An unusual presentation of Vilanova disease (erythema nodosum migrans) with superficial histologic changes

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INTRODUCTION

Erythema nodosum migrans (ENM) is a rare clinical variant of erythema nodosum (EN). It is a septal panniculitis characterized by asymmetrical, mildly tender, nodules that migrate over time.

Fever, sore throat, or arthralgias may also accompany the cutaneous manifestations of the disease; however, this is uncommon. The treatment of choice for ENM is potassium iodide. We present a case of a 77-year-old woman with a 5- month history of ENM that was treated successfully with topical clobetasol.

CASE REPORT

A 77-year-old woman was referred to our dermatology clinic with a 5-month history of an enlarging, erythematous to brownish indurated lesion on the left thigh. The lesion was mildly tender and expanding centrifugally over the last 5 months. No physical trauma or infection preceded the lesion. Her medical history included a hip replacement surgery, cataracts, and hypothyroidism. She was taking L-thyroxin for her hypothyroidism and her thyroid-stimulating hormone levels were within the normal range. Physical examination found a large annular plaque on the medial left thigh with an erythematous mildly tender border (Fig 1). No overlying ulceration was observed. Based on the clinical presentation, our differential diagnosis included eosinophilic cellulitis, necrobiosis lipoidica, and morphea. A punch biopsy was done to confirm the diagnosis; however, results were inconclusive. At the second clinical visit, the lesion appeared to have migrated from the left medial thigh to the left lower limb (Fig 2). A punch biopsy of the new tender area found mixed histopathologic findings consistent with the clinical course (Figs 3 and 4). The histopathologic findings were mild lymphohistiocytic infiltrate with vascular proliferation in the superficial and deep dermis. The subcutaneous tissue showed lymphohistiocytic infiltrate and slight thickening. Some of the lymphohistiocytic infiltrate extended to the periphery of the subcutaneous lobules. No neutrophilic infiltrate or well-formed granulomas were seen. Overall, the changes were that of dermal and subcutaneous septal lymphohistiocytic infiltrate with vascular proliferation. The subcutaneous changes of septal panniculitis observed were consistent with the diagnosis of ENM.

Abbreviations used:

EN: erythema nodosum
ENM: erythema nodosum migrans

DISCUSSION

ENM was first described by Bafverstedt in 1954.[1] Subsequently, Vilanova and Pinol Aguade[2] referred to the condition as *subacute nodular migratory panniculitis*, and it has now come to be known as *Vilanova disease*. The condition is characterized by deep seated nodular lesions that often enlarge via confluence or peripheral extension to form large plaques.[2] Classically, the lesions appear unilaterally as single, discrete erythematous plaques. The nodules do not ulcerate, and over weeks or months the nodules may migrate and become crescentic in nature.

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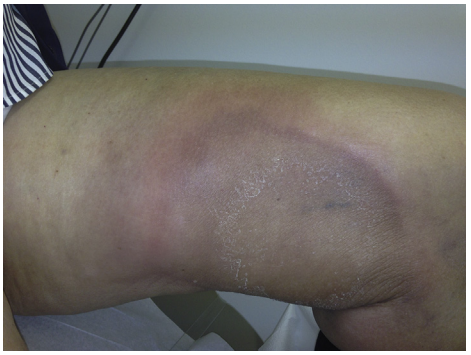

Fig 1. Large subcutaneous plaque on the left medial thigh.

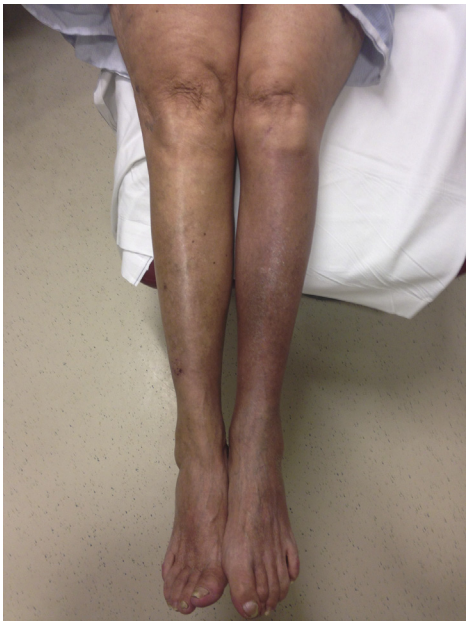

Fig 2. Large migratory plaque now observed on the left shin.

