

Nodular cutaneous amyloidosis effectively treated with intralesional methotrexate



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INTRODUCTION

Amyloidosis is a group of conditions that involve the deposition of amyloid proteins in various tissues. It is generally classified as either systemic or cutaneous. Primary localized cutaneous nodular amyloidosis (PLCNA) is the rarest form of cutaneous amyloidosis with the amyloid depositing in the dermis, subcutis, and blood vessel walls.

Management of PLCNA is challenging, as there is no consistently effective treatment, and local recurrence is common. Management options include surgical excision, dermabrasion, electrodesiccation and curettage, cryotherapy, carbon dioxide laser, pulse dye laser, intralesional steroids, and cyclophosphamide. We report a case of PLCNA in which intralesional methotrexate has been used with good effect.

CASE REPORT

A 77-year-old man was referred to a dermatologist for management of large firm nodules and plaques on his lower face and neck. The lesions had been present for approximately 2 months before dermatology review. No treatments were trialed before dermatology referral (Fig 1). A biopsy of a lesion on the patient's chin found nodular deposits of eosinophilic hyalinized globular material for the whole thickness of the dermis, patchy plasmacytic chronic inflammatory infiltrate in adjacent stroma, and normal epidermis. Stains with Periodic acid–Schiff crystal violet and Congo red were strongly positive, and polarized light of Congo red–stained sections showed characteristic yellow-green birefringence. Histologic findings were classic

Abbreviations used:

LFT: liver function test
 PLCNA: primary localized cutaneous nodular amyloidosis



Fig 1. PLCNA pretreatment with intralesional methotrexate.

for PLCNA (Fig 2). Similar lesions were present on the patient's neck but were not biopsied. Routine blood test results showed abnormal blood protein levels, and subsequent serum electrophoresis confirmed monoclonal gammopathy of unknown significance. The potential for secondary systemic amyloidosis was then investigated with serum and urine protein electrophoresis, echocardiogram and bone marrow biopsy. In the absence of investigation findings for secondary systemic amyloidosis, the condition was classified as primary cutaneous

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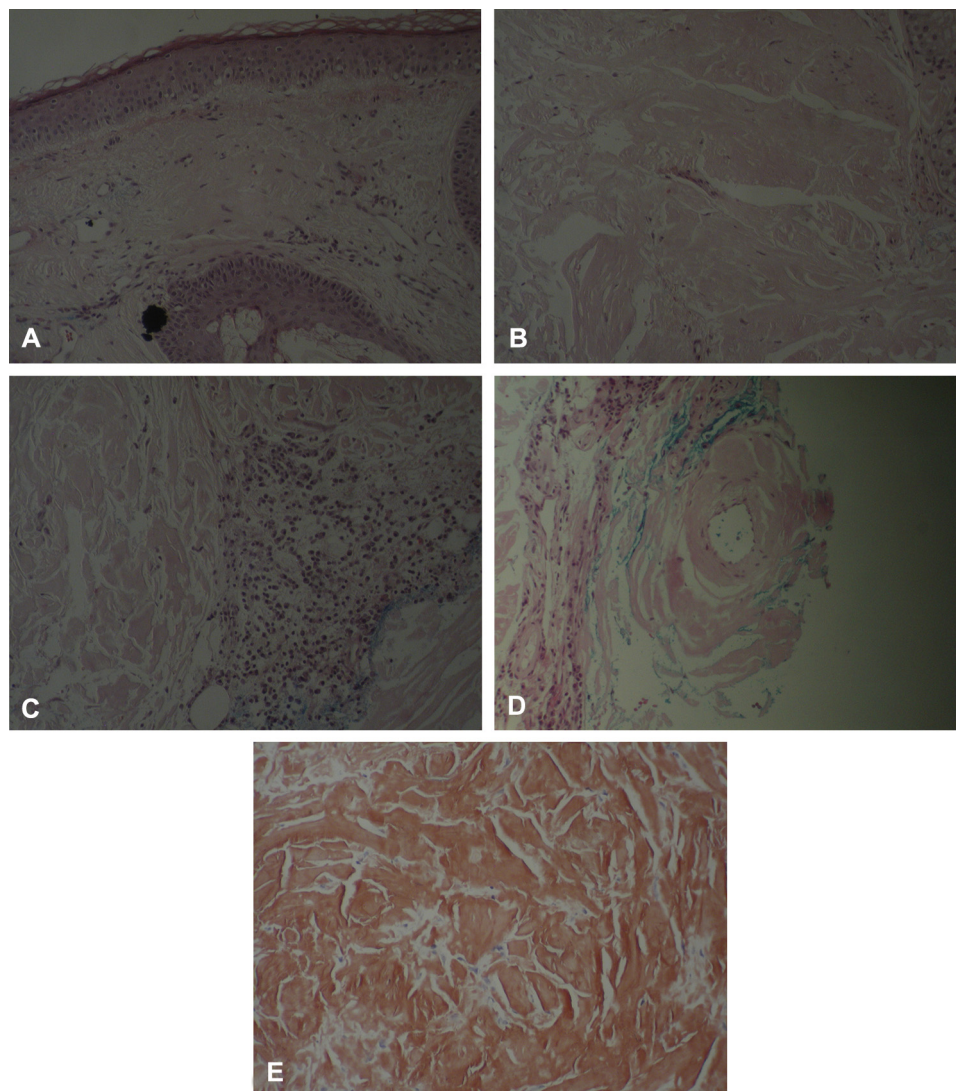


Fig 2. A-E, Histology findings.

amyloidosis in conjunction with histopathology findings.

Initially, the patient was given betamethasone dipropionate 0.1% twice daily for the nodules on the face and neck. After 4 weeks of topical corticosteroid treatment, there was no improvement in the patient's condition. Two intralesional corticosteroid injections (triamcinolone, 40 mg/mL) were administered to the nodular lesions over a 2-month period without any improvement in conjunction with topical betamethasone dipropionate 0.1% twice daily on a second weekly basis. Intralesional methotrexate injections were then started for lesions on cheeks, upper lip, and chin. Initially, only 2 nodules on the cheeks were treated, but eventually all nodules on both cheeks, upper lip, and chin region were injected. Excision or pulse dye laser therapy is being

considered for lesions on the neck, as they are less conspicuous.

The initial dose of methotrexate was 12.5 mg, which was titrated up to 25 mg then 40 mg then 50 mg. The period between treatments ranged between 2 and 4 weeks in the early stages to determine whether the patient would tolerate methotrexate. Upon ascertaining tolerance, 50-mg injections were administered on a weekly basis over a 6-month period without folic acid. This regimen resulted in a significant reduction in size of the treated lesions, and they remain stable (Fig 3). However, untreated nodules have recently started growing, and new lesions are developing in untreated areas on the back of the patient's neck and eyebrows. Weekly to fortnightly monitoring of full blood count, electrolytes, urea & creatinine, and liver

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