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A Case of Multifocal Recurrent Nonpainful Myositis

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- Abstract -

Focal myositis is a benign inflammatory pseudotumor of a skeletal muscle that clinically mimics a tumor of soft tissue, but the cause of which is obscure. I report here a case of multifocal recurrent nonpainful myositis found in a 68-year-old man who showed a subacute multifocal recurrent nonpainful inflammatory myopathy affecting discrete muscle groups with spontaneous remission and/or some medication.

Key Word : Multifocal recurrent nonpainful myositis

A number of conditions may lead to localized, grossly evident enlargement of a single muscle or muscle group. These entities include hemmorrhage, amyloidosis, fasciitis, hypertrophy, abscess, cysticercosis, myositis ossificans, proliferative focal myositis, and focal myositis(FM). And polymyositis(PM), typically a diffuse disease from the beginning, may also occasionally start as a focal process[1-4].

I report here a case of multifocal recurrent nonpainful myositis found in a 68-year-old man who showed a multifocal recurrent nonpainful inflammatory myopathy.

CASE REPORT

A 68-year-old man who was in his usual state of good health until approximately three years ago. While playing golf, he noted tightness and swelling of his right lumbar paraspinal muscles. This progressed over several hours and then remained unchanged. He was seen at that time by a chiropractor and a masseur but there was no improvement. He was treated with an anti-inflammatory drug by his internist three weeks later. Approximately after one more month, the muscle swelling in the back slowly improved.

About one month after this, he noted the muscles of his right triceps was swollen and hard. It was not painful. He again was treated with an anti-inflammatory drug and this gradually improved after six weeks. Several months after this episode, he had a similar tightness and swelling of a muscle in his left posterior shoulder, again this episode was resolved sponta-

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neously over 1~2 months. About one year after this, a small knot appeared in his right anterior chest. Over several weeks swelling occurred over his entire right chest. This was not painful but several weeks later, he was not able to raise his arm above shoulder level. Then the symptoms disappeared spontaneously in about three weeks. About one year after this, firm nonpainful mass occurred on the left pectoralis muscle. The mass progressed for several days, then it improved in about one week by oral prednisone therapy.

Past medical history was unremarkable except for a vocal cord cancer which was treated with surgery and radiation nine years ago. And he had hernia repair 11 years ago. Family history was unremarkable.

Physical examination at the last recurrence showed an elongated firm nonpainful mass which measured 5~10cm in the middle portion of the left pectoralis muscle running from the axilla toward the sternum. Other physical and neurological examinations were unremarkable and he did not show any systemic manifestation.

Laboratory study showed that total CPK was 97IU/L, SGOT was 16IU/L, creatinine was 1.2mg/dl and alkaline phosphatase was 86U/L. Serum protein electrophoresis was normal except for a slightly increased beta globulin. CBC was unremarkable with a hematocrit of 44 and a white blood cell count of 5900. Platelet count was 217,000. Differential count was 59% segment, 17% lymphocyte, 11% monocyte and 3% eosinophil. Absolute eosinophil count was 100, which is slightly below normal. ESR was 4mm/hr. And rheumatoid factor, ANA, thyroid function, general chemistry, trichinella antibodies and chest X-ray were unremarkable. MRI scan of the back showed a possible inflammation of the back muscle.

EMG of both pectoralis muscles showed a fibrillation, positive sharp wave and small amplitude short duration polyphasic motor unit potential with reduced interference pattern and early recruitment as seen in active myopathy. Conventional nerve conduction studies and EMG of the right triceps and lumbar paraspinal muscles were normal. Muscle biopsy of the left pectoralis muscle showed an endomysial and perivascular inflammatory cells, phagocytosis, regeneration and floccular change of muscle fibers, and increased endomysial connective tissue as seen in inflammatory myopathy(Fig. 1).

DISCUSSION

FM is a benign, self-limited inflammation of the skeletal muscle, which presents as a soft tissue pseudotumor frequently mimicking a sercoma or abscess.

It was first described as a distinct clinicopathologic entity by Heffner et al in 1977[5], thereafter a number of cases have been reported occurring in the upper and lower limb, trunk, neck, tongue, rectus abdominal muscles, upper eyelids and tem-

