ATYPICAL MULTICENTRIC RETICULOHISTIOCYTOSIS

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A 38-year- old male had arthritis since 8 years and erythematous papules, plaques, cutaneous and subcutaneous nodules over face, ears, trunk, and extensors of arms since 2 years. Histopathologically, presence of multiple foreign body giant cells confirmed the clinical diagnosis of multicentric reticulohistiocytosis. Unusual associations were: tapered fingers with depressed scars on their tips, low ESR - (5mm Ist hour). Raynaud's phenomenon and exaggeration of lesions after methotrexate.

Key words: Multinucleated giant cells, Arthritis, Rheumatoid factor, Multicentric reticulohistiocytosis

The term"multicentric reticulohistiocytosis" (MR) was coined by Orkin and associates in 1968.[1] MR can be cutaneous or cutaneous with systemic involvement.

Symmetrical polyarthritis with negative RA factor precedes the nodular eruption in nearly two-third cases. The interphalangeal joints are most commonly affected, followed by knees, shoulders, wrists, hips, ankles, feet, elbows and vertebral joints. The arthritic process is destructive and in advanced cases, opera gass hand, telescopic fingers, accordian or concertina hands deformity can occur.

Multiple, hemispherical, non-tender, yellowish to reddish brown, pruritic nodules, 2-20 mm in diameter are seen mainly on face, nose, paranasal areas, ears, retroauricular areas, neck, upper trunk, extensors of elbows, forearms, hand, nail folds and knees. Other features are weight loss, fatigue, pyrexia, lymphadenopathy, hepatosplenomegaly, moderate elevation of BSK, reversal of A:G ratio, mild anaemia, hyperproteinaemia and pulmonary infiltration. Associated diseases include cancer (of colon, breast, bronchus, cervix, ovary), thyroid disorders like hypothyroidism, thyrotoxicosis, goitre, tuberculosis, and diabetes.[3]

Histopathologically, multinucleated ginat cells, large PAS positive lipid containing histiocytes with ground glass cytoplasm are seen.

Case Report

A 38-year-old male had arthritis since 7-8 years, affecting joints of hips, neck, shoulders, hands, knees, elbows, wrists, and ankles. Arthritis improved initially for 2-3 years with steroid and remained well for next 3 years without steroids but relapsed and progressed with associated generalised myalgia since 2 years.

He had asymptomatic progressive, ery-

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thematous, papulonodular lesions which affected both elbows and ears, face, neck, scalp, trunk upper thighs and palmar surfaces of both hands and nail folds. Rapid increase occurred since 3 months after methotrexate and antituberculosis treatment. Numerous, 1-2cm, grouped/discrete, freely mobile, bright-red nodules, some with smooth shiny surface and others with peau'd orange appearance were seen on scalp, forehead, face, V of neck, ears (Fig. I) and upper arms. Purplish-red, smooth, 1-3cm shiny, firm, dome-shaped non-tender freely mobile nodules were seen on extensors of elbows, (Fig.2) metacarpophalangeal and interphalangeal joints, palms and thighs. Tips of all fingers showed tapering and firm, purplish-red papules with central depression/superficial ulceration and a few scars of old healed lesions. Nail folds revealed typical coral beaded papules.

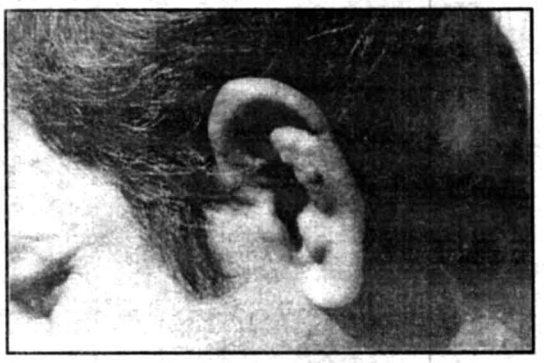

Fig.1 - Multiple, 1-2cm, mobile, bright red nodules some with smooth shiny and others with peau'd orange surface on pinna of left ear.

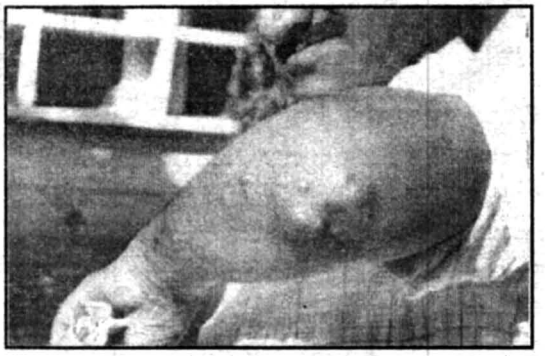

Fig.2 - Multiple, 1-3cm, purplish-red, smooth, shiny, firm, mobile, telangiectatic nodules on extensor surface of left elbow and arm.

Patient had polyuria (8L/day), Raynaud's phenomenon and generalized body aches. General physical and systemic examinations were normal. He was diagnosed as a case of pulmonary tuberculosis on the basis of positive Mantoux test, bilateral pleural effusion on x-ray chest although sputum was negative for AFB. Two months after ATT, he developed maculopapular rash all over the body associated with pruritus and ATT was then discontinued.

Investigations revealed ESR-5mm (Ist hour) serum calcium 11.2g%, 24 hour urinary calcium-1400mg/, RA factor-positive (1:4), ANF - positive (nucleolar pattern) Mantux test positive (15x25mm), x-ray knees soft tissue swelling, x-ray hands - cystic lesions with generalised osteoporosis and USG abdomen-splenomegaly. Rest of the detailed investigations including Hb, total and differential leucocyte count, urinalysis, kidney and liver function tests, serum cholesterol, widal test, pulmonary function test, synovial fluid cytology, C3 levels, x-ray skull and chest and bone marrow biopsy were normal. Screening for HIV was negative.

Skin biopsy revealed numerous large histiocytes showing an abundance of eosinophilic, homogenous to fine granular cytoplasm having a ground glass cytoplasm. The cells had either single or numerous irregularly distributed nuclei. Focal lymphocytic infiltrate was seen.

Discussion

Multicentric reticulohistiocytosis is a rane disease. Its atypical form is still rarer. The case reported here has all features suggestive of multicentric reticulohistiocytosis. It was atypical in that urinary calcium was 1400mg/24 hour, Mantoux test was highly positive (15x25mm), ESR was low (5mm-Ist hour) and there was exaggeration of lesions after methotrexate. Other unusual associated features were tapered fingers with depressed scars on their tips, positive Raynaud's phenomenon, ANF and RA factor. Sarcoidosis in our case was excluded mainly histopathologically.

References

1. Orkin M, Goltz RW, Good RA, et al. A study of multicentric reticulohistiocytosis. Arch Dermatol, 1964; 89: 640-645.

2. Montgomery H, Polley HF, & Pugh DG. Reticulohistiocytoma (Reticulohistiocytic granuloma). Arch Dermatol 1958;77:61-72.

3. Barrow MV, Holubar K. Multicentric reticulohistiocytosis. Medicine (Baltimore) 1969;48:287.

4. Lever WF, Lever GS. Lipidoses, in: Histopathology of the Skin, 7th edn., Lippincott, Philadelphia, 1988;400-402.