ENM is more common in women, and it is often compared with EN, which is the most common cause of panniculitis. Lesions in ENM are less tender than those in EN. The subcutaneous nodules or plaques in ENM persist for a longer period compared with EN nodules.[3] Furthermore, ENM is typically unilateral, whereas EN is typically bilateral. Most cases of ENM are idiopathic in origin; however, a significant proportion of EN cases are preceded by infection, autoimmune disease, medication adverse effect, and cancer. Although an underlying etiology is not usually identified in ENM, streptococcal infection has been implicated in some cases.[3] Generally, EN is self-limiting, whereas ENM lesions can last for months.

Histologically, ENM may appear very similar to EN. Classic EN shows septal panniculitis with superficial and deep perivascular inflammatory

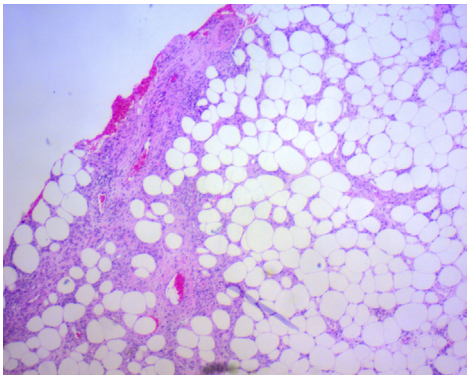

Fig 3. Low-power image shows septal involvement of the subcutaneous tissue. The dermis shows increased dense vascularity.

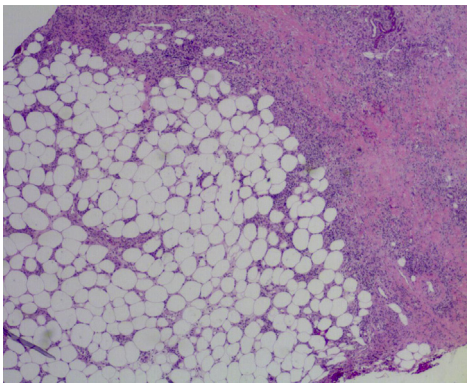

Fig 4. Inflammatory infiltrates extending into the subcutis with numerous histiocytes and lymphocytes in a septal distribution.

lymphocytic infiltrate. As the disease progresses, periseptal fibrosis, giant cells, and granulomatous tissue appears.[4,5] ENM, however, typically show greater septal thickening and prominent granulomatous inflammation along the subcutaneous septa.[1] Vascular hyperplasia may also be present in some cases.

Differential diagnosis for ENM includes EN, pancreatic panniculitis, morphea, granuloma annulare, necrobiosis lipoidica, and panniculitis caused by foreign body and lupus panniculitis. Laboratory studies helpful in diagnosis include erythrocyte sedimentation rate (ESR), antistreptolysin O titer, complete blood count, antinuclear antibody, and α-1antitrypsin levels. The ESR is commonly elevated, and antistreptolysin O is elevated in some patients. CBC helps to rule out any infection, whereas antinuclear antibody, α-1antitrypsin, serum amylase, and serum lipase can help rule out other forms of panniculitis. In our patient, the ESR was 25 mm/h, within the normal range (2–30 mm/h), whereas the neutrophil count was slightly elevated at 8.5×10^9/L

(normal, 2.0–7.5 × 10^9). All other laboratory test results were within the normal range, ruling out any form of panniculitis. The lack of clinical presentation of necrobiosis lipoidica, including telangiectasia, atrophy, and yellowish hue, along with the absence of histopathologic changes of granuloma and necrobiosis, exclude necrobiosis lipoidica from the differential diagnosis.

Treatment options for ENM patients are generally limited to potassium iodide and intralesional corticosteroids.[6,7] Indomethacin and dapsone may also be used to treat ENM; however, the evidence is very limited.[8,9] Untreated ENM can last from months to years without treatment. In the case of our patient, we prescribed topical clobetasol propionate cream (0.05% twice daily for 2 weeks) to reduce the inflammation, and her lesions have cleared up completely. It is important to note that administration of iodine-containing medications (eg, potassium iodide, amiodarone) to patients being treated for hypothyroidism, like our case, may lead to iodine excess and subsequent Wolff-Chaikoff phenomenon.[10] In such patients, treating with topical clobetasol may be a safer option.

This case shows the importance of considering ENM in the differential diagnosis of suspected morphea, necrobiosis lipoidica, and lower limb cellulitis. Certain variants of the disease may present with vascular changes in the dermis, as was seen in our case. This finding is quite atypical, and the diagnosis is made primarily based on the characteristic septal subcutaneous panniculitis. Although ENM is a variant of EN, the correct diagnosis is essential for appropriate management and patient care.

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