Multicentric Reticulohistiocytosis: A Case with Minimal Cutaneous Features

Sir,

Multicentric reticulohistiocytosis (MRH) is a rare, multisystem non-Langerhans cell histiocytosis of unknown etiology that is characterized by symmetric polyarthritis and papulonodular skin lesions. Very few case reports exist worldwide and only 12 cases have been reported from India.

A 40-year-old male presented with debilitating joint pain and morning stiffness involving multiple joints associated with swelling of both wrist and knee joints as well as multiple asymptomatic skin colored lesions over hands, feet, ears, and forehead for the past eight months. On examination, there was painful swelling of both wrist and knee joints [Figure 1a and b]. Dermatological examination revealed discrete, firm, skin colored to reddish brown, non-tender papules of size 0.2 × 0.2 mm to 1 × 1 cm over hands, bilateral ears, forehead (Figure 1c-e). Similar lesions were also seen over the feet and a typical coral bead appearance of lesions was seen in th periungual rehion [Figure 1f]. Rest of the systemic examination was normal and there was no evidence of lymphadenopathy. Differential diagnosis of rheumatoid nodules and multicentric reticulohistiocytosis was kept. Routine hematological investigations, lipid and thyroid profile, rheumatoid factor, anti-nuclear antibodies, and C-reactive protein were normal. Malignancy screening (including ultrasound abdomen, stool for occult blood, urine analysis, and peripheral smear) was also negative. Radiological examination of knee joints showed bilateral subchondral sclerosis and suprapatellar effusion [Figure 2a and b]. Skin biopsy from papular lesions over the elbow revealed diffuse infiltration of the dermis by multinucleated histiocytes with an eosinophilic ground glass cytoplasm. CD68 stain was diffusely positive and the cells were negative for S100 [Figures 3 and 4]. The patient was started on tablet methotrexate, at a dose of 7.5 mg/week along with non-steroidal anti-inflammatory drugs (NSAIDs) as well as physiotherapy. Carbon dioxide laser ablation was planned for skin lesions. Significant improvement in joint pain and mobility was seen after 6-8 months of follow-up.

In approximately 60 to 70% of patients, polyarthralgia is the first manifestation, in 30% skin manifestations occur first and around 29% of the patients present with both skin and joint symptoms.[1] It is often associated with constitutional symptoms such as fatigue, weight loss, and fever but our case had no such symptoms. In the absence of effective treatment, the destruction of the proximal and distal interphalangeal joint may result in "arthritis mutilans" and "opera-glass hand" deformity. Our case had involvement of both knee and wrist joints.

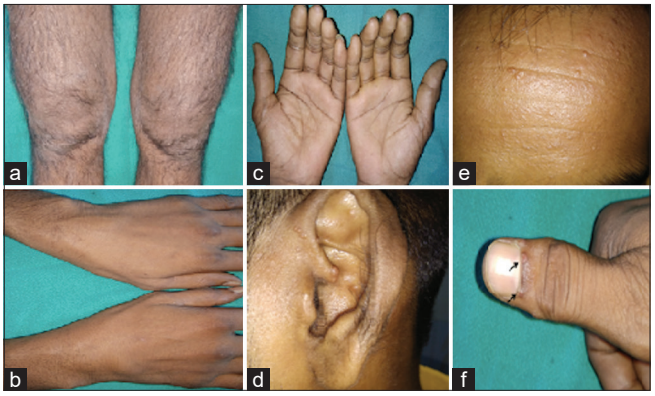

Figure 1: (a and b): Swelling of both knee and wrist joints. (c-e) Skin colored to reddish brown, papules and nodules over hands, feet, elbow, face, and bilateral ear. (f) Coral bead appearance of lesions in periungual area

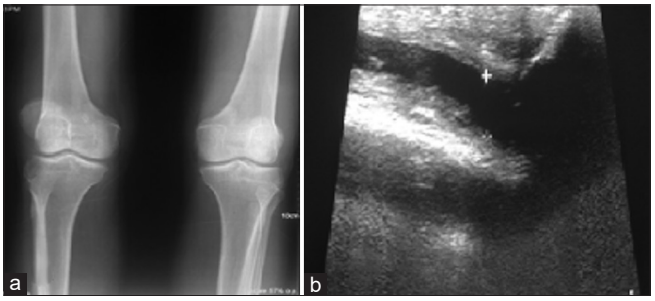

Figure 2: (a) X-ray of knee joints showed bilateral subchondral sclerosis (b) Ultrasound (USG) showing suprapatellar effusion in knee joint

