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Case Report

Pseudotumor of muscle - focal myositis

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ABSTRACT

Focal myositis is a rare disease, and to date, 250 cases have been reported in literature. It is a benign dysimmune disease of unknown etiology. It is defined as myopathy affecting a single muscle without systemic manifestation with a historically proven inflammatory myositis process. It usually presents as a mass ranging from 1 to 20 centimeters, growing insidiously over weeks to months, and may be painless or tender. Or the growth can be rapid and with a lot of pain and disability. It's common in lower limbs and rare in facial muscles. It usually regresses spontaneously and does not invade the surrounding structures. In 18% of cases, there may be a relapse. Laboratory studies may show elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Creatine kinase (CK) is usually not elevated significantly. Autoantigens are often normal or may be marginally elevated. Magnetic resonance imaging (MRI) is a diagnostic tool of choice and is done with relative ease. Electromyography (EMG) occasionally shows recordable spontaneous and repetitive discharges. Nerve conduction velocity (NCV) may be helpful if nerves are involved. A muscle biopsy usually confirms the diagnosis with specific features. Tumours of muscle and Inflammatory myositis need to be differentiated. Sometimes focal myositis may be associated with immune-mediated inflammatory disease (IMID), neoplasms, radiculopathy, and trauma. Focal myositis is usually self-limiting and benign. It usually responds to NSAID and occasionally may require a short course of steroids, especially in those with elevated ESR and CRP.

Keywords: Myositis, Focal, Pseudotumors of muscle, Creatine kinase, Myositis panel, Inflammatory myositis, Rhabdomyosarcoma

INTRODUCTION

First described by Heffner et al in 1973, focal myositis (FM) is a rare disease associated with isolated inflammatory pseudotumors restricted to skeletal muscle, with focalized abnormalities in magnetic resonance imaging (MRI) or electromyography (EMG) and myositis on pathological examination.[1] Among the heterogeneous group of inflammatory myopathies, FM is a benign dysimmune disease with unknown etiology.[2] Although it can be associated with root and nerve lesions, muscle trauma, malignant disorders and autoimmune diseases, its triggering factors are not certain. Flaisler et al defined FM as a "myopathy affecting a single muscle without systemic manifestation with a historically proven inflammatory myositis process."

This disease can occur at any age but adults are often affected. Both males and females are equally affected. Maybe multiple muscles from different regions are involved. The prevalence is less than 1/1,000,000. To date, there have been 250 cases reported in the literature. It usually presents as a mass ranging from 1 to 20 centimeters, grows insidiously over weeks to months, and may be painless or tender. Sometimes growth can be rapid and with excessive pain and disability.[3] Usually lower limb muscles are involved, the most common being abductor, vastus lateralis and gastrocnemius. Rarely occurs in

muscles of the head and neck. Systemic symptoms such as fever, myalgia, general weakness, and weight loss can occur. It usually regresses with time and doesn't invade surrounding structures, tendons, fascia, and skin. Recurrence is possible in 18% of cases. Classical inflammatory myopathies may present focally occasionally, resembling focal myositis, creating diagnostic difficulties.[4]

Laboratory studies including erythrocyte sedimentation rate (ESR), creatinine kinase (CK), and c-reactive proteins (CRP) are often normal. Often, autoantigens are not significantly elevated. Elevated CK and ESR were observed in 25% of cases as reported by Yamnaz. Nerve conduction (NCV) and EMG studies may help to determine the involvement of nerves and focal or generalized abnormalities of the disease. Nerve conduction studies were normal when the nerve was not involved. Spontaneous activity with complex repetitive discharges on EMG is occasionally recorded in the muscles involved.[5]

MRI is a diagnostic tool that is both convenient and effective. Usually, an MRI shows a muscle mass or enlargement. It appears hyper-intensity on T2 STIR and iso or hyper-intensity with a homogeneous or heterogeneous signal on T2 weighted images. Ultrasound examination performed by an expert could provide an idea of myositis.[6]

A muscle biopsy confirms the diagnosis. Histological features include marked variations in fiber size with atrophic or hypertrophic changes. It is possible to see fibers that are degenerative or regenerative but with either significant or minimal fibrosis.

Benign tumors involving muscles, such as rhabdomyoma, fibromatosis, and intramuscular lipoma, may mimic myocytic pseudotumor. Malignant tumors such as rhabdomyosarcoma, liposarcoma, leiomyosarcoma, and metastasis may be confused with myositis in the early stages when they have not yet infiltrated neighboring structures. Pathologically, it can resemble muscular dystrophy or inflammatory myopathy at times.

The natural course of the disease is benign with occasional recurrence. Non-steroidal anti-inflammatory drugs may suffice. For non-responders, a short course of oral steroids may be beneficial, especially in individuals whose CK and ESR are elevated.

CASE REPORT

A 23-year-old male patient, a software engineer by profession, working at Hyderabad, was admitted to our hospital on 26 December 2019. He was not a smoker, alcoholic, diabetic, or hypertensive.

