle weakness [3]. Juvenile Myasthenia Gravis (JMG) is defined as myasthenia gravis in children younger than 18 years of age [5]. The occurrence of myasthenia gravis in childhood is strongly influenced by genetic and environmental factors [6].

Patients with myositis usually have constant weakness, on the other hand patients with MG have weakness that worsens with activity and as the day progresses. In the majority of MG patients, the ocular muscles are involved first, causing intermittent diplopia and ptosis. These symptoms are not typically observed in myositis [3]. The diagnosis of MG can be made by detecting antibodies recognizing the AChR in the blood. Specialized electrophysiological testing, such as repetitive nerve stimulation (RNS) can also be used. While myositis is mostly diagnosed by Creatinine kinase (CK) elevation, Electromyography (EMG) and muscle biopsy [3].

Here, we report a juvenile case of Myasthenia Gravis and myositis overlap, also known as myositis-MG association.

CASE PRESENTATION:

A 13-year-old male patient presented to neurology department with difficulty in standing from sitting position and raising arms above head. He had bilateral ptosis since childhood and his milestones were delayed. He often complained of easy fatigue and could not play outdoor games. An initial work-up showed Acetylcholine receptor antibodies as positive. He was treated on the lines of myasthenia gravis to which he responded partially.

On that note, rheumatology review was sort. On interviewing it was revealed he had muscle weakness with gradual worsening over a period of three years. Initially, it was at the end of the day. He described increasing difficulty climbing stairs, getting up from chair or picking up heavy objects, brushing hair. There was no history of dysphagia, cough, photosensitivity, rash, no fever at onset of weakness or weight loss. On examination he had bilateral ptosis with proximal muscle weakness of both upper and lower limbs. There was decreased muscle power but, no pain during the movements of involved musculature. However, on palpation patient reported soreness of the proximal upper and lower limbs but no signs of joint involvement, and no nail fold capillary abnormalities. On investigations, his Creatinine kinase levels were elevated and consistent with polymyositis. MRI, Electromyography (EMG) (Figure 1), and Muscle biopsy also confirmed the diagnosis of polymyositis. On CT- scan chest, there was no thymoma. Repetitive Nerve Stimulation (RNS) demonstrated decremental response as shown in figure 2.

The final diagnosis was polymyositis overlap with myasthenia gravis. He was treated with prednisolone 40 mg/day, methotrexate 15 mg/week along with folic acid and pyridostigmine.

On follow-up, his muscle power improved, and muscle enzymes level declined.

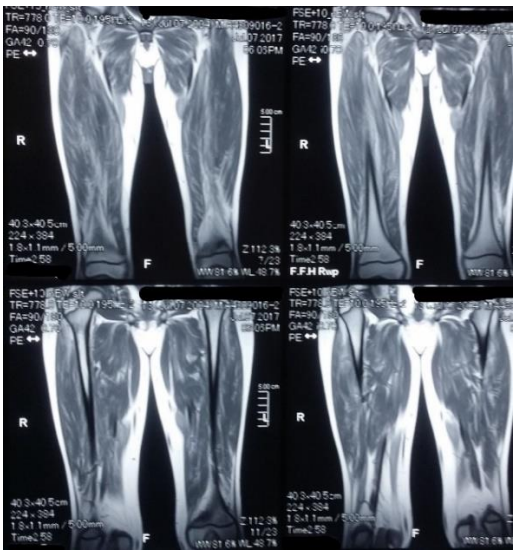

Figure 1: Coronal MRI of Thighs: Muscle Edema along with atrophic changes involving both thighs. MR features are suggestive of acute on chronic polymyositis.

