

*Case Report*

## **Eosinophilic Fasciitis: Role of Contrast Enhanced MRI as A Substitute to Biopsy**

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### **ABSTRACT**

Eosinophilic fasciitis is rare conditions that presents with skin thickening involving the extremities and usually spring the hands, with peripheral eosinophilia. Raynaud's phenomenon is usually absent and hands are typically spared. We describe a case of 20 year old male patient who presented in immunology OPD with generalized myalgia and skin tightening over the trunk, arms and forearms. Examination showed peripheral eosinophilia, biopsy confirmed eosinophilic infiltrates in skin and subcutaneous tissues and MRI showed characteristic abnormal fascial signal intensity and enhancement.

**Key words:** Eosinophilic infiltrate, Fascial thickening, MRI.

### **INTRODUCTION**

Eosinophilic fasciitis (EF), also called Shulman syndrome, is a rare, localized fibrosing disorder of the fascia, characterized by peripheral eosinophilia and fasciitis. Fascial thickening in the setting of eosinophilia, elevated erythrocyte sedimentation rate, and hypergammaglobulinemia are the critical elements of the syndrome. The diagnosis of eosinophilic fasciitis is suspected in a patient presenting with characteristic skin changes and consistent laboratory findings. It is confirmed with full-thickness biopsy or characteristic MRI findings. Here we present a biopsy proven case of eosinophilic fasciitis with characteristic MRI finding.

### **CASE SUMMARY**

A 20 year male patient presented in the immunology OPD with complaints of generalized myalgia and skin tightening over the trunk, arms and forearms (left>right) for 3 months. He had lost around 3 kg

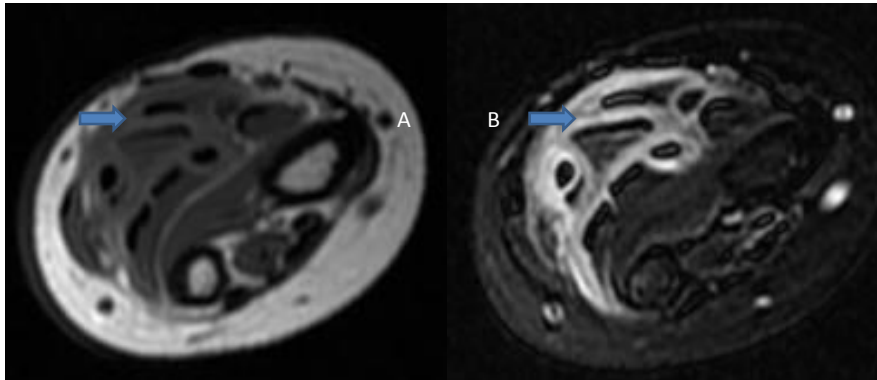
weight over the last 3-4 months. No other comorbid conditions were present. No complaints regarding cough, breathlessness, chest or abdominal pain were present. Laboratory examination revealed mild anemia, raised total leukocyte count (14500 cells/mm<sup>3</sup>) with 12% eosinophils. ESR was 30mm. Renal and Liver function tests were normal.

Patient was sent for MRI examination of the left forearm with contrast. MRI was done on a 3-T scanner (Signa, General Electric Medical Systems, Milwaukee) using an extremity coil in a neutral position. The parameters for these MRI examinations were as follows: axial T1-weighted conventional spin-echo images (TR/TE, 1360/7.2); axial fat-suppressed T2-weighted fast spin-echo images (TR/TE, 5400/111); axial and coronal post contrast fat suppressed T1 (TR/TE; 1000/7.3)

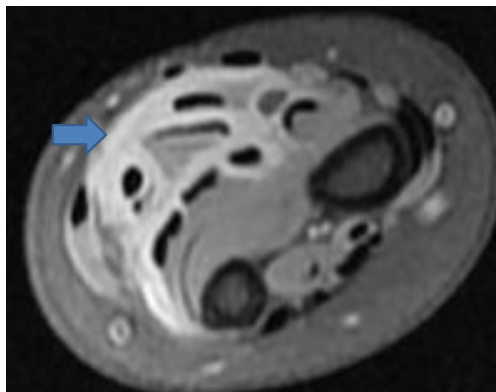
T1WI showed thickened superficial and deep fascia along the forearm with signal intensity more than the muscle. Fat-

suppressed T2WI showed increased signal intensity in the fascial layers (Figure 1). Post contrast T1W images (Figure 2) showed intense enhancement in the areas that showed increased T2 signal intensity. No evidence of muscle edema or adjacent bony involvement was noted. Rests of the extensor and flexor compartment tendons were normal.

