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Case Study

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MULTICENTRIC RETICULOHISTIOCYTOSIS: A COMPREHENSIVE CASE REPORT

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ABSTRACT

Multicentric reticulohistiocytosis (MRH), previously known as lipoid dermatoarthritis, is a rare systemic disorder characterized by cutaneous nodules and progressive polyarthritis. It primarily affects middle-aged women and can lead to joint destruction and deformities resembling rheumatoid arthritis. The etiology remains unclear, with possible associations with malignancies and autoimmune diseases. We present a case of a 52-year-old woman with multiple skin nodules and severe polyarthritis, leading to a diagnosis of MRH based on histopathological and immunohistochemical findings. Early diagnosis and intervention were critical in managing symptoms and preventing joint damage. This report highlights the diagnostic challenges and therapeutic options available for this rare condition.

INTRODUCTION

Multicentric reticulohistiocytosis (MRH) is a rare systemic histiocytic disorder first described by Weber and Freudenthal in 1952 (Weber & Freudenthal, 1952). It is characterized by a combination of cutaneous and articular manifestations, with nodular skin lesions and a rapidly progressive, erosive polyarthritis that can lead to severe disability (Barrow & Holubar, 1991). Although the exact pathogenesis remains unclear, MRH is considered a disorder of non-Langerhans histiocytes, with a possible link to underlying malignancies, autoimmune diseases, and metabolic disorders (Gibson et al., 2019).

Clinically, MRH presents in the fourth to sixth decade of life, with a higher prevalence in women. The skin lesions, often affecting the hands, face, and extensor surfaces, are composed of dermal histiocytic infiltrates (Winkelmann et al., 1975). The associated arthritis mimics rheumatoid arthritis (RA) but lacks rheumatoid factor and anti-citrullinated protein antibodies (ACPA) (Selva-O'Callaghan et al., 2006). Approximately 25-30% of cases are associated with malignancy, making cancer screening essential in affected patients (Kaye et al., 2021).

Due to its rarity and overlapping features with other arthropathies, MRH poses a diagnostic challenge. This case report presents a patient with MRH, emphasizing the clinical, histopathological, and therapeutic aspects of this uncommon disorder.

Case details

A 52-year-old woman presented to the clinic with a 12-month history of progressively worsening polyarthritis and multiple skin nodules. She reported pain and stiffness in her hands, wrists, and knees, with morning stiffness lasting more than an hour. The cutaneous nodules initially appeared on the dorsal aspects of her hands and later spread to the face and elbows. She denied systemic symptoms such as fever, weight loss, or night sweats.

Clinical Examination: On examination, the patient had multiple, firm, reddish-brown papulonodular lesions on the dorsum of her hands, perioral region, and extensor surfaces of the elbows. The affected joints were swollen, warm, and tender, with limited range of motion, particularly in the small joints of the hands and wrists. The characteristic "coral bead" sign, with periungual papules, was noted. There was no lymphadenopathy or hepatosplenomegaly.

Laboratory Investigations: Routine laboratory tests showed mild anemia (Hb 11.2 g/dL) and elevated inflammatory markers (ESR 45 mm/hr, CRP 22 mg/L). Rheumatoid factor (RF) and ACPA were negative. ANA and ANCA were also negative. Serum lipid profile and metabolic panel were within normal limits. A malignancy workup, including chest X-ray, mammography, and abdominal ultrasound, was unremarkable.

Radiological Findings: Hand radiographs revealed marginal erosions and joint space narrowing, particularly

in the interphalangeal joints, but no osteophyte formation or periarticular osteopenia, distinguishing it from RA. MRI of the hands showed synovial thickening and erosions without significant pannus formation.

Histopathological and Immunohistochemical Analysis: (Figure a, Figure b)
A biopsy of a skin nodule showed dermal infiltration by histiocytes with eosinophilic granular cytoplasm, multinucleated giant cells, and an absence of epidermal involvement. Immunohistochemical staining revealed strong positivity for CD68 and CD163, confirming the histiocytic origin. S100 and CD1a were negative, ruling out Langerhans cell histiocytosis. The findings were consistent with MRH.

