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Eosinophilic Fasciitis (Shulman Syndrome) in Association with Morphea and Systemic Sclerosis

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Abstract. A case of Shulman syndrome or diffuse fasciitis with eosinophilia is reported. Both superficial scleroderma (morphea) and systemic sclerosis accompanied the subcutaneous changes.

Key words: Shulman syndrome; Diffuse fasciitis with eosinophilia; Morphea; Systemic sclerosis

Shulman (3, 4) described a syndrome which he termed diffuse fasciitis with eosinophilia. Over 20 cases have now been reported (1) in which the usual features are a rapidly progressing sclerosis of the skin over limbs, contractures, eosinophilia and immunoglobulin G elevation, but an absence of Raynaud's phenomenon and a good response to anti-inflammatory treatment. The fascia between fat and muscle is greatly thickened, with collagen hypertrophy and infiltration by lymphocytes and plasma cells.

We report such a case which in addition showed both superficial and systemic involvement, so linking the Shulman syndrome with the more commonly encountered forms of scleroderma.

CASE REPORT

A 59-year-old man reported an 8-week history of progressive tightening of skin over arms and legs following ankle swelling attributed to unaccustomed exertion. Skin on forearms and shins was pigmented and firmly bound down to muscle and bone. Proximal limbs and trunk were less affected. Wrist, elbow, knee and ankle joint movement was limited. Digits were unaffected. The margins of the sclerosis on feet and wrists showed erythema.

Investigations showed: erythrocyte sedimentation rate, 36–40 mm in first hour; eosinophil count in peripheral blood, $152-1008/\text{mm}^3$; serum immunoglobulins normal; negative RA Latex test; antinuclear antibodies present to dilution of 1 in 16; partial right bundle branch block on ECG; normal peristalsis on barium swallow; no calcification of subcutaneous tissue in limbs; chest X-ray showed heart size at upper limit of normal; fibrosis was present in both lower lobes; respiratory function studies indicated restrictive and obstructive defects; carbon monoxide transfer factor was significantly reduced at 18 ml/min/ mm^2/hg . Electromyography of limb muscle showed normal activity. Biopsy of abdominal skin showed thick collagen in the dermis, consistent with morphea or localized superficial scleroderma, while a deep forearm biopsy not only showed excess collagen in dermis but also the presence of thick collagen bundles in subcutaneous fat and a marked thickening and oedema of the muscle fascia in which there was a heavy perivascular infiltrate composed mainly of lymphocytes with moderate numbers of plasma cells. Occasional histiocytes and polymorph leukocytes were also seen, but eosinophils were scanty. Blood vessel walls were slightly oedematous but there was no evidence of vasculitis. Direct and indirect immunofluorescence examination for C3, fibrinogen and immunoglobulins G, M and A in epidermis, dermis, fascia and muscle was negative.

During initial treatment with D-penicillamine in a dose of 125 mg daily for 4 weeks, increasing to 250 mg daily for a further fortnight, there was resolution of sclerosis in the trunk skin. Weight loss of 10 kg during this time led to the substitution of 40 mg prednisolone daily for the D-penicillamine. Over the following 8 weeks, there was complete clearing of sclerosis from upper arms and thighs except for some puckering of skin on the inner aspects, a feature often reported in diffuse fasciitis. However, there has been no obvious improvement on forearms and lower legs where the skin remains firmly bound down.

DISCUSSION

Except for the absence of demonstrable immunoglobulin abnormalities in the tissues, our patient shows the constellation of clinical and histological features described in the Shulman syndrome or diffuse fasciitis with eosinophilia. In addition, however, he has both dermal sclerosis and lung involvement consistent with systemic sclerosis. A recent review of 16 cases of the Shulman syndrome reported from the Mayo Clinic (2) showed that 5 had superficial dermal changes, while another 4 had pulmonary and 3, oesophageal involvement.

We would support this view of the Shulman syndrome as being a form of subcutaneous scleroderma that has a greater inflammatory element than other forms of scleroderma and hence a greater likelihood of responding to prompt anti-inflammatory therapy.

but which may also progress to mild systemic sclerosis.

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Localized Polyarteritis Nodosa in the Lower Limb with New Bone Formation

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Abstract. A 61-year-old male patient, with a burned-out ankylosing spondylitis, developed polyarteritis in the right lower limb, associated with periosteal new bone formation in tibia and fibula. An acute exacerbation may have been precipitated by an infection with yersinia. HLA type: A 28, 26, B 5, 27, Cw1.

Key words: Polyarteritis nodosa; Vasculitis; New bone formation; Yersiniosis

Localized polyarteritis nodosa is a vasculitis in small and medium-sized muscular arteries in cutis, subcutis and occasionally skeletal muscles, particularly in the lower limb (1, 2). The clinical picture is characterized by recurring pain in the involved extremity, livedo reticularis, subcutaneous nodules,

ulcerations, and peripheral neuritis. The disease runs a benign course, in contrast to the systemic form.

A few cases of localized polyarteritis nodosa with simultaneous periosteal new bone formation have been published (5, 8). In 1974, 3 cases with new bone formation were reported from the Mayo Clinic (8) and a further 7 cases were collected from the literature. We report here one additional case on this rare pathological entity.

CASE REPORT

A 61-year-old male was admitted to hospital on suspicion of a tumour in the right lower limb.

At the age of 45 he had low back pain from an ankylosing spondylitis. The pains disappeared and the spondylitis burned out, with typical roentgenological sequelae.

Two years prior to hospitalization, scaly, red plaques, bluish discoloration and periodic ulceration appeared on the upper part of the right calf. After nearly 2 years the patient developed increasing swelling, pains, and weakness in the right calf. During this exacerbation there was fatigue and a slightly elevated temperature.

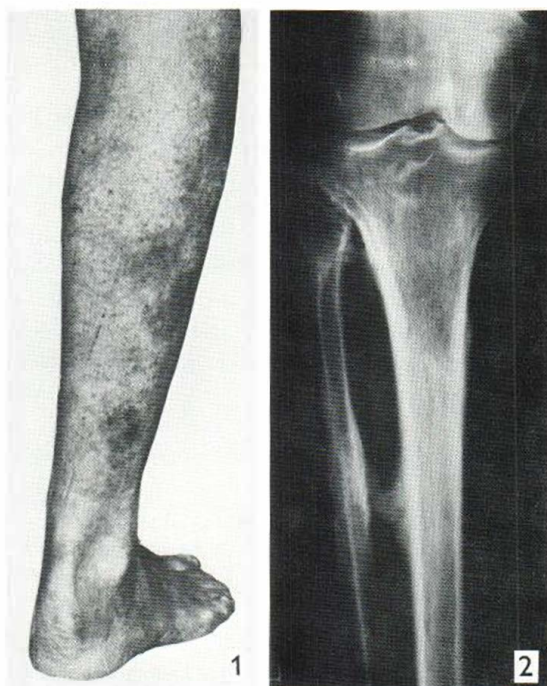


Fig. 1. Blotchy, bluish discoloration on and firm infiltration of the right calf.

Fig. 2. Radiograph of right calf. Periosteal new bone formation and sclerosis of the endosteal osseous structure are revealed.