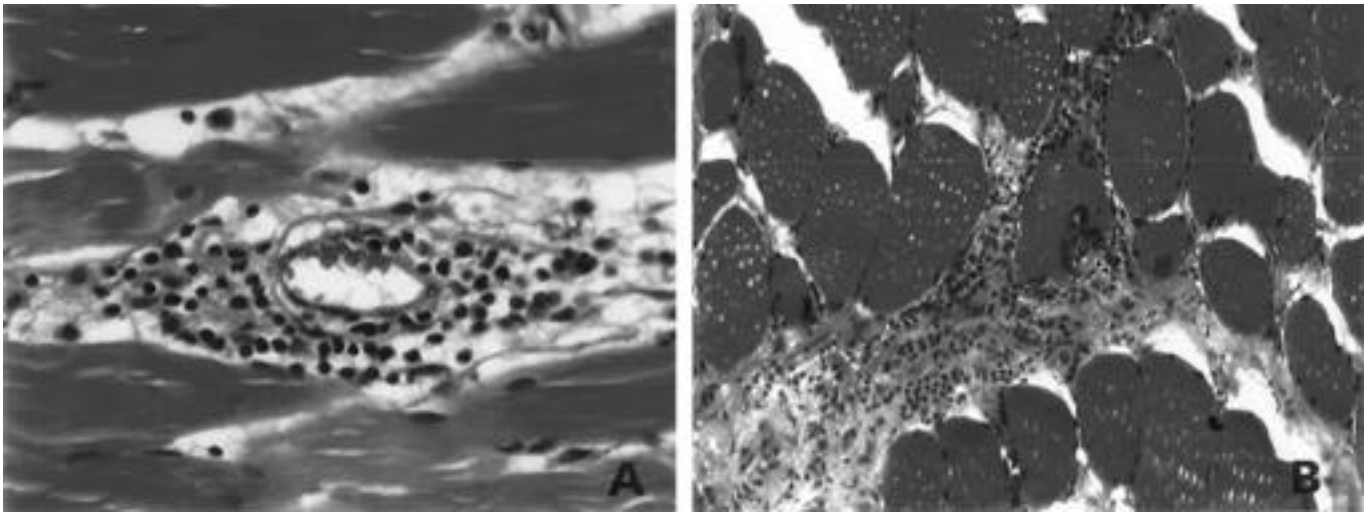

Figure 1. A. *Muscle biopsy shows perivascular inflammatory cells(H & E, ×400).* ***B.*** *Phagocytosis of muscle fibers(H & E, ×400).*

poral muscle[3,5-21].

But historically a patient with a similar conditon, "relapsing myositis", was described by Mcletchie and Aikens in 1952[22]. Inflammatory myopathy with a focal presentation was also described by Cumming et al in 1977 as localized nodular myositis[23]. And a patient with multifocal interstitial myositis associated with localized lipoatrophy was reported by Palliyath and Garcia in 1982[24].

Myositis has a histologic definition, according to the features described, but clinical presentations may be variable and several classifications have been proposed with nosologic problems. Therefore, several names have been given to this disease as localized nodular myositis, localized interstitial PM, interstitial nodular myositis and focal nodular myositis[3].

The etiology of FM is unknown. But FM has been reported after trauma, excessive physical activity and following infection with Borrelia burgdorferi[3,5,13,19]. No direct relationship to trauma has been found, but subclinical episodes of mild trauma can not ruled out as a contributing factor[8,12]. Other infections which can affect skeletal muscles including bacterial, viral, and parasitic agents like trichinosis, echinococcosis, cysticercosis, or toxoplasmosis have to be included[3.5,13,19]. But viral particles have not been identified in specimens of FM despite careful searching with electron microscopy[5]. Some authors suggest that a denervating process plays an important role in the pathogenesis of FM[1,16]. However, it remains unclear whether a focal inflammatory lesion is the response to local nerve damage or whether histologically proven denervation is caused by inflammatory infiltrates[12,19]. A genetic predisposition is possible. An association with HLA typing was suggested(HLA B8-DR3-DW6), but no studies exist on FM[3]. My case had a spontaneous onset.

This condition characteristically develops during several weeks into a circumscribed painful enlargement within the soft tissue, usually of an extremity, leading to a preoperative diagnosis of malignacy[5]. No age and sex predominance have been reported. In every case the myositis appeared as an enlarging mass within the muscle. Usually, the mass was painful, elicited by palpation, and unattached to the subcutaneous tissues. A rapid enlargement developed inside a few weeks. In a few cases, general symptoms were present, including fever, generalized muscle weakness, and weight loss. No family history of muscle disease or connective tissue disorders have been reported[3,23]. My case was an idiopathic multifocal recurrent nonpainful nonsuppurative myositis.

Flaisler et al[3] reviewed 39 published cases and their own case, and reported that ESR was normal in 24 cases, elevated in 8 and undetermined in 8. And serum enzymes including CPK were normal in 24 cases, elevated in 8 and undetermined in 8. EMG often showed myopathic potentials suggestive of PM and showed brief duration, small amplitude motor unit potentials and fibrillations in the affected limbs[25-28].

