Myositis involving Masticatory Muscles in Behcet's disease

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Abstract

Muscular involvement in Behcet's disease is rare manifestation in spite of basic characteristic that is vasculitis can invade multi-organ of the entire body. A few cases has been announced involving the lower extremities, the arm and generalized weakness. Like our case, myositis involving the masticatory muscles with clinically diagnosed Behcet's disease was presented with the magnetic resonance imaging (MRI) and the clinical findings, which is, not yet documented in the literature.

Key words

Behcet's disease, Myositis, Masticatory muscles

INTRODUCTION

The clinical triad of uveitis with oral and genital ulceration was probably first recognized by Hippocrates and in 1937 Hulusi Behcet described two cases of recurrent oral & genital ulcers with relapsing iritis[1]. This syndrome, thought initially to be a rare disorder found mainly in the Estern Mediterranean area, now appears to have world wide distribution. As the year have passed, the clinical spectrum has greatly enlarged and it is known that the disease is a more complicated entity affecting every tissue and organ in the body without exception. However, the disease has a predilection for certain organs and tissues such as mucocutaneous, ocular, articular, vascular, pulmonary, gastrointestinal and nervous system. Muscular involvement in Behcet's disease, rarely described in the literature, is usually mild and manifesting as myalgia or muscle weakness, and would be in most cases involving the lower extremities localized predominantly with pain and swelling but in a few cases presenting the arm and generalized weakness[2-7]. This case report is to present the MRI and clinical features of myositis involving the masticatory muscles which has not been documented.

CASE REPORT

A 33-year-old female whose clinical diagnosis was Behcet's disease was consulted from department of Dermatology, Chungbuk National University Hospital, for the evaluation and proper treatment of limitation of mouth opening and tenderness on left preauricular area since 3 weeks ago. She presented to Behcet's disease with 4 years history of intercurrent oral and genital ulcer, folliculitis, erosive gastritis, arthralgia, myalgia, swelling of finger and ankle joint, general weakness etc. She was able to open her mouth only 7 mm because of severe pain. On physical examination, painful swelling of left preauricular and temporal area was identified with tenderness and local heating. Oral examination revealed no evidence of infectious lesions in oral & maxillofacial region and patient had experienced no febrile history. There was no definite bony abnormality in plain x-ray view. Basic initial laboratory studies including serum enzymes, creatinine phosphokinase(CPK) and glutamic

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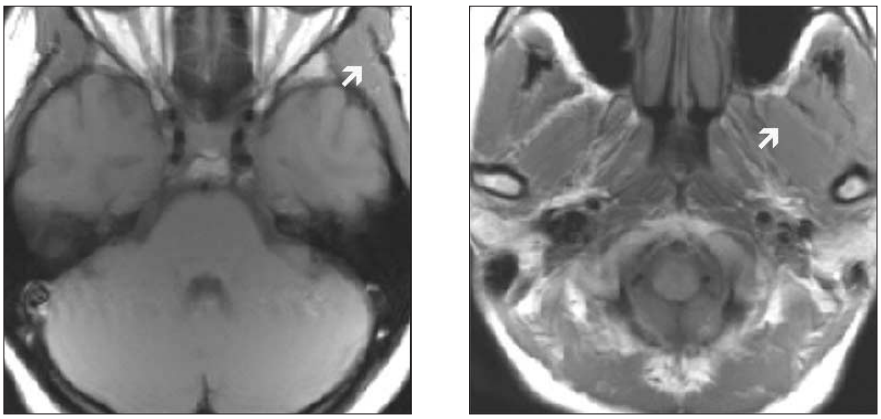
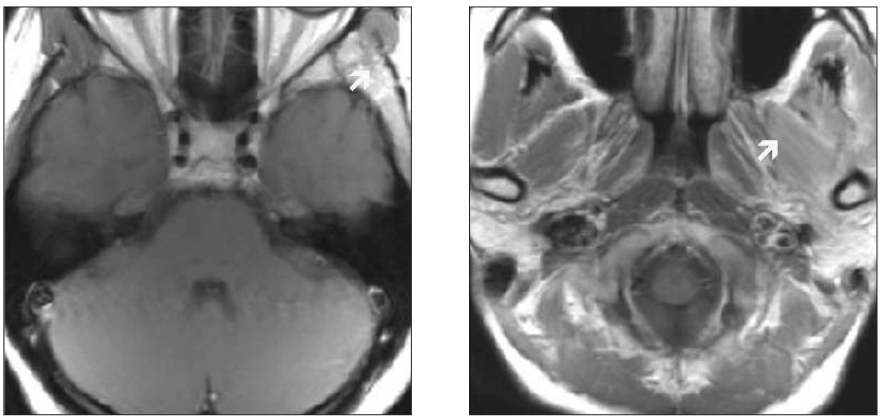
oxaloacetic transaminase(GOT) were within normal ranges except mild increased erythrocyte sedimentation rate(ESR). Symptom was not decreased to antibiotics and anti-inflammatory medication for initial diagnosis. Diagnostic infiltration anesthesia and arthrocenthesis of temporo-mandibular joint failed to improve restriction and pain on mouth opening. On MRI findings, it revealed mild swelling and abnormal signal intensity in left temporal, internal and external pterygoid and masseter muscles. Those muscles showed diffuse and patch hyperintensity on spin echo T2 weighted images and strong contrast enhancement on T1 weighted images(Fig.