He developed pain in his right heel radiating to his calf posteriorly on 9 December 2019. At that time, he was not feverish. An orthopedic surgeon was consulted and an X-ray of his ankle and foot was performed, which did not reveal any pathology. He was given non-steroidal anti-inflammatory drugs. His urticaria rash was treated with pheniramine maleate and dexamethasone intramuscularly for two days, and it eventually disappeared. He experienced a partial decrease in the severity of his heel and calf pain. Within two days, the pain in the right heel and calf intensified, and he also experienced pain in the left shoulder and wrist, with minimal swelling. At that stage, he consulted us and was admitted to our hospital. He was investigated.

The total white cell count was 17,000, with polymorphs accounting for 80%, lymphocytes 10%, eosinophils 5%, and monocytes 5%. ESR was 22 mm 1st hour, CPK was normal 0.6 mg/dl (0.5 to 1.4), CRP was elevated 155 mg (0-6 mg/l) and serum glutamic pyruvic transaminase (SGOT) increased 316 IU/l (5-42).

An ultrasound scan of the right calf and tendo-achilles showed subtle changes in the echogenicity of the calf muscles, suggesting myositis (Figure 1).

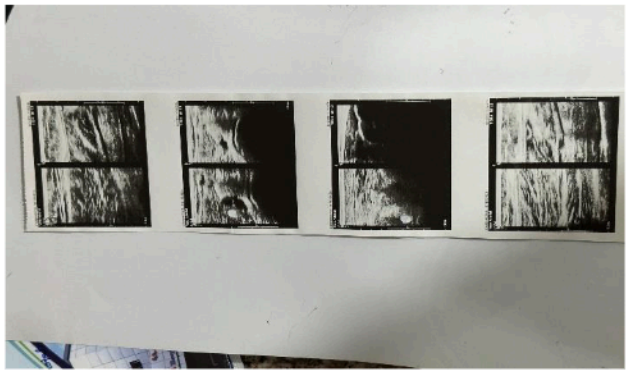

Figure 1: Ultrasound scan of right calf and tendo-achilles showing subtle changes in the echogenicity suggestive of myositis.

MRI of the right leg revealed T2/STIR hyperintensity of soleus, flexor hallucis longus and flexor digitorum longus muscles suggesting myositis (Figure 2). The anterior aspect of the left forearm showed subcutaneous edema on MRI. T2/STIR hyperintensity has been noted in the flexor pollicis longus, flexor carpi radialis longus, extensor carpi radialis longus, and brachioradialis muscles of the left forearm, suggesting myositis (Figure 3).

There were no significant abnormalities found in NCV and EMG studies, and the antinuclear antibody profile was normal.

In view of clinical presentation and laboratory evidence of myositis, a clinical diagnosis of infectious polymyositis was made. He was given intravenous piperacillin tazobactam 4.5 grams three times, and clindamycin 600 milligrams three times, in addition to dexamethasone 4 milligrams twice a day. He responded well to the regimen,

his muscle swelling and pain subsided completely. He was discharged on 60 mg of prednisolone per day, with a gradual reduction of the dose by 20 mg every 5 days.

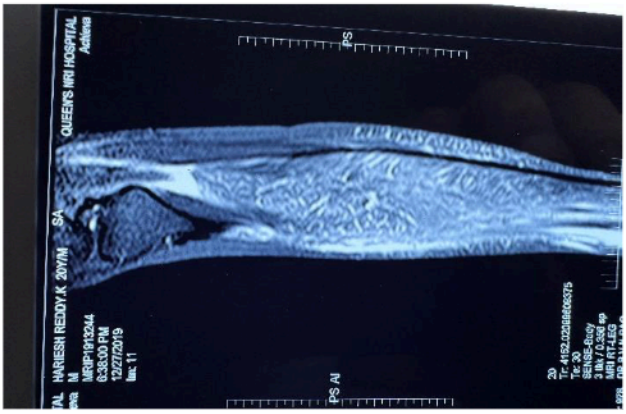

Figure 2: MRI of right leg indicating myositis.

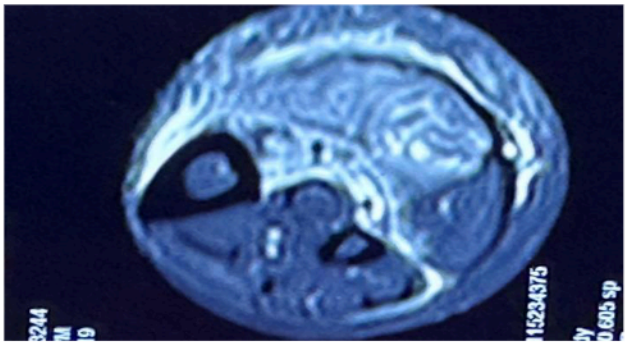

Figure 3: MRI of left forearm showing hyper intensity suggestive of myositis.

