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Two cases of inclusion body myositis presenting with unusual symptoms, head drop and facial diplegia and different responses to intravenous immunoglobulin treatment

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Inclusion body myositis (IBM) is a challenging type of myopathy due to insidious course, mixed electromyographic (EMG) findings and unfavorable response to treatment. Although the majority of patients have classic symptoms at onset including progressive asymmetric quadriceps muscle wasting and weakness of the proximal and distal upper extremities, some have atypical presentations consisting of a prominent facial or cervical muscle weakness.[1] These unusual manifestations may be confused with other myopathies and even motor neuron disease.

The present article described two patients with IBM presented with rare symptoms, including severe facial diplegia (case 1) and isolated head drop (case 2). Both patients previously misdiagnosed with other neuromuscular disorders due to their uncommon presentations. IBM ultimately diagnosed based on their conclusive muscle biopsy although they had different responses to intravenous immunoglobulin (IVIG) treatment. In this study, we tried to address the wide clinical spectrum of IBM and discuss the benefit of early diagnosis in treatment of the eligible patients.

Patient 1: A 51-year-old woman referred to our clinic with dysphagia and drooling.

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She reported a history of progressive bilateral facial weakness since three years ago which followed by dysphagia a year later and caused significant weight loss. from six months ago, she experienced clumsy hands and difficulty of climbing stairs. Lately amyotrophic lateral sclerosis (ALS) was diagnosed for the patient according to the dysphagia, muscle wasting and irritative neurogenic electromyographic finding and Riluzole was started subsequently. On neurologic examination severe symmetric facial palsy was prominent including disability to closing the eyes, elevating eyebrows, smiling and rounding lips to whistle. Neck flexion was partially weak (force 3/5). A mild weakness of distal upper and proximal lower limbs (force 4/5) and slight asymmetric atrophy of quadriceps muscles detected. Deep Tendon Reflexes were normal. Creatine kinase (CK) was slightly elevated up to 650. Electromyographic study showed mixed neurogenic and myogenic changes in upper and lower limbs with irritative features. Muscle biopsy of left vastus lateralis revealed marked atrophy with presence of multiple necrotic and degenerative/regenerative fibers associated with multiple foci of endomysial chronic inflammation as well as partial invasions. Based on the histopathologic findings the diagnosis of inclusion body myositis was made (Figure 1). She received IVIG for three courses; however, medication discontinued because of no significant improvement.

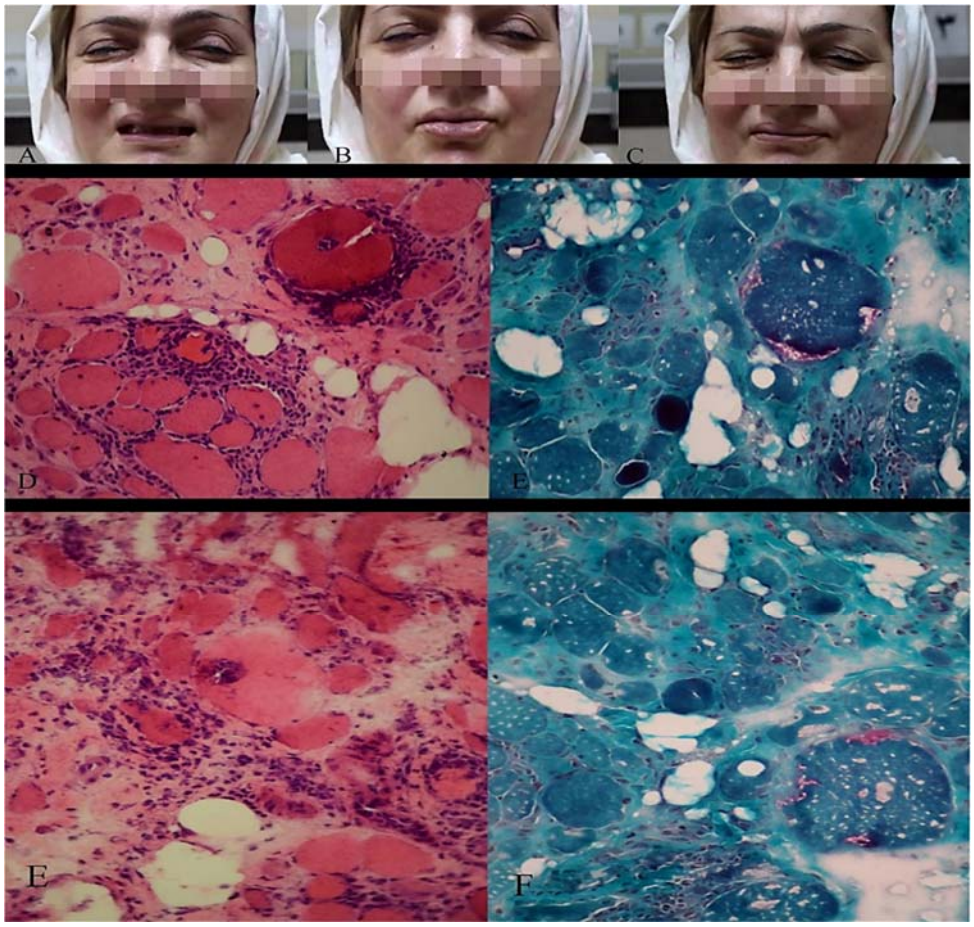

Figure 1. Inclusion body myositis (IBM) with severe facial diplegia
A: Hematoxylin-eosin (H&E) staining, marked atrophy with presence of multiple necrotic and degenerative/regenerative fibers (arrow head) and endomysial chronic inflammation (arrow) associated with prominent adipose tissue replacement. B: Modified Gomori trichrome staining, red-rimmed cytoplasmic vacuoles (white arrow) no congophilic inclusion was found on Congo red stained slide. No cytochrome oxidase (COX)-negative fiber was identified. IBM with isolated dropped head. C: Inflammatory myopathy in hematoxylin-eosin staining with, D: rare red-rimmed vacuoles on Modified trichrome stain (white arrow). Neither congophilic inclusion, nor Cox-negative fiber was seen

