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Groove Sign as an Eosinophilic Fasciitis Clue

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ABSTRACT

Eosinophilic fasciitis is a rare variant of scleroderma with an unknown etiology, predominantly affecting middle age group. Clinically it presents with local oedema, erythema, woody induration with an "orange peel" appearance, progressing rapidly to fibrosis associated with hematological changes mainly eosinophilia. A deep fascial biopsy and imaging studies aid the diagnosis. Classical skin findings help in early diagnosis thereby preserving the mobility and preventing the joint contractures. This report presents a case of 59-year-old male with features suggestive of eosinophilic fasciitis and its importance in diagnosis.

Keywords: Groove sign, peau d'orange appearance, dermascopy, Eosinophilic fasciitis, Shulman syndrome.

INTRODUCTION

Eosinophilic fasciitis (EF) or Shulman syndrome first described by Shulman in 1975, an autoimmune disease of rare entity characterized by progressive induration, symmetrical thickening and edema. These changes are later replaced by fibrosis of fascia resulting in cobblestone appearance or *peau d'orange* appearance and the groove sign, complicated by joint contractures predominantly involving the extremities. (1)
Biochemical abnormalities include eosinophilia, elevated erythrocyte sedimentation rate, high C-reactive protein & hypogammaglobinaemia (2). Histological diagnosis requires a deep biopsy including deep fascia for a definitive diagnosis of the disease. Here we report a classical case of eosinophilic fasciitis in a 59 year old male with grooves sign and peau d'orange appearance and its dermascopic features.

CASE REPORT

A 59-year-old man who is a driver by occupation presented with tightening of skin with stiffening and edema of bilateral lower limbs. His symptoms started 2 months back with swelling and pain followed by restriction of motility involving both lower limbs. He also gave history of gradual progression of stiffness of lower limbs from below upwards. He had associated itching and tingling sensation. History of significant weight loss present. He denied history of trauma and strenuous work. No history suggestive of Raynaud's phenomena. No history of chest pain or dyspnea. The physical examination revealed edema and woody induration showing peau d' orange appearance with loss of hair over both lower legs. Indentations of skin along the course of superficial veins (Groove sign) present

over both lower legs and forearms (Fig.1a&b).

Laboratory investigations revealed elevated eosinophils, elevated erythrocyte sedimentation rate and negative for antinuclear antibodies and anti-SCL70. Blood counts, chest x-ray was normal.

Skin biopsy taken from right lower leg revealed thickened and hyalinized collagen bundle in superficial and deep dermis with mild perivascular and interstitial lymphocytic infiltrate and atrophy of fat lobules in subcutis (Fig.2a) confirming the diagnosis of eosinophilic fasciitis. Dermoscopy showed peau d'orange appearance and groove sign (Fig.2b). Based on history, clinical and histopathological examination a diagnosis of eosinophilic fasciitis was achieved.

Patient was started with oral prednisolone 1mg/kg/day at tapered doses combined with oral methotrexate at a dose of 10mg/week and topical tacrolimus 0.1% ointment after which the patient had significant improvement in skin thickening.

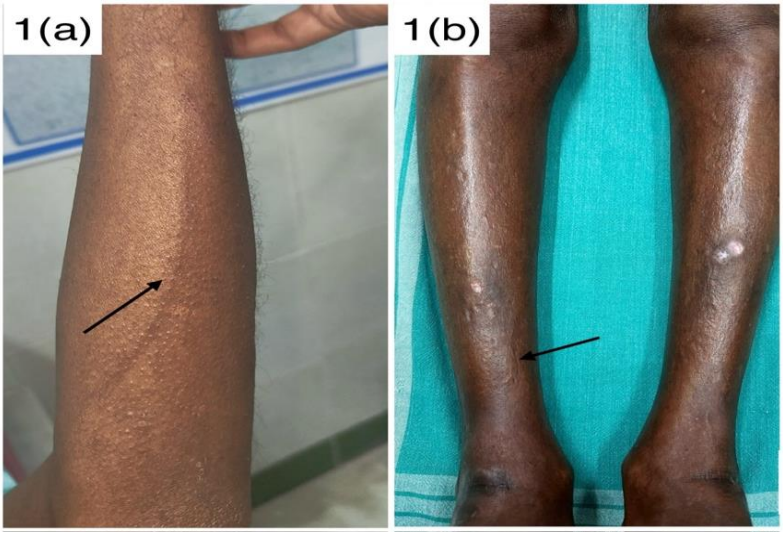

Fig.1a- Indentation of skin along the course of superficial veins along forearm (black arrows).
Fig.1b- Indentation of skin along the course of superficial veins along both the legs (black arrows).