a) precontrast Spin echo T1 weighted axial images



b) postcotrast Spin echo T1 weighted axial images

Fig. 1. Mild swelling, abnormal signal intensity and strong contrast enhancement in left temporal, internal & external pterygoid and masseter muscles.

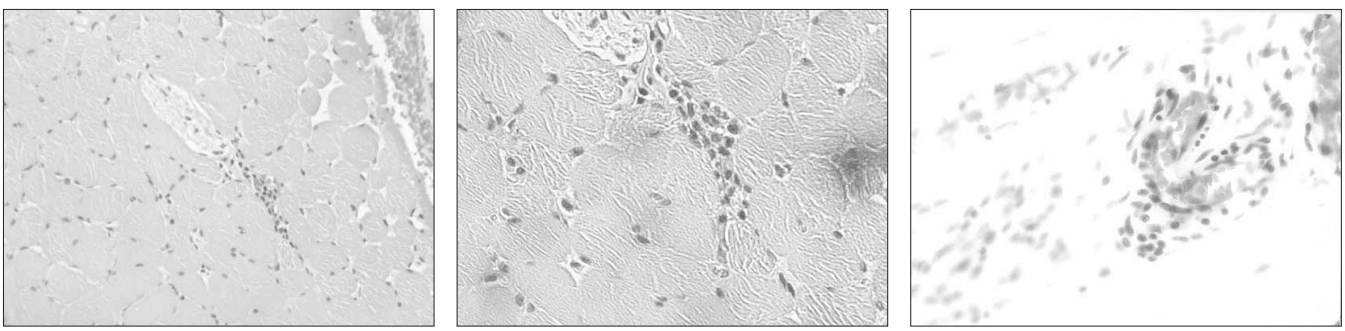


Fig. 2. Biopsy specimen from left temporal muscle shows perivascular inflammatory infiltration by mononuclear cells on light microscopy with H&E staining.

1). Also subtle signal abnormality in the right pterygoid muscle was present. No adjacent or remote disease which could explain the possible cause of the myositis was found on MRI of head & neck. Muscle biopsy was tried in the left temporal muscle which showed abnormal signal intensity on MRI. On light microscopic examination with H&E staining, endomysial perivascular inflammation was evident and mononuclear leucocytes were dominant with a few polymorphonuclear infiltrates and macrophages(Fig. 2). There were no bacteria or fungi. Three days of high dose intraoral steroid treatment resulted in the improvement of painful restriction of mouth opening.

