EOSINOPHILIC FASCIITIS

(A Case Report)

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Introduction

Over 200 cases of eosinophilic fasciitis have been reported after the initial report of 2 cases by Shulman in 1974 [1]. The syndrome of eosinophilic fasciitis is now accepted as a specific clinical condition characterized by firm "puckered" skin bound to the underlying tissue, predominantly involving the extremities along with peripheral eosinophilia, raised erythrocyte sedimentation rate and hypergammaglobulinemia. The hands and feet are often spared and Raynaud's phenomenon is not present in contrast to scleroderma. Histopathological examination shows marked thickening of fascia with inflammatory cell infiltrate consisting of lymphocytes, plasma cells and eosinophils [2,3]. Majority of cases show prompt response to prednisolone therapy [4,5].

A case of eosinophilic fasciitis is being reported for its rarity and good response to steroids.

Case Report

A 45-year-old male was admitted with complaints of swelling, redness and induration of skin over extremities, trunk and chest of 9 months duration. The swelling, redness and pain started around the ankles after physical exertion. It extended upwards in symmetrical fashion involving legs, thighs, arms, forearms, trunk and chest. There was no history of fever, sore throat, burning micturition, dysphagia, dyspnoea, palpitation, drug intake, diabetes mellitus or Raynaud's phenomenon.

Dermatological examination revealed symmetrical, non-tender induration of skin involving arms, forearms, thighs, legs, trunk and chest. Hands, feet and digits were spared. The induration was irregular giving rise to puckering and cobblestone appearance to the skin. General physical and systemic examination of cardiovascular, respiratory system and abdomen were within normal limits.

Investigations : HB 13.5 gm per cent, TLC 7800/Cumm,

P-43 L-24, M-2, E-31 (per cent), ESR 36 mm fall at the end of 1st hour (Wintrobe method), absolute eosinophil count 2420 eosinophils per cumm and platelet count 1,70,000 per cumm. Bleeding time, clotting time, peripheral blood smear, blood urea and sugar were within normal limits and no haemoparasites were detected. No abnormality was detected on urinalysis and stool examination. Liver function tests, SGOT, SGPT, CPK, ASO titre and serum creatinine were within normal limits. Rheumatoid factor, LE cell phenomenon, VDRL and AMF were negative. Immunoglobulin profile, Ig G-837 mg, IgA-412 mg, and IgM 363 mg (all in per cent). No β -haemolytic streptococci were grown on throat swab culture. Electrocardiographic tracings were within normal limits.

Radiograph of skull, chest, knees and ankles were normal. Histological examination of skin biopsy including deep fascia overlying muscle showed marked thickening of fascia with inflammatory cell infiltrate consisting of lymphocytes, plasma cells and eosinophils. Epidermis, dermis and subcutaneous tissue were normal.

Taking into account the clinical profile, peripheral eosinophilia, raised erythrocyte sedimentation rate, increased level of IgA, IgM and histological findings; the patient was diagnosed as a case of eosinophilic fasciitis. The patient was treated with oral prednisolone 40 mg daily for a period of 3 months. The patient showed good response in the form of regression of swelling, redness and induration. After 3 months the dose of prednisolone was gradually reduced and then tapered off over a period of 2 months. The patient was followed up for 1 year. There was no recurrence or residual lesions.

Discussion

It may be difficult to differentiate eosinophilic fasciitis from the skin involvement of progressive systemic sclerosis. Though, some authors consider eosinophilic fasciitis to be a form of scleroderma, the unique clinical appearance and histopathology merit its consideration as a separate disease [2,3].

Induration and tightness of skin predominantly over the extremities with "peau d' orange" appearance, sparing of hands, feet and digits; absence of

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systemic involvement of Raynaud's phenomenon differentiate it from progressive systemic sclerosis. More than 90 per cent cases show peripheral blood eosinophilia at some time during the course of disease, though it may be variable and transient. In over 50 per cent cases erythrocyte sedimentation rate is elevated [3-5].

In original studies hypergammaglobulinemia was described but in subsequent studies it has only been observed in 25 to 72 per cent of cases [1-3].

Mild elevation of muscle enzymes has been observed in most of the cases [2-4]. Positive antinuclear factor and rheumatoid factor have been reported in less than 20 per cent of the cases [5,6].

Apart from characteristic induration of the skin over extremities, trunk and chest, our patient had peripheral blood eosinophilia varying from 31 to 11 per cent during the course of disease. He also had high erythrocyte sedimentation rate along with elevated levels of IgA and IgM. Histopathological findings of fascial thickening with inflammatory

cell infiltrate consisting of lymphocytes, eosinophils and plasma cells in our patient were consistent with diagnosis of eosinophilic fasciitis.

REFERENCES

1. Shulman LE. Diffuse fasciitis with eosinophilia: a new syndrome. *Arthritis Rheum* 1977; 20 suppl: 2055-155.
2. Barnes L, Rodnan GP, Medager TA, Short D. Eosinophilic fasciitis: a pathologic study of twenty cases. *Am J Pathol* 1979; 96: 493-517.
3. Kent LT, Cremer SF, Moskowitz RW. Eosinophilic fasciitis: clinical, laboratory and microscopic consideration. *Arthritis Rheum* 1981; 24: 677-9.
4. Michet CJ, Doyle JA, Ginsburg WW. Eosinophilic fasciitis: report of 15 cases. *Mayo Clin Proc* 1981; 56: 27-34.
5. Lakhanpal S, Ginsburg WW, Michet CJ, Doyle JA, Moore SB. Eosinophilic fasciitis: clinical spectrum and therapeutic response in 52 cases. *Semin Arthritis Rheum* 1988; 17: 221-3.
6. Sontheimer RD, Eumer RL, Geppert TD, Cohen SB. Connective tissue diseases. In: Moschella SL, Hurley HJ, editors. *Dermatology*. 3rd ed. Philadelphia: WB Saunders, 1992; 1217-51.