Management and Follow-Up: The patient was started on methotrexate (15 mg/week) and low-dose prednisone (10 mg/day), leading to partial symptom relief. Given the severity of arthritis, adalimumab (40 mg biweekly) was added, resulting in significant improvement in joint symptoms and reduction in skin lesions. At one-year follow-up, she demonstrated stable disease with no radiographic progression or new nodular lesions.

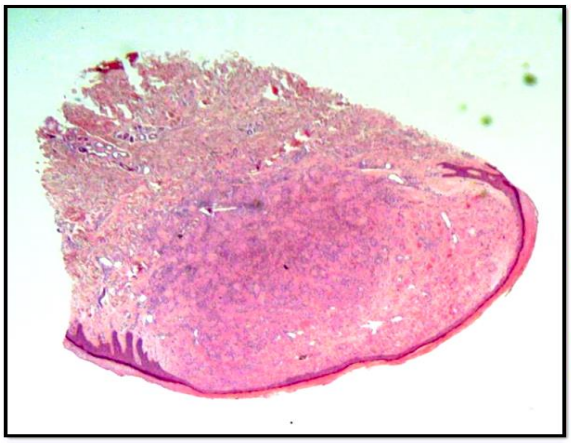

Figure a) 4x view

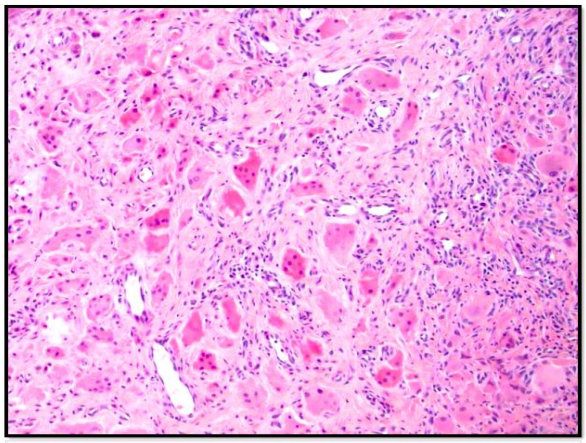

Figure b) 20x view

DISCUSSION

MRH is a rare disorder with significant clinical and diagnostic overlap with inflammatory arthritis and other histiocytic disorders. The cutaneous manifestations, particularly the characteristic periungual papules, are crucial for early recognition (Gibson et al., 2019). The histopathological presence of multinucleated giant cells with a ground-glass cytoplasm is a hallmark of MRH (Winkelmann et al., 1975).

The pathogenesis remains poorly understood but is thought to involve an aberrant histiocytic response with cytokine dysregulation. The association with malignancies, particularly breast, lung, and ovarian cancers, suggests a potential paraneoplastic phenomenon (Kaye et al., 2021). Autoimmune associations, including dermatomyositis and Sjögren's syndrome, have also been reported (Selva-O'Callaghan et al., 2006).

Management of MRH primarily involves immunosuppressive therapy to control arthritis and prevent joint destruction. Traditional DMARDs such as methotrexate, azathioprine, and hydroxychloroquine have shown variable success (Barrow & Holubar, 1991). Biologic agents, including TNF inhibitors and IL-6 inhibitors, have demonstrated efficacy in refractory cases (Gibson et al., 2019). Corticosteroids are useful for acute flares but are not recommended for long-term management due to side effects.

Regular malignancy screening is advised, given the reported cancer associations. Long-term prognosis varies, with some patients experiencing spontaneous remission, while others develop severe joint destruction necessitating surgical intervention (Weber & Freudenthal, 1952).

CONCLUSION

MRH is a rare systemic histiocytic disorder characterized by progressive polyarthritis and distinctive skin lesions. Due to its rarity and resemblance to other rheumatologic conditions, early recognition and histopathological confirmation are crucial. Immunosuppressive therapy, particularly biologics, plays a significant role in disease management. This case emphasizes the need for heightened awareness among clinicians to ensure timely diagnosis and intervention, ultimately preventing joint deformities and disability.

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