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Multicentric Histocytosis Mimicking Histoid Leprosy- A Case Report

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***Abstract*: -** *Multicentric reticulohistiocytosis (MRH) is a very rare systemic disease with variable phenotypic presentation having predominant joint and cutaneous manifestations most commonly affecting women in middle age. The diagnosis is often overlooked because of the rarity of this condition.*

Here in, we are reporting a case of Multicentric reticulohistiocytosis presenting as multiple erythematous nodules all over the body mimickin as Histoid leprosy along with systemic involvement.

***Keywords*: - Multicentric reticulohistiocytosis, ground glass appearance, giant cells, pulmonary fibrosis.**

Introduction

Multicentric reticulohistiocytosis (MRH) is a rare histiocytic disease of unknown etiology being first described by Weber and Freudenthal [1] in year 1937 but the term was given by Goltz and Layman in 1954 [2]. MRH is characterized by distinctive cutaneous papulonodular lesions predominantly affecting extensor surfaces and destructive polyarthritis with other systems being rarely affected .Around 20% cases have been found to have an internal malignancy.

The diagnosis is based on the Histopathological findings showing presence of multinucleate giant cells and oncocytic macrophage with finely granular cytoplasm and CD68 positivity on immunohistochemical staining.

Case report

A 45 year old Indian female patient presented to us with complaints of generalized itching and presence of multiple, reddish-brown, non-tender elevated lesions all over body predominantly over both palmar & dorsal aspect of hands, ears, toes, knees and elbows and face since 4 months. She also had joint pain and stiffness involving knee, feet, elbows, shoulders and ankles since 3 months. Pain responded poorly to NSAIDS. There were no other systemic complaints.

Dermatological examination revealed multiple flesh colored to reddish –brown, discrete, glistening papulo- nodular lesions of size varying from 0.1-0.6cm present all over the body predominantly over the dorsal and ventral surface of fingers and toes, being more prominent over joints and palmer aspect of hands, knees, elbows, face, lower lip and upper chest area. General physical examination showed pallor and bilaterally enlarged inguinal lymph node.

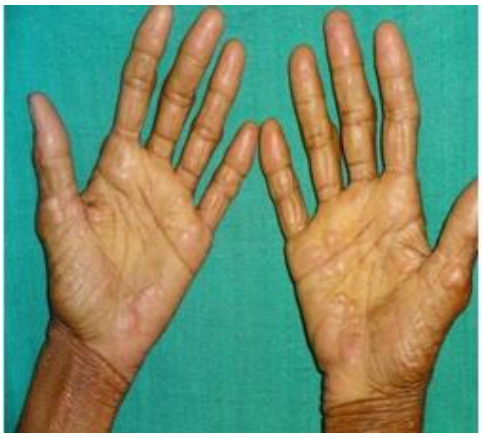

Figure 1(a) Nodular lesions involving palmer aspect of both hands.

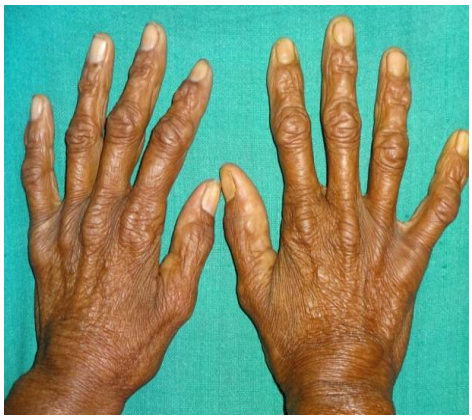

Figure 1(b): Nodular lesions involving dorsal aspect of both hands.

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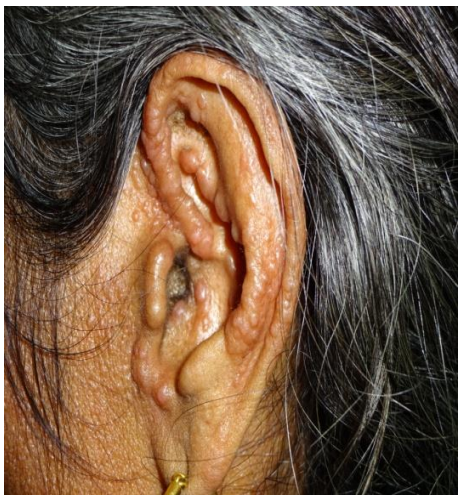

Figure 2: Multiple erythematous nodules over face including ears.

Laboratory investigations were done and patient had Hemoglobin- 8.6, CRP +ve, ANA+ve and ESR < 002 mm/ 1st hr as significant findings. Peripheral blood film showed microcytic hypo-chromic anemia. Total lipid profile was within normal range, Mantoux test was negative abdominal USG showed no Organomegaly. On chest X-ray radio-opaque shadow was present. In left middle zone. On further evaluation patient showed apical fibrosis in both lungs along with mediastinal, pre and para aortic, pretracheal lymph node enlargement on HRCT.