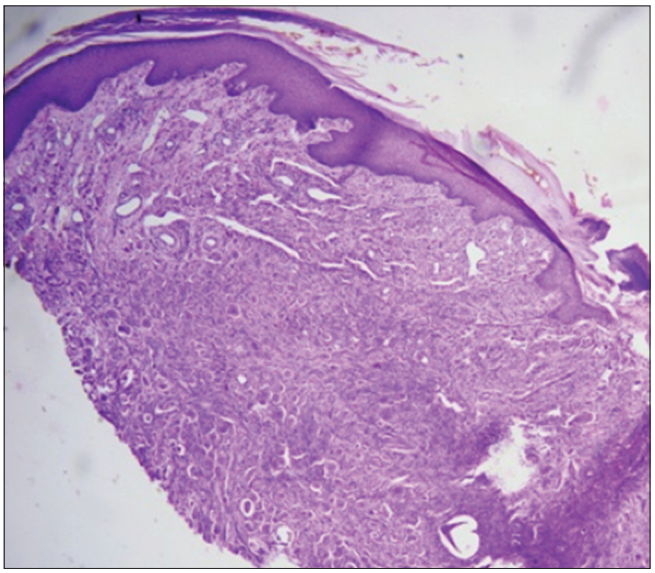

Figure 3: Diffuse infiltration of dermis by histiocytes (H and E, ×40)

Cutaneous manifestations are invariably seen and generally follow the joint involvement as slowly appearing translucent brown-reddish to flesh-colored papulonodular lesions. The

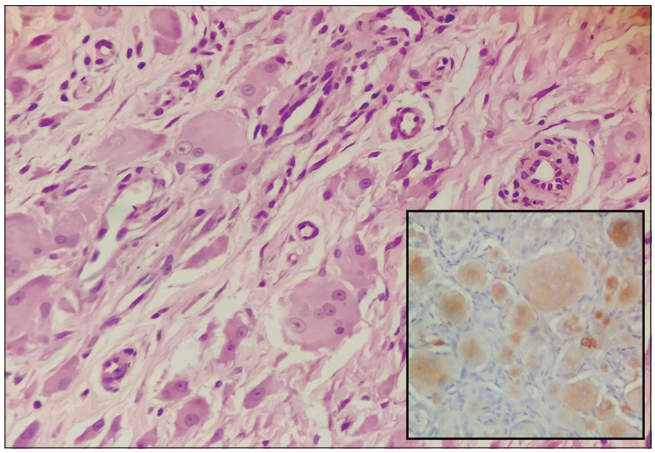

Figure 4: Higher magnification shows mononucleated and multinucleated histiocytes with an amorphous, eosinophilic, ground glass cytoplasm. The nuclei of cells were haphazardly arranged. There was a background of lymphocytes and collagenization (H and E, ×400). Inset: CD68 stain was diffusely positive (IHC ×400)

periungual lesions have a classic "coral beads" appearance. In our case, lesions were primarily papules with few papulonodules. MRH may also affect other organs like heart, lung, liver, and lymph nodules. Approximately, 25% of MRH cases have been associated with the presence of neoplasia like breast, cervix, and colon. Our case was not associated with any systemic involvement or malignancy. Coexistence of MRH with autoimmune diseases is reported in approximately 15% of cases.[2] Spontaneous remission often occurs within 10 years.

Diagnosis is based on histological and immunological features of the proliferating histiocytes. Early lesions manifest with lymphohistiocytic infiltrate in the dermis containing small histiocytes while more advanced lesions with pathognomonic multinucleated giant cells with ground glass eosinophilic cytoplasm. Additional staining includes a strong positivity for periodic acid-Schiff (PAS), CD68, andCD45, acid phosphatase, nonspecific esterase, and lysozyme. However, there is conspicuous negativity for S-100 protein, CD1a, and factor XIIIa.[3]

Due to the rarity of MRH, it has been difficult to establish well-defined treatment guidelines for this aggressive disease. NSAIDs and corticosteroids are generally utilized as first-line agents, with the latter used systemically and by intraarticular injection. Limited evidence supports the use of disease-modifying antirheumatic drugs, such as methotrexate, leflunomide, hydroxychloroquine, and azathioprine.[4] Methotrexate is usually given at low dose in combination with NSAIDs or corticosteroids for several months to years. Bisphosphonates and biologicals (infliximab, etanercept, adalimumab, and anakinra) have been proposed as promising therapeutic agents.[5,6] Skin lesions may be treated by carbon dioxide laser application or surgical excision.[7]

We report this case because of its rarity and characteristic histopathological findings. Our patient had an early form of disease as suggested by mild cutaneous and radiological features.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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