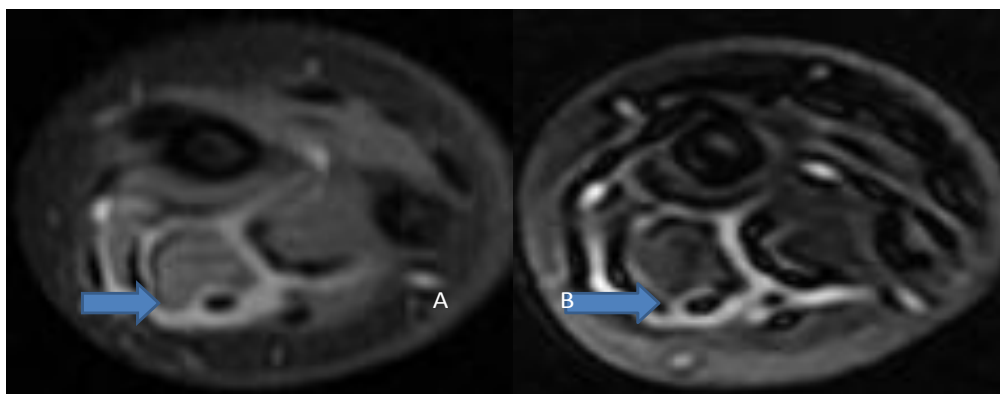
Patient was kept on oral steroids and was followed up after 6 weeks. Repeat MRI was done (Figure 3) which revealed reduction in the T2 signal intensity and post contrast enhancement along the fascial layers which corresponded to mild clinical improvement.



**FIGURE 1:(A) T1WI showed thickened superficial and deep fascia along the forearm (arrow) with signal intensity slightly more than the muscle. (B) Fat-suppressed T2WI showed marked increased signal intensity in the fascial layers**



**FIGURE 2:Axial post contrast T1WI showing intense enhancement (arrow) in the areas that showed increased T2 signal intensity**



**FIGURE 3: Follow up after 6 weeks: A) Axial T1WI post contrast image and (B) Fat suppressed T2WI showing mild reduction in extent,thickness and intensity of the fascial layers after 6 weeks of steroid treatment.**

## DISCUSSION

Eosinophilic Fasciitis (EF) also known as Shulman's syndrome or diffuse fasciitis with eosinophilia is an uncommon disease clinically characterized by painful thickening and induration of the skin and subcutaneous tissues of the affected limbs. [1] Disease onset may follow an episode of strenuous physical activity.

This disorder is characterized by peripheral eosinophilia and fasciitis that could be differentiated from scleroderma by the distinctive pattern of skin involvement that spares the digits, involves fascia rather than dermis, and is not accompanied by Raynaud phenomenon. The disease may affect the upper and lower extremities and the trunk but spares the head. Very rarely, the hands and feet may be involved

The age range in eosinophilic fasciitis is 1-88 years, although most patients present during the third to sixth decade. The clinical presentation of EF is variable and nonspecific. Patients may present with muscle weakness, swelling, and stiffness of the extremities and skin changes. Joint contractures may occur as a sequela of induration and sclerosis of the subcutaneous tissue. [2] The skin is thickened with induration and hyperpigmentation. Visceral organs are not involved in EF, a finding that helps differentiate eosinophilic fasciitis from systemic sclerosis and other differential.

Characteristic laboratory findings of eosinophilic fasciitis (EF) include peripheral blood eosinophilia, hypergammaglobulinemia and an increase in the erythrocyte sedimentation rate (ESR). Peripheral blood eosinophilia is described as a hallmark of the disease, although its severity varies over time, even in the absence of specific treatment. [3]

The pathophysiology underlying eosinophilic fasciitis is postulated to involve an inflammatory response resulting in an activated inflammatory cell infiltrate of affected tissues and subsequent

dysregulation of extracellular matrix production by lesional fibroblasts.

MRI findings in eosinophilic fasciitis are highly characteristic and include fascial thickening that appear brightly hyperintense on fat sat T2 sequence. The thickened fascias also appear mildly hyperintense to muscle on T1W sequence and demonstrates intense enhancement after administration of intravenous contrast. So MRI may be a helpful guide for the area to be biopsied in order to reduce false negative results. [4] MRI signal characteristics may correlate with the disease activity as highlighted in previous case reports. [5] We support this observation as we have also noted that the MRI signal characteristics reflect clinical disease activity, especially the degree of T2 signal hyperintensity within the fascia and corresponding areas of fascial enhancement on post contrast images. The T2 hyperintensity and post contrast enhancement usually disappear after treatment. MRI may be useful as a non radiation modality in reevaluating these patients after treatment for remission, recurrence or exacerbation of the disease. However, still the gold standard for the diagnosis of eosinophilic fasciitis is a full-thickness skin biopsy.

Eosinophilic fasciitis is generally corticosteroid-responsive, and initial treatment regimens are based on this therapy. Multiple additional agents have been used in steroid-refractory disease.

## CONCLUSION

MRI findings in eosinophilic fasciitis are characteristic and consist of abnormal fascial signal intensity and enhancement. Though histological examination remains the golden standard, MRI may be helpful to confirm fascial inflammation and may serve as an alternative to biopsy, if biopsy is not meaningful or not possible.

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