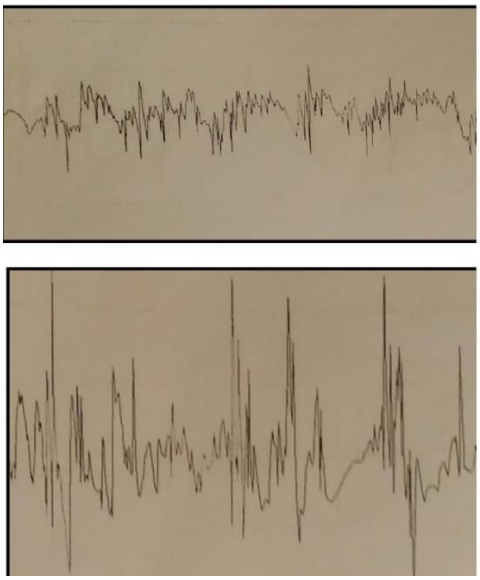

Figure 2: EMG Right Deltoid: Demonstrate fibrillation and sharp waves suggestive of myositis.

DISCUSSION:

The myositis-MG is a rare entity to be found. Such associations have been identified in less than 50 cases so far [4]. Interestingly, only one similar case of juvenile myositis-MG overlap has been observed in the literature [7]. Our case didn't have any other autoimmune disorder beside the association. A case-series reported that, in 67% of their patients an additional overlap with other autoimmune diseases like ulcerative colitis, Sjogren's, scleroderma, thyroid disease and autoimmune hepatitis was observed [3]. That may point towards an autoimmune link in myositis-MG overlap.

Diplopia and ptosis, as seen in our patient, are classic features of MG that are never encountered in patients with myositis alone. In a case series reporting this association, about half of the subjects had either ptosis or diplopia [3]. The most important clinical feature of Inflammatory myositis is symmetric proximal muscle weakness. It is presented as difficulty in lifting heavy objects, getting up from sitting position, climbing stairs, and overhead abduction of the arm [2]. In addition to muscle weakness, patients with dermatomyositis (DM) often present with a skin rash that include Gottron papule and heliotrope rash. Dermatomyositis is bimodal with respect to age distribution with one peak at 5 to 15 years and another

at 45 to 60 years [2]. On the other hand, Polymyositis rarely occurs in the pediatric age group, and the mean age is between 50 to 60 years [2]. Interestingly, Patient in our case report had polymyositis. It is rare finding this association let alone with polymyositis which is seldom to be seen in adolescents. However, another case of the MG-myositis association has been reported in a 14-year-old girl who also presented with polymyositis, followed by development of generalized myasthenia gravis 2 years later [7].

In previous cases with myositis-MG overlap, biopsy findings of the muscle were suggestive of polymyositis in 63% of the cases while dermatomyositis 25% and granulomatosis in about 12% [4]. A cohort discussing 13 patients with this association have demonstrated that, in 10 cases the onset of both pathologies was simultaneous, while in two cases MG occurred before myositis and in one case myositis developed before MG. In our case, MG developed in childhood and polymyositis at the age of 10 [4].

Our patient didn't have thymoma on CT but Thymus pathology is quite frequently associated with MG. It was reported in a cohort that 10 out of 13 patients had thymus pathology [4]. Thus mediastinal imaging either by CT or MRI is very important in evaluating patient with myositis-MG [3].

Management of MG comprises of symptomatic treatment with anticholinesterase. All patients with MG may not respond to this medication, but in a case series describing myositis-MG overlap, 83% patients had improvement with pyridostigmine[3]. As in myositis, steroids and immunomodulators along with IVIG are commonly used in the management of patients with MG. It is recommended that treatment for MG be commenced with low dose steroids and then titrated up as opposed to high dose with subsequent tapering, in order to prevent further muscle weakness associated with a higher dose regimen.

CONCLUSION:

Myositis-MG is a rare association, even more infrequent in adolescents. We suggest that patients should be interrogated for the duration and fluctuating nature of the weakness so that the overlap can be easily diagnosed. Any patient with partial response to treatment of myasthenia gravis, should be assessed for inflammatory myopathies. Given that the approach to management may be significantly different in patients with MG versus myositis, it is important to recognize when patients may have an overlap of these two conditions. Treatment may involve low dose steroids, methotrexate and pyridostigmine.

Learning points:

- Myasthenia Gravis always presents with fluctuating muscle weakness. The initial symptoms may be limited to diplopia or ptosis while myositis have muscle weakness that is present constantly.

- Patients with myositis-MG overlap require a different management protocol as opposed to the approach followed by these when they present separately.

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