

# Cutaneous involvement in a case of multifocal idiopathic fibrosclerosis



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## INTRODUCTION

Multifocal idiopathic fibrosclerosis (MIF) is a rare progressive fibrosing disorder with reported manifestations in myriad tissues, most commonly the retroperitoneum.<sup>1</sup> Retroperitoneal fibrosis commonly presents with complications such as hydronephrosis or sequelae of fibrotic tissue surrounding blood or lymphatic vessels.<sup>1</sup> Other tissues affected by MIF may include the thyroid, mediastinum, and, less commonly, the gallbladder, orbit, and pancreas.<sup>2</sup> To date, only 1 case of MIF with cutaneous involvement has been reported.<sup>3</sup> We report a case of MIF with cutaneous and retroperitoneal involvement, the cutaneous manifestations of which were successfully treated with intralesional triamcinolone.

## CASE REPORT

A 64-year-old woman presented with painful, indurated, hyperpigmented plaques involving the lower legs (Fig 1, A and B). Her medical history included retroperitoneal fibrosis with biopsy-proven involvement of the para-aortic fascia. Prior treatments included methotrexate, prednisone, and hydroxychloroquine. Azathioprine had recently been added to her treatment regimen following recurrent right kidney hydronephrosis related to ureteral stricture.

An incisional biopsy from the left leg found diffuse fibrosis with multinodular and perivascular lymphoplasmacytic inflammation (Fig 2, A-C). Immunohistochemistry for IgG subclasses showed diffusely positive IgG1 staining and scant IgG4 staining. Serum IgG1 was elevated with no elevation of serum IgG4.

On follow-up, the patient presented with palpable firmness focally involving the mid back, along with worsening of the aforementioned

### Abbreviation used:

MIF: multifocal idiopathic fibrosclerosis

plaques on the lower extremities. A telescoping punch biopsy specimen was taken from the back. Histologic examination found focal deep dermal fibrosis with hyalinized collagen bundles; the lymphoplasmacytic inflammation noted in the prior biopsy was not seen. Intralesional triamcinolone (0.8 mL of 20 mg/mL) was infiltrated into several of the lower extremity plaques with improvement in both induration and pain.

## DISCUSSION

MIF is a fibrosing disorder described as part of the IgG4 spectrum of diseases, but without elevation of IgG4 in serum or IgG4-positive plasma cell infiltrate on biopsy, as previously reported by Ayli and Mutasim.<sup>3,4</sup> A suggested definition of MIF is 2 or more areas of progressive fibrosclerosis present in the same patient.<sup>3</sup> The disease's manifestations are protean and include Riedel thyroiditis, retroperitoneal fibrosis, sclerosing pancreatitis, pseudotumor of the orbit, and cholangitis.<sup>1-4</sup> To our knowledge, only 1 previous case of MIF with cutaneous involvement has been reported.<sup>3</sup> Histopathologic features of MIF include dense interstitial lymphoplasmacytic infiltration, perivascular sclerosis, fibrous tissue with scant inflammation, and vasculitis.<sup>3-5</sup> While the cutaneous biopsy findings in our case differed somewhat in that the lymphoplasmacytic infiltrate was nodular rather than interstitial, the clinical presentation, the prior para-aortic tissue biopsy showing sclerosis and lymphoplasmacytic inflammation, as well as the

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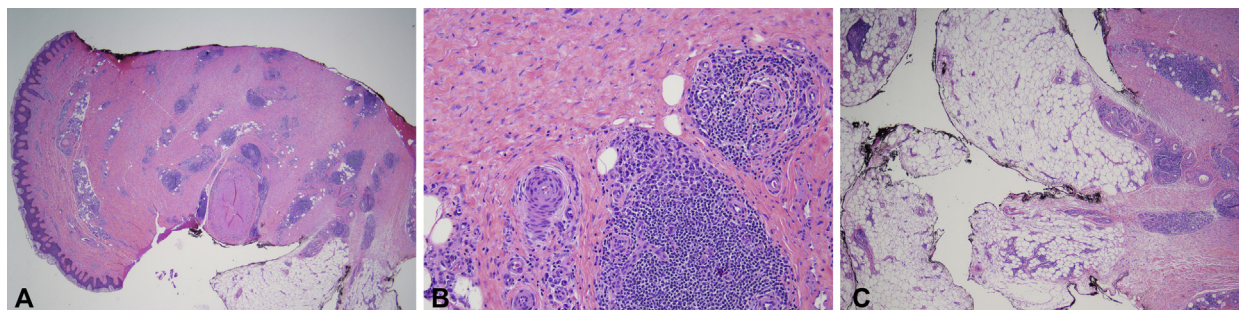
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**Fig 1.** Clinical presentation of cutaneous MIF. **A** and **B**, Indurated poorly defined hyperpigmented plaques involving the distal lower extremities were tender to palpation.



**Fig 2.** Histopathology of cutaneous MIF. **A**, Punch biopsy specimen from the left leg shows a multinodular inflammatory infiltrate extending to the deep reticular dermis with diffuse fibrosis. **B**, Higher-power magnification shows a lymphoplasmacytic infiltrate. **C**, This is also seen extending into the subcutis. (**A-C**, Hematoxylin-eosin stain; original magnifications: **A**,  $\times 20$ ; **B**,  $\times 200$ ; **C**,  $\times 40$ .)

IgG serology and exclusion of other diagnoses, were most consistent with the diagnosis of MIF.

Consistent with the previous case of cutaneous MIF described by Ayli and Mutasim,<sup>3</sup> our patient has a history of visceral fibrosis, IgG4-negative serology, and cutaneous involvement with histopathologic changes similar to those of the visceral lesions. This present case is distinguished by the documented successful treatment of cutaneous manifestations with intralesional triamcinolone. The etiology of MIF has yet to be elucidated, but reports of successful treatment with immunosuppression have raised the notion of an autoimmune pathogenesis.<sup>1</sup> There is scant evidence regarding treatment options for MIF, but there are reports of remission of visceral disease with medium-to-high dose corticosteroids, tamoxifen, and immunosuppressive medications such as cyclosporine and mycophenolate mofetil.<sup>1,3</sup> Here we propose intralesional steroid injection as a potential option for the treatment of these patients.

We report this case to increase awareness of the potential for cutaneous involvement of multifocal idiopathic fibrosclerosis and to suggest a possible therapeutic modality for the management thereof.

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