DISCUSSION

The common histopathological lesion of Behcet's disease is vasculitis[8]. It is virtually unparalleled among the vasculitides in its ability to involve blood vessels of nearly all sizes and types, ranging from small arteries to large ones, and involving veins as well as arteries. Because of the diversity of blood vessels which affected, manifestations may occur at various sites throughout the body. For this reason, Behcet's disease is a multi-system disorder with frequent mucocutaneous and ocular lesions. Besides typical manifestations, clinical symptom also comprises non-specific inflammatory signs, like elevated serum immunoglobulins, elevated body temperature and increased ESR. Especially, Muscle involvement in Behcet's disease was rare and about 12 cases(18 patients) were reported in literature. They were mostly of lower extremity (13/18), two of generalized weakness (2/18), two with clinical evidence of peripheral neuropathy (2/18), four patients presented unilateral myalgia or muscle weakness (4/18), but none of other localization like our case. These papers were shown that myositis from Behcet's disease is usually localized, mild, short-lasting, may involve both extremities or generalized form and may recur. Laboratory findings such as creatine phosphokinase(CPK), aspartate aminotransferase(AST), alanine aminotransferase(ALT) and aldolase enzymes are usually normal, and those findings were normal in our case[8]. Because of such nonlinear view and lack of definitive laboratory screening or test, diagnosis is achieved clinically. Radiologic investigation such as ultra-sonography and MRI, and myopathology such as vasculitis appearances which are thickening of capillary basement membrane, inflammatory cell infiltration etc. and muscle fiber degeneration can be used by a diagnostic methods. MRI findings have been described as focal muscular lesions showing increased T2 and decreased T1 signal intensity with contrast enhancement in Behcet's disease which is known to involve muscles[9]. It is similar to our case that hyperintensity on spin echo T2 and strong contrast enhancement on T1, and allowed us to diagnose myositis due to Behcet's disease in our patient. Although MRI findings are usually clear-cut with typical clinical appearances, biopsy may be necessary in rare atypical cases. The myopathologies of Behcet's disease are known to include thickening of capillary basement membrane, excessive pleating of sarcolemma, subsarcolemmal aggregates of mitochondria and glycogen, disorganization and breakdown of myofibrils, central nucleation and the presence of a variety of cytoplasmic inclusions[8]. Previous reports of muscular involvement in lower legs described that the early phase inflammation is granulocytic-monocytic, and lympho-monocytic patterns represent the late stage manifestation[7]. Our case may be in between two phases since the temporal muscle specimen showed mononuclear cells as dominant infiltrates, but there were polymorphonuclear cells and macro- phages also observable. Although muscles showed abnormal features on MRI definitely, but pathological specimen had little evidence of diffuse muscular involvement except the presence of perivascular inflammation. The discrepancy between the MRI findings and the light microscopic features may be attributable to the fact that no special technique such as high resolution light microscopy, immunohistochemistry or electron microscopy was performed on the biopsy specimen, which could reveal the histological abnormalities in the affected muscle fibers that was undetected on conventional H&E staining. Even with the myopathy confirmed on electron microscope that is the most accurate histopathological examination of all, many revealed no evidence of abnormalities on the routine light microscopy. And it was reported that clinically silent specimens also showed muscular lesions on the sofisticated pathological evaluations[10]. The normal level of serum creatinine kinase in this patient may reflect the presence of rather circumscribed myositis than extensive myopathy with severe degeneration.

In the clinical differential diagnosis of myositis, infective cellulitis is the main consideration[11]. MRI is helpful in this situation by localizing the involvement to the muscle. Since our patient had known, longstanding

Behcet's disease and the MRI findings were typical, and a favorable response to treatment with anti-inflammatory and steroid medications further secured the diagnosis. The other differential diagnosis for MRI findings would include pyomyositis, viral myositis, compartment syndrome and traumatic muscle injury. Bacterial or viral myositis could produce similar MRI findings, but would be unlikely to present without abnormal laboratory tests. Compartment syndrome would result in a more diffuse abnormality involving the entire compartment, frequently with neural dysfunction caused by increased pressure. The lack of a history of trauma or heavy exercise prior to symptoms also helps exclude compartment syndrome as well as muscle contusion and strain, both of which could mimic the MRI findings.

SUMMARY

We report myositis involving the masticatory muscles occurred from Behcet's disease in a 33-year-old female, it is very rare, but should be considered as a possible manifestation.

REFERENCES

1. Serdaroglu P: Behet's disease and the nervous system. J Neurol 1998;245:197-205.
2. Arkin AK, Rothchild BM, Florendo NT, Popoff N: Behcet's syndrome with myositis: a case report with pathologic findings. Arthritis Rheum 1980;23:600-604.
3. Yazici H, Tuzuner N, Tuzun Y, Yurdakul S: Localized myositis in Behcet's disease. Arthritis Rheum 1981;24:636.
4. Di Giacomo V, Carmenini G, Meloni F, Valesini G: Myositis in Behcet's disease. Arthritis Rheum 1982;25:1025.
5. Finucane P, Doyle C, Ferriss J, Molloy M, Murnaghan D: Behcet's syndrome with myositis and glomerulonephritis. Br J Rheumatol 1985;24:372-375.
6. Lang BA, Laxer R, Thorner P, Greenberg M, Silverman E: Pediatric onset of Behcet's syndrome with myositis: case report and literature review illustrating unusual features. Arthritis Rheum 1990;33:418-425.
7. Worthmann F, Bruns J, Turker T, Gosztonyi G: Muscular involvement in Behcet's disease:case report and review of the literature. Neuromusc Disord 1996;6:247-253.
8. Sarui H, Maruyama T, Ito I, Yamakita N, Takeda N, Nose M et al: Necrotising myositis in Behcet's disease. Ann Rheum Dis 2002;61:751-752.
9. Gur Akansel, Yonca Akgoz, Ercument Ciftci, Arzu Arslan, Ali Demirci: MRI findings of myositis in Behcet's disease. Skeletal Radiol 2004;33:426-428.
10. Frayha R: Muscle involvement in Behcet's disease. Arthritis Rheum 1981;24:636-637.
11. Uziel Y, Lazarov A, Cordoba M, Wolach B: Paediatric Behcet´s disease manifested as recurrent myositis: from an incomplete to a full-blown form. Eur J Pediatr 2000;159:507-508.