Histopathological examination from a nodular lesion revealed thinning of epidermis with flattening of rete ridge and dermal infiltration by numerous large histiocytic showing abundant eosinophilic granular cytoplasm with typical ground glass appearance surrounded by presence of chronic inflammatory cells. Cytoplasm showed PAS positive material.

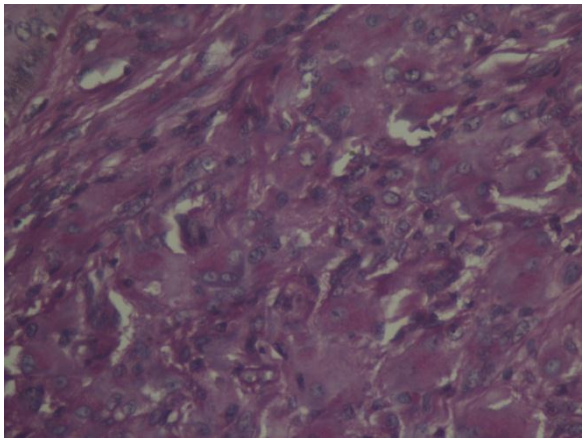

Figure 3: Skin biopsy specimen (H&E stain) in high power view showing diffuse infiltration of dermis by histiocytic cells with scattered multinucleated giant cells and abundant eosinophillic granular cytoplasm and presence of chronic inflammatory cells.

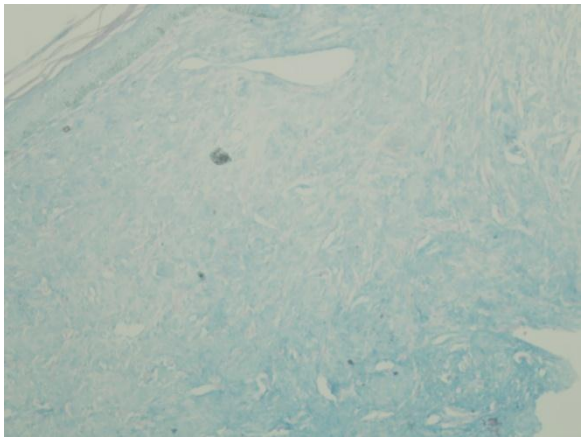

Figure 4: CD68 positivity on 100X magnification.

Histopathology was also positive for CD68 and vimentin. These findings were consistent with the diagnosis of Multicentric reticulohistiocytosis.

Discussion

Multicentric reticulohistiocytosis, also known as lipoid dermatoarthritis is a non-langerhans cell histiocytosis (class II) disorder with unknown etiology. The condition typically affects middle aged women, with a female to male ratio of 3:1 [3] and rarely affects children [4].

The typical cutaneous manifestations consists of flesh colored to reddish brown yellow papules and nodules that may be found anywhere over the body with a predilection for face, hands and around joints [5]. Coral bead like lesions may occur around nail folds, which may result in nail dystrophy. About 25% of patients complain of pruritus associated with skin lesions. These features were present in our case.

Differential diagnosis of these lesions includes lepromatous leprosy, sarcoidosis, xanthomatosis, histiocytosis X, juvenile and adult xanthogranuloma, generalized eruptive histiocytoma and neurofibromatosis. Mucosal involvement involving the mouth, gingiva, pharynx, larynx and sclera have been noted in more than 50% patients which is absent in our case.

Polyarthritis typically affecting the hands is seen in 60% of patients. The involvement is diffuse, symmetric, progressive and destructive with a predilection for distal interphalangeal joints [6]. Rarely other joints can be involved viz. the knees, shoulders, wrists, hips, ankles, feet, elbows, spine and temporomandibular joints.

Radiographic findings include destruction of the articular surfaces, with bone resorption and eventually secondary osteoarthritis. In our case, although there was presence of joint pain and stiffness, there was no significant abnormality on radiological evaluation.

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Constitutional symptoms like pyrexia and weight loss may occur. Other systems that are less commonly involved include bone marrow, skeletal muscle, tendon sheath, lymph nodes, heart, pericardium, lungs, pleura, bones, liver, duodenal mesentery and kidneys. In our case there was pulmonary involvement in the form of apical lung fibrosis on chest X ray and HRCT-chest.

Reported associated conditions include diabetes [7], Sjogren syndrome[8], hypothyroidism, hyperlipidemia, primary biliary cirrhosis, tuberculosis, systemic sclerosis, myopathy and pregnancy. Malignancies that are associated include carcinoma of breast, cervix, colon, ovary [9], stomach and bronchus, melanoma, mesothelioma, lymphoma, sarcoma and leukemia. However, no such associations were found in our patient.

The prognosis of the disease is good in the absence of underlying malignancy, the disease becoming quiescent in 7-8years. Various treatment modalities like systemic steroids, bisphosphonates, cyclophosphamide, azathioprine, cyclosporine, methotrexate, etanercept and infliximab have been tried with fairly good results.

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