

**CASE REPORT**

# Polymyositis with antisynthetase syndrome presented with lobular panniculitis: A case report

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**Key Clinical Message**

Panniculitis is rarely seen in polymyositis. To the best of our knowledge, we report the first case of panniculitis which precedes the antisynthetase syndrome.

**KEY WORDS**

antisynthetase syndrome, autoimmune disease, Panniculitis, polymyositis

## 1 | INTRODUCTION

Panniculitis can occur in many conditions such as trauma, infection, neoplasm, enzymatic insufficiency, and connective tissue disease.<sup>1</sup> Among connective tissue diseases, panniculitis is mostly associated with lupus erythematosus over the others<sup>2</sup> and quite rare in dermatomyositis.<sup>3,4</sup> Panniculitis is even scarcer in polymyositis, with only two cases reported in PubMed Database.<sup>5,6</sup> To the best of our knowledge, panniculitis in antisynthetase syndrome has never been reported. Herein, we report the first case of a Thai male who presented with panniculitis and further investigation revealed he had polymyositis with antisynthetase syndrome.

## 2 | OUR CASE

A 68-year-old Thai man with 1 year of ankylosing spondylitis suddenly developed tender erythematous skin eruptions on both thighs for two weeks (Figure 1A). His medication was prednisolone 10 mg per day, azathioprine 75 mg per day, and methotrexate 7.5 mg per week. He was initially diagnosed and treated as cellulitis with antibiotics. Hence, immunosuppressive agents except prednisolone were stop. Unfortunately, his lesions were not improved, so the skin biopsy was done. The result showed lobular panniculitis with sparse lymphocytic and histiocytic infiltrations with foci of lipomembranous changes (Figure 2). The tissue cultures

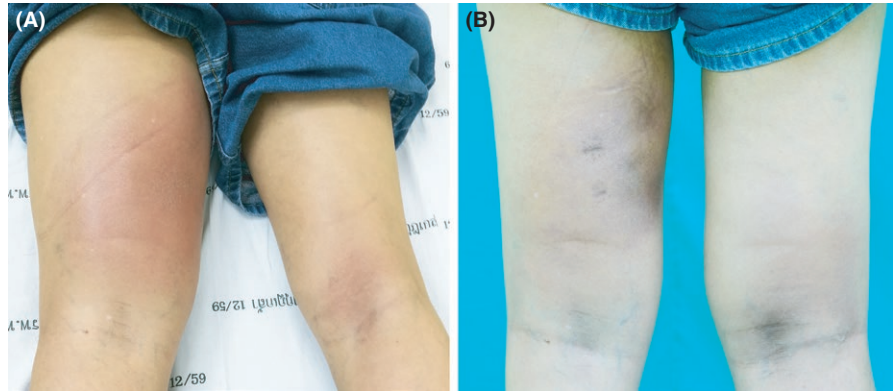
for bacteria, fungus, and mycobacterium were all negative. Laboratory investigations showed strongly positive for Anti-Jo1 and Anti-R052 but negative for ANA, anti-dsDNA, antiphospholipid antibodies, and Anti-Sc170. He had neither cutaneous signs of lupus erythematosus nor dermatomyositis. His panniculitis was improved (Figure 1B) upon increasing the dosage of prednisolone to 30 mg per day. Two months later, he developed proximal muscle weakness with high serum creatine phosphokinase. The muscle biopsy was compatible with inflammatory myopathy. The screening investigations for malignancy showed all negative results. Only the CT chest revealed interstitial lung disease. Finally, he was diagnosed with polymyositis with antisynthetase syndrome.