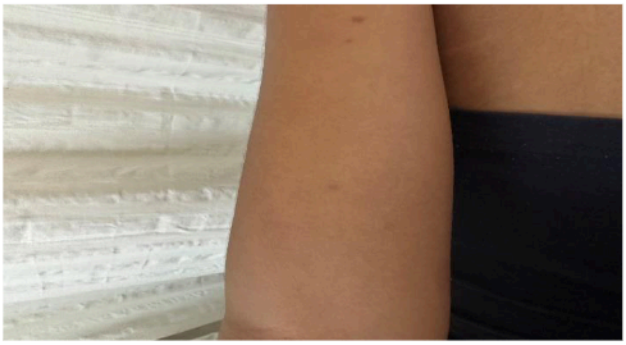

Figure 4: Swelling left proximal forearm.

He was referred to a rheumatologist who reviewed the case. The myositis profile test revealed positive results for Mi-2 beta and PL-7. He considered evolving Inflammatory Myositis as a provisional diagnosis. In addition to steroids, he added mycophenolate mofetil (MMF) 500 mg two tablets every third day. He continued treatment under the telephonic guidance of the Rheumatologist for 6 months but lost follow-up due to the corona pandemic.

He was readmitted on 26 July 2023 with pain full swellings of right heel and right & left calf, right middle finger, left proximal forearm (Figure 4), left distal forearm (Figure 5) and left brachioradialis. The investigation revealed that there were 9,600 white cells, 38 IU/l SGPT, 3.6 mg/dl CRP, 8 mm/1 hour of ESR, and 64 U/l CK. Repeated myositis profile showed borderline positivity for EJ antigen.

In view of borderline and transient elevated myositis markers, there was weak evidence in favor of inflammatory polymyositis. CPK levels were never elevated, which pointed against inflammatory myositis. Elevated CRP, episodic nature of the disease, and myositis in focal groups of muscles explain the diagnosis of a rare disease "focal myositis".

The patient was explained the nature of his illness and discharged on a short course of steroids and colchicine.

Reviewed every month for one year, there was no recurrence of symptoms and he was not on any medications.

Figure 5: Swelling left distal forearm.

DISCUSSION

A 23-year male, presented to us with recurrent muscle pains and swelling. Initial presentation he had a high leukocyte count, high ESR, high CRP and high SGPT. Ultrasound and MRI scans were suggestive of myositis. His EMG and NCV were normal. CK was not elevated, and the autoantibody screen was negative. Muscle swellings were localized and did not invade fascia and surrounding structures. Hence clinical diagnosis of infectious myositis was made and was treated with antibiotic and steroidal anti-inflammatory drugs. Infections also could trigger myositis.[7] He responded well to treatment. At times it may be difficult to differentiate inflammatory myopathies and benign focal myositis. Absent myopathic antigen markers, and normal CRP, the focal nature of the disease favors focal myositis. However, it may be the tip of the iceberg we may be seeing, and it actually may be an early stage of inflammatory myositis. The fact warranted a chronological observation of the patient. A myositis profile done by another physician at a later date gave a transient elevation of MI 2 and PL 7 raising the doubt of an inflammatory myositis and landing

the patient in exposure to immune suppression unnecessarily.

Years later he presented with similar complaints of painful pseudo tumors of muscle. 18% of people with focal myositis have recurrence as reported earlier. This time in myositis profile, EJ only transiently elevated pointing against inflammatory myositis. His leukocyte count, ESR, CPK, CRP and SGOT were normal. Focal pseudotumors of muscles, normal CK, and transiently elevated myositis markers made focal myositis more likely than inflammatory myositis.

Sometimes tumors of muscle may need to be differentiated from myositis swellings. MRI examination and Pathological testing usually clearly delineate the two. As the treatment modalities are entirely different, a clinician should know both.

Inflammatory myopathy may present as a mass in the muscle closely resembling benign myositis. Hence those patients diagnosed with focal myositis need prolonged clinical follow-up.

There were rare reports of focal myositis presenting as sartorius muscle contraction requiring surgery.[8] Gallay et al in their article on focal myositis detailed its association with various diseases. 32% had immune-mediated inflammatory diseases (IMID), 24% had neoplasia, 11% had radiculopathy, and 2% had trauma. Many other rheumatological disorders are associated with focal myositis. Behcet's disease is one of the common IMIDs associated with focal myositis.[9,10] This study details the importance of searching for any underlying significant pathology before tagging focal myositis benign.

Focal myositis is usually self-limiting and benign. It may respond to non-steroidal anti-inflammatory drugs or may require a short course of steroids, especially those having higher CRP and ESR.

CONCLUSION

Focal myositis is a rare disorder that may be misdiagnosed, leading to unnecessary investigations or delayed treatment. Clinicians should consider focal myositis in the differential diagnosis of muscle tumors and inflammatory lesions, ensuring appropriate management and avoiding unnecessary interventions.

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