MULTICENTRIC RETICULOHISTIOCYTOSIS

Mohan B Gharpuray, Vinay Kulkarni and Raju Shah

A 55-year-old male had papulo-nodular and plaque type lesions on upper eyelids, chest, neck and extremities. He had joint pains and abnormal lipid profile. The histopathology showed typical picture of reticulohistiocytosis.

Key words : Multicentric reticulohistiocytosis.

Multicentric reticulohistiocytosis (MR), also called lipoid dermatoarthritis or reticulocytoma cutis,[1] is a rare systemic granulomatous disease of unknown aetiology. It is characterized by involvement of skin, mucosa, synovia, bones and internal organs and distinct histopathological features. The cutaneous nodules and destructive arthritis are the most common and prominent clinical features. Paraneoplastic character of MR is possible.[2]

Caro and Senear[3] in 1952 first coined the term reticulohistiocytic granuloma. Ackermann[4] reintroduced it recently. Lever and Schumburg-Lever[5] use the term reticulohistiocytic granuloma when the lesions are solitary. However, majority still stick to the term MR used by Goltz and Laymon[2] in 1954, because it emphasizes its multifocal origin[6] and alludes to its systemic nature. Hardly about 80 cases of this disease have been reported.[2]

The sex distribution in MR is 3 : 1 in favour of females.[2] The hallmark of the disease is a granulomatous proliferative process of histiocytes, some of which are multinucleated and lipid laden. The aetiological stimulus is not known. Neither any metabolic factor, nor any infective agent has been identified. No data as regards a possible compromise of host immune system is available.

Clinically, in two-third patients arthritis develops prior to the development of skin lesions. In one out of five, skin lesions precede bone involvement, while in another one out of five both get affected simultaneously. About 50% patients eventually develop mucous membrane lesions.

The most commonly affected areas of the skin are face, especially the nose and paranasal areas, hands, especially nail folds; ears, forearms, retro-auricular areas of scalp; neck and occasionally trunk. Hemispherical, non-tender, reddish-brown nodules varying in diameter from a few mm to several cm are seen. Pruritus is a common symptom but the nodules rarely ulcerate. The joint involvement is usually symmetrical and interphalangeal joints are most commonly affected, followed by knees, shoulders, wrists, hips, ankles, feet, elbows and vertebral joints. The arthritic process is destructive (arthritis mutilans) and leads to deformities. The process of destruction may be rapid, but it gradually burns out. Radiological involvement is often severe when the clinical picture is mild. In far advanced cases, fingers may be considerably shortened but can be pulled out to full length. This deformity is often called opera glass hand, telescopic fingers or concertina hand. The mucous membrane lesions mimic skin lesions. The other features which may be present are xanthelasmata, infiltrates along the tendon sheaths, myocardium, lungs and lymph nodes.

The results of laboratory investigations are non-specific. The histopathology is characteristic, and multinucleated giant cells and large

From the Dermatology Clinic, Continental Chambers, Karve Road, Pune-411 004, India.
Address correspondence to : Dr M B Gharpuray.

PAS positive lipid-containing histiocytes with ground glass cytoplasm are seen. Electron microscopy has failed to reveal the presence of Langerhan's granules. The cellular infiltrate otherwise is non-specific and lymphocytes, neutrophils and eosinophils are seen in the lesions having scales.

A possible paraneoplastic character has been considered and Catteral and White[7] have reported the presence of malignancies of ovary, stomach, colon, breast and cervix in patients with MR.

The treatment is unsatisfactory and local, systemic and intralesional corticosteroids; and anti-inflammatory drugs have all failed to give relief. Recently, encouraging results have been obtained with topical nitrogen mustard.[8]

We are reporting a case of MR.

Case Report

A 55-year-old male developed asymptomatic nodular lesions on both the eyelids $2\frac{1}{2}$ months ago. Similar lesions then appeared on the neck, arms, forearms and thighs. The onset was acute. He suffered from joint-pains off and on prior to the appearance of skin lesions. There was no history suggestive of any associated disorder like diabetes mellitus, hypertension, ischaemic heart disease, acute pancreatitis or any internal malignancy. He was under treatment for cervical spondylosis and had a non-healing ulcer following an injury on the left leg for which skin grafting was done 7 months back.

The lesions were nodules and well-defined plaques, soft to firm in consistency, non-tender and yellow to skin-coloured, present on both eyelids, nape and V of neck, arms and forearms, groins and thighs. These were bilaterally symmetrical. Multiple small joints of the hands were inflamed and tender suggesting arthritis. Systemic examination revealed no abnormality.

Laboratory investigations showed hemoglobin 8.5gm%, TLC, DLC, urine were normal, blood sugar fasting was 82 mg% and post-prandial 112 mg%, serum cholesterol 272 mg%, beta lipoproteins 475 mg%, pre-beta lipoproteins 416 mg% and chylomicrons 35 mg% (suggestive of type IV hyperlipidemia). X-ray chest and ECG were normal. X-ray of hands showed only osteoporosis, there was no cartilage or bone destruction.

Skin biopsy showed epidermal atrophy, the dermis showing multiple, large, histiocytes with ground glass cytoplasm. Majority of these cells were multinucleated. An occasional giant cell showed wreath pattern.

He was given anti-inflammatory drugs but had very little benefit.

Comments

Our patient showed characteristic cutaneous and bone involvement. The joint involvement had preceded the cutaneous manifestations. The mucous membranes were not involved. Apart from type IV hyperlipidemia, there were no other metabolic abnormalities. The histopathology was characteristic. Giant cells with ground glass cytoplasm is a characteristic of MR, but the presence of wreath pattern is not described. It was probably related to the abnormal lipid metabolism. This biochemical and histopathological association has not been reported and may be a co-incidental finding.

References

1. Greaves MW : Histiocytic proliferative disorders, in : Textbook of Dermatology, 4th ed, Editors, Rook A, Wilkinson DS, Ebling FJG et al : Oxford University Press, Bombay, 1987; p 1709-1710.
2. Holubar K : Multicentric reticulohistiocytosis, in : Dermatology in General Medicine, 3rd ed, Editors, Fitzpatrick TB, Eisen AZ, Wolff K et al : McGraw Hill, New York, 1987; p 1870-1874.
3. Caro MR and Senear FE : Reticulohistiocytoma of the skin, Arch Dermatol, 1952; 65 : 701 (Quoted by 2).

4. Ackermann AB : Histologic Diagnosis of Inflammatory Skin Diseases, Lea and Febiger, Philadelphia, 1978; p 472.

5. Lever WF and Schaumburg-Lever G : Lipidoses, in : Histopathology of the Skin, 6th ed, JB Lippincott, Philadelphia, 1983; p 400-402.

6 Fast A : Cardiopulmonary complications of multicentric reticulohistiocytosis, Arch Dermatol, 1976; 112 : 1139.

7. Catteral MD and White JE : Multicentric reticulohistiocytosis and malignant disease, Brit J Dermatol, 1978; 98 : 221-224.

8. Brand F : Topical nitrogen mustard therapy in multicentric reticulohistiocytosis, J Amer Acad Dermatol, 1982; 6 : 260.