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MULTICENTRIC RETICULOHISTIOCYTOSIS

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This is the first reported case from India of multicentric reticulohistiocytosis, manifesting as multiple cutaneous nodules on the face, ears, elbows, wrists, knees and hands with mild pruritus. Polyarthritis involved the small and big joints. Histopathology confirmed the diagnosis. All the lesions disappeared in 1½ years time.

Key words : Reticulohistiocytosis, Multicentric.

Multicentric reticulohistiocytosis is a rare disease of skin and joints with distinctive histopathologic findings. Weber and Freudenthal in 1937[1] described systemic lesions in multicentric reticulohistiocytosis. Caro and Senear in 1952[2] reported two cases of multicentric reticulohistiocytosis and provided evidence that it was disorder of the reticuloendothelial system. Synovial membrane biopsy changes were similar to the changes seen in the skin. In 1954, Croltz and Laymon[3] used the term multicentric reticulohistiocytosis to differentiate those patients who had cutaneous nodules and definite arthritis from those with only cutaneous lesions.

Clinically, multiple, yellowish to brownish, pruritic nodules 2 to 20 mm in diameter may be present on the scalp, face, neck, ears, upper part of trunk, extensor surfaces of elbows, knees and hands. Mucosal papules are seen in about half the cases.

A symmetrical polyarthritis, insidious in onset, precedes the nodular eruption in nearly two-thirds of the cases. Women are affected more commonly. Large joints such as knees, hips and shoulders as well as smaller joints are effected. There is destruction of the interphalangeal joints which leads to accordion or concertina hands deformity. The arthritis is variable in behaviour, it persists in some cases and disappears in others without any reason. Orkin et al in 1964[4] reported that one-fifth of the patients had mutilating arthritis of the hands. Other features were weight loss, fatigue, pyrexia, lymphadenopathy, hepato-splenomegaly, tuberculosis, moderate elevation of ESR, reversal of A/G ratio, mild anaemia, hyperproteinaemia and pulmonary infiltration. Barrow in 1967[5] reported nail atrophy, longitudinal ridging, brittleness and hyperpigmentation etc. Taylor in 1977[6] treated one case of multicentric reticulohistiocytosis with systemic corticosteroids.

The striking histopathologic pattern is the presence of aggregates of histiocytic giant cells and histiocytes in the skin, synovia, mucosa and even bones. The histiocytic multinucleated giant cells can be as large 100 mm and multiple (upto 20) haphazard nuclei may be seen. Histiocytes and giant cells have finely granular pale eosinophilic cytoplasm with a ground-glass appearance. Nodular infiltrates may occupy the entire dermis and are unencapsulated. Other cell types are lymphocytes, eosinophils, plasma cells, RBCs and fibroblasts. The connective tissue stroma is pushed aside by the cellular masses and becomes vascularised. Histochemical studies indicate that the giant cells and histiocytes contain a PAS-reactive material that may be glycoprotein or mucoprotein, neutral fat, phospholipids, iron and melanin.

Case Report

A 32-year-male had rheumatoid arthritis of six months duration, first starting in the right knee and then involving the left knee, both elbows, wrists and hand joints within two

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months. At that stage, he also developed a generalised nodular eruption especially marked on the face, ears and extensor surfaces of elbows, knees and hands with mild pruritus. There was no fever or systemic illness, but he had loss of weight and fatigue. General physical and systemic examination were normal. There was swelling of the small joints of both hands, wrists, elbows and knees and painful restricted movements. Multiple, firm nodules, yellowish to brown in colour, varying in size from 5 mm to 2 cm and mobile over the underlying structures were present all over the body but were more prominent over the pinna of both ears and extensor aspect of elbows, knees and hands. There were no lesions on the mucosae. The investigations revealed, Hb 10 gm%, TLC 9700/cmm, DLC (P 69, L 26, M 3, E 2), rheumatoid factor negative twice, ESR 30 mm, serum uric acid 3 mg%, and serum cholesterol 235 mg%, X-ray both knees showed soft tissue swelling (Fig. 1). Histopathology showed large, round to oval histiocytes with abundant, eosinophilic, finely granular cytoplasm with a ground-galss appearance and with irregularly distributed multiple nuclei. There were empty spaces and fibrous tissue surrounding the giant histiocytes. Some histiocytes had single nuclei and a few inflammatory cells were also present (Fig. 2).

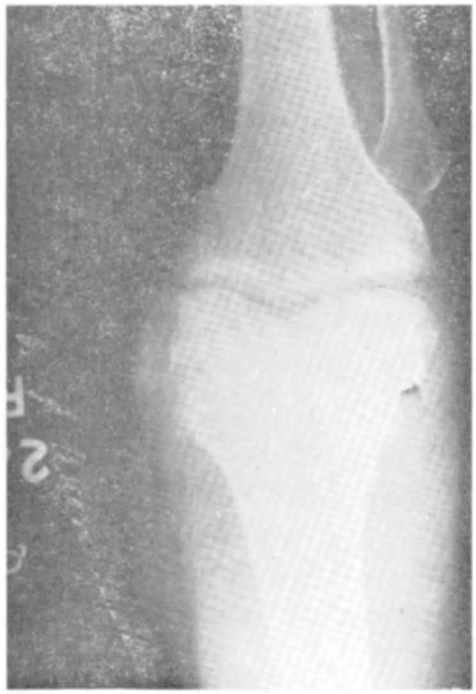

Fig. 1. Skiagram of right knee showing soft tissue swelling.

Fig. 2. Large histiocytic giant cells with peripheral empty spaces, with abundant cytoplasm and irregularly distributed multiple nuclei. Some histiocytes with single nuclei and a few inflammatory cells are also seen (H & E×400).

Comments

Exact aetiology of multicentric reticulohistiocytosis is not known. Johnson and Tilden in 1957[7] reported that though giant cells contain little or no sudanophilic lipid material, yet this condition may be related to the other histiocytic diseases such as xanthomatoses and the so-called lipoid storage diseases of the reticulo-endothelial system as many multicentric reticulo-histiocytosis

cases also have high serum cholesterol, xanthomatous lesions of eyelids and PAS positive glyco or lipoprotein and lipoid complex as in giant cells. So it was believed that mutlicentric reticulo-histiocytosis is a specific type of benign, non-neoplastic histiocytosis affecting the skin and joints as suggested by Croltz and Laymon.[3] It was thought that the histiocytic reaction may be in response to primary degeneration of the collagen.

Montgomery et al[8] however, wrote that this disorder was not an essential disorder of lipoid metabolism or rheumatic disease or neoplasia and there was no explanation why some cases improved after a few years while the disease was progressive in others. In our cases also, the skin as well as joint lesions completely disappeared after about 1½ years. Orkin et al[4] thought that histiocytosis-X shared certain facts with multicentric reticulo-histiocytosis and suggested that the suffix osis was preferable to oma since the latter signifies a neoplastic process.

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