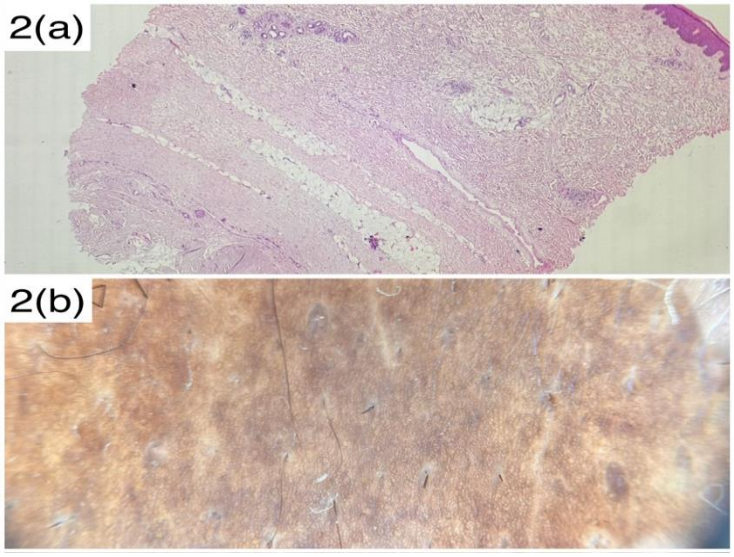

Fig.2a- Histopathology section showing hyalinised collagen bundle in superficial and deep dermis.
Fig.2b- Dermoscopy showing peau d' orange appearance and groove sign.

DISCUSSION

EF is a rare connective tissue disorder involving bilateral extremities with scleroderma like skin changes sparing the fingers and toes, affecting both the genders equally, mostly with an unknown etiology. [1] Few studies over 30-40% suggests an infectious etiology such as borrelia burgdorferi and mycoplasma arginine; others being severe trauma, exercise, splenectomy and drugs.[1]

Early cutaneous manifestation includes burning sensation, pain & edema; later progressing to fibrosis and induration showing peau d'orange appearance. A classical clinical finding of eosinophilic fasciitis is the 'groove sign' which is a linear depression along the course of veins, accentuated by limb elevation, due to inward tethering of the skin. [3] Many cases were found to be associated with plaque type of morphea (29-41%). Extra cutaneous complications associated are joint contractures (50-56%), arthritis (40%), carpel tunnel syndrome, vasculitis and rarely visceral involvement like pleural, pulmonary and pericardial involvement. [2]

Laboratory investigations of the affected individuals usually show peripheral eosinophilia (58-85%), hypergammaglobulinemia (36-46%), monoclonal gammopathy and elevated ESR.[2]

Histopathological examination of a full thickness skin biopsy including the fascia remains the standard diagnostic test which reveals thickening, hyalinization & fibrosis of deep reticular dermis and subcutaneous tissue with mild inflammatory infiltrates. [4]

Additional imaging studies of deep fasci and muscles aids the diagnosis. Magnetic resonance imaging being a non-invasive and a rapid investigation reveals increased T2 signal in thickened deep fasciae and fascia. MRI aids in the diagnosis, to study the disease activity and treatment response. [5] 18-F-fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PT/CT) or ultrasound can be tried in cases were MRI couldn't be performed.[2]

The main differential diagnosis of EF is systemic sclerosis and the features that distinguishes both these conditions are, in EF there is no involvement of distal extremities, Raynaud's phenomena, nailfold capillary changes or digital ulcers. [2]

EF responds well with systemic glucocorticoids hence being the first line of management & can be combined with steroid sparing agents, among which Methotrexate being the common agent with a better prognosis when diagnosed early to prevent long term complications. [1] In 10-20 % patients, spontaneous resolution can be seen.[6]

At present there are new therapies which can produce a better outcome and prognosis in these patients. Studies in which treatment with IL-6 targeted drugs, such as Tocilizumab at a dose of 162mg for 3 months in steroids refractory cases of EF with improvement have been described. [7]

Early detection and timely management of eosinophilic fasciitis may improve morbidity, quality of life and remission of the disease. [6]

CONCLUSION

Shulman syndrome, also known as Eosinophilic fasciitis, is a rare disease with a varying clinical spectrum, age of onset and laboratory changes making the diagnosis and treatment a challenge. Though this disease being rare, a complete knowledge regarding the clinical signs of this entity helps to arrive at the diagnosis even in its initial phase. Dermascopy being an upcoming noninvasive tool for the diagnosis

of many dermatological entities also aids in the early recognition of these signs. Our report emphasizes the pivotal role of dermoscopy in facilitating the early diagnosis of eosinophilic fasciitis.

Declaration by Authors

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