Patient 2: A 58-year-old man was referred with dropped head to consult. The patient described his symptom started from 6-month ago, whilst a mild neck weakness developed into complete head drop in few weeks and forced him to use artificial neck support. He also complained of dysphagia during this period which caused noticeable weight loss. He complained of no other muscle weakness. The patient had been treated with prednisolone in favor of polymyositis for 3 months, although no suggestive muscle biopsy or electromyographic support were documented. No clinical improvement reported by the patient after corticosteroid therapy and medication discontinued. In neurologic examination, neck extension force was completely lost and neck flexion force was 2/5. Extremity muscles were all intact. CK level was slightly elevated up to 297. In needle examination, severe myogenic changes with mild irritative features detected only in cervical paraspinal muscles and other regions were spared. Biopsy of the involved muscles revealed inflammatory myopathy with rare red-rimmed vacuoles on Modified trichrome stain. The diagnosis of inclusion body myositis was suggested in case of clinical correlation (Figure 1). The patient treated with IVIG (total dose 2 g/kg) and according to remarkable improvement of both symptoms, he periodically received a maintenance dose (1 g/kg every month) for several months. In a follow up of 18 months, patient was able to hold up his head unassisted although difficulty in neck flexion and intermittent dysphagia to solid food were still complained. Moreover, no additional limb weakness was detected.

Discussion

In general, about one third of patients with IBM illustrated to have mild facial weakness including incomplete burying of the eyelashes on forceful eye closure. Severe weakness with incomplete eye closure have been observed in a few patients with very advanced disease and prominent early involvement has been reported twice in previous studies.[2,3] likewise, neck extensor weakness, causing dropped head, develops with more extended IBM[4] and it has been reported minimally in the literature as a presenting symptom.[5]

Another topic is the favorable response to IVIG in our second case. Due to the rare nature of the disease, the best treatment modality is unknown in IBM. Some experts have suggested the relative benefit of IVIG for selected patients with prominent bulbar involvement.[6,7] There is a positive response to IVIG in a 59-year-old woman diagnosed with IBM with head tilting forward and dysphagia (similar to the second case of ours) in previous reports. The author proposed that following unsuccessful prednisone therapy, repeated intravenous administration of IVIG improved the posture of neck and dysphagia disappeared.[8]

There are two potential explanations for good response to IVIG in these patients with head drop; First, would be the short "symptom to treatment" course. This is unlike to the most cases of IBM that symptoms progressed for years before proper diagnosis, therefore lots of muscle fibers undergone degenerative process and replaced by adipose tissue. As the muscle staining of our patients shown, the amount of necrotic and adipose tissue in the second case are less prominent in comparison to the first case (6 months course versus 36 months course respectively). Second, we hypothesize that different muscular zone involvement in the cervical region in compare to extremity muscles, could be responsible for the favorable outcome after IVIG treatment in both bulbar and head drop cases and requires further complementary evidence.

In conclusion, in this study, we attempted to highlight the clinical manifestations and histopathological findings of two IBM patients with rare presenting symptoms, to propose that these symptoms should be in the corner of mind in the evaluation of IBM. This would not only prevent from adding unnecessary costs to the patients and healthcare system by diminishing differential diagnosis, but also be as a promising hope for treatment of early diagnosed IBM cases presented with head drop syndrome in the future.

Conflict of Interests

The authors declare no conflict of interest in this study.

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References

1. Dimachkie MM, Barohn RJ. Inclusion body myositis. Neurol Clin 2014; 32(3): 629-46.
2. Ghosh PS, Laughlin RS, Engel AG. Inclusion-body myositis presenting with facial diplegia. Muscle Nerve 2014; 49(2): 287-9.
3. Cummins G, O'Donovan D, Molyneux A, Stacpoole S. Facial diplegia as the presenting

symptom of inclusion body myositis. Muscle Nerve 2019; 60(2): E14-E16.
4. Ma H, McEvoy KM, Milone M. Sporadic inclusion body myositis presenting with severe camptocormia. J Clin Neurosci 2013; 20(11): 1628-9.
5. Hund E, Heckl R, Goebel HH, Meinck HM. Inclusion body myositis presenting with isolated erector spinae paresis. Neurology 1995; 45(5): 993-4.
6. Cherin P, Pelletier S, Teixeira A, Laforet P, Simon A, Herson S, et al. Intravenous immunoglobulin for dysphagia of inclusion body myositis. Neurology 2002; 58(2): 326.
7. Takamiya M, Takahashi Y, Morimoto M, Morimoto N, Yamashita S, Abe K. Effect of intravenous immunoglobulin therapy on anti-NT5C1A antibody-positive inclusion body myositis after successful treatment of hepatitis C: A case report. eNeurologicalSci 2019; 16: 100204.
8. Ehler E, Melekova A, Latta J, Mandysova P, Vojtisek P, Zamecnik J. Inclusion body myositis with neck muscle weakness and positive effect of immunoglobulins - a case report. Cesk Slov Neurol N 2013; 76(3): 362-6.