CT findings include irregularity and enlargement of the muscle involved, with diffuse, poorly defined fatty infiltration of the muscle planes, but no evidence of an associated mass. And CT was helpful in determining the nature and extent of the abnormality for needle biopsy and follow up[14]. MRI showed no abnormality of signal on T1 and T2 weighted images. MRI allowed only a diagnosis of a muscular pseudotumor[3]. Despite an accurate iconography, patients were often given a presumptive diagnosis of infiltrative soft tissue neoplasm, most likely a rhabdomyosarcoma[3].

The diagnosis can be confirmed only by biopsy and in all cases histologic features of inflammatory myopathy were observed as inflammatory infiltration(plasma cell, lymphocyte) collected in sheets, in clusters within interstitium, sometimes around blood vessels, necrosis and regeneration of muscle fibers with intense phagocytic activity, variable fiber size and interstitial fibrosis. Inflammatory changes consisting of perimysial collections of mononuclear cells and single muscle fiber necrosis were observed in biopsies from the involved limbs and were absent in muscle from the noninvolved extremity. These data were found in variable percentages, with interstitial edema in acute lesions and focal fibrosis in older lesions predominant[3,4,18,28]. My

case showed evidence of inflammatory myopathy.

FM is defined by two precise features as the myositic process affects a single skeletal muscle without systemic manifestations, and it presents as a benign inflammatory pseudotumor. The histologic study shows the appearance of a severe myopathy with inflammatory infiltration and alteration of muscle fibers as described in the typical form of PM[3]. It is important to determine if FM is an individualized entity or a rare form of PM. In FM, the pseudotumor is unique, usually painless or dull sensation of discomfort, laboratory abnormalities are generally absent, no recurrence is observed after surgical excision, some regression after incisional biopsies, no development of a systemic disease after years of follow up, and biopsy shows muscle fiber hypertrophy but no infarct-like necrosis of the affected muscles, and tumor may disappear spontaneously. On the other hand, in the localized form of PM, patients very often present more than one muscle, these masses are painful and early recurrence are observed after biopsies. Furthermore, ESR and/or CPK are elevated early in the course of the disease, and patients quickly develop a characteristic pattern of PM with systemic manifeststions, including fever, malaise, dysphagia and arthralgia. Biopsy also does not show muscle fiber hypertrophy but infarct-like necrosis is present. But histology can not definitely separate the two entities[3,10,18,25,29].

FM should be considered in the differential diagnosis of soft tissue masses involving skeletal muscle. The most common misdiagnosis is a soft tissue sarcoma. Unlike proliferative myositis, FM does not exhibit prominent fibroblastic proliferation in the connective tissue matrix, nor do the lesions contain large basophilic giant cells. Fibrous tissue proliferation in FM appears indolent with little fibroblastic activity. Complete replacement of muscle fibers over a large circumscribed area, as in nodular fasciitis or myositis ossificans, is never seen. The zonal phenomenon with varying maturity of ossification typical of myositis ossificans is not encountered. Unlike nodular fasciitis, FM does not primarily involve the fascia or subcutaneous tissue, nor are there any hypercellular pseudosarcomatous areas composed of proliferating capillaries, immature fibroblasts, multinucleated giant cell and mitotic figures. Eosinophilic myositis, which may be focal in nature, was excluded by the absence of a frank eosinophilic infiltrate in the biopsy specimen. Giant cell or granulomatous myositis was excluded by the absence of granulomata in the biopsy specimens and the lack of evidence to support a diagnosis of sarcoidosis. In the head and neck region, salivary gland lesions and hypertrophic branchial myopathy are additional clinical considerations[4-6,9-11,20,30].

Flaisler et al[3] reported that 21 cases(among 39 cases) with normal ESR and CPK had a favorable prognosis with spontaneous disappearance of the lesion. Consequently, normal muscle enzymes and biologic inflammatory tests suggest a benign evolution whereas elevated ESR or CPK increase the possibility of developing PM. Liefeld et al[10] reported that once the diagnosis is established, no further treatment is required, and patients can be followed up in the anticipation of this benign muscle enlarging disorder resolving. Issacson et al[13] and Naumann et al[19] reported that drug therapy with nonsteroidal antiphlogistics, or in more severe cases, with glucocorticosteroids, has proven benifial. But surgical excision of the affected muscle is not recommended since surgery did not alter the clinical course of those patients in whom it was taken. And other authors reported that corticosteroid was effective in FM and a poor course took place usually within one year of follow up[3,15]. Initially my case did not show good response to anti-inflammatory drug therapy, but later showed spontaneous remission and good response to prednisone therapy. He showed frequent recurrence but does not show any systemic manifestations.

FM, a rare pathology, presents with an alarming initial, veritable pseudotumoral mask. Surgical biopsy is indispensible to confirm the diagnosis. When it is true focal form(without evolusive potential), there is no therapeutic indication, and simple follow up is sufficient. If the picture is that of an initial focal form of PM, it develops

within a few weeks to a year, with the rapid appearance of clinical signs, laboratory findings and EMG indications[15].

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