## 3 | DISCUSSION

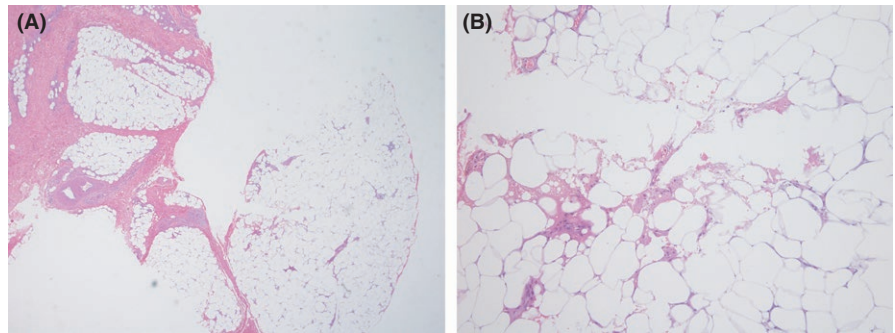
Antisynthetase syndrome is an autoimmune disease which is a separate entity from dermatomyositis or polymyositis. It is characterized by antibodies directed against an aminoacyl transfer RNA synthetase with clinical features of interstitial lung disease, myositis, Raynaud's phenomenon, and arthritis<sup>7</sup> according to the diagnostic criteria proposed by Connors et al<sup>8</sup> and Solomon et al.<sup>9</sup> To our knowledge, there is no report of any patient with polymyositis and antisynthetase syndrome presenting with panniculitis.

Regarding the differential diagnosis of panniculitis, especially in immunocompromised host, the infectious etiologies

**FIGURE 1** A, Tender ill-defined erythematous indurated subcutaneous plaques on posterior left thigh and right popliteal fossa; B, After treatment, the lesions improved and became hyperpigmented 1859 × 821 mm (72 × 72 DPI)



**FIGURE 2** The histopathology from left thigh showed (A) lobular panniculitis with sparse cells infiltration (Hematoxylin-eosin staining, ×4) (B) lymphocytic and histiocytic infiltrations with some foci of lipomembranous change (Hematoxylin-eosin staining, ×40) 541 × 203 mm (150 × 150 DPI)



**TABLE 1** Comparative features between lupus panniculitis and dermatomyositis panniculitis

	Lupus panniculitis	Dermatomyositis/Polymyositis
Presentation	Tender subcutaneous nodules and plaques; may have ulceration and lipoatrophy. <sup>14</sup>	Painful, erythematous, indurated, nodules and plaques; may have ulceration and lipoatrophy. <sup>15</sup>
Distribution	Face (esp. cheeks), upper arms, scalp, shoulders, hips, buttocks, breasts, trunk	Lower and upper extremities, usually spare face. <sup>16</sup>
Associated features	Discoid rash or other lupus specific rashes  Systemic features of lupus, such as arthralgia or Raynaud phenomenon. <sup>14,17</sup>	Typical cutaneous findings including heliotrope rash, Gottron's papules in dermatomyositis  Symmetrical proximal muscle weakness. <sup>16</sup>
Investigations	ANA, anti-dsDNA, anti-RNP, leukopenia, hypocomplementemia and elevate ESR. <sup>14,17</sup>	Elevate serum CPK, Anti-Jo1 Electromyography (EMG) and muscle biopsy show evidence of myositis. <sup>6</sup>
Histopathology	Lobular or mixed panniculitis composed of lymphocytes and plasma cells  Mucin deposition Lipomembranous change Nuclear dust and lymphoid aggregations in some cases. <sup>14</sup>	Lobular or mixed panniculitis composed of lymphocytes and plasma cells Fat necrosis Lipomembranous changes Some lymphoid aggregations May have calcification. <sup>16</sup>

should be ruled out first. In our case, all cultures were negative and histopathology excluded infections, instead suggesting connective tissue diseases that is lupus panniculitis and dermatomyositis. However, due to the similarities of clinical presentation and histopathology of both diseases, associated

features and further investigations were required (Table 1). There were no specific rashes in this patient. However, the presence of myopathy, arthralgia, and interstitial lung disease favored the diagnosis of polymyositis and antisynthetase syndrome.

Panniculitis in dermatomyositis and polymyositis is a rare finding and may precede, coexist, or appear after the onset of disease.<sup>4,10</sup> There were only two case-reports of panniculitis in polymyositis. In the first case,<sup>6</sup> a 58-year-old woman presented with panniculitis on her thigh, abdomen, and buttocks that preceded the development of polymyositis. The histopathological features showed lobular panniculus infiltrations with lymphocytes and plasma cells. This case was similar to our patient, which responded well to high dose prednisolone and had no evidence of malignancy. The second report<sup>5</sup> was a 35-year-old woman who developed cutaneous ulcer and multiple panniculitic nodules on her abdomen and breasts after the onset of polymyositis. This case developed panniculitis while she was treated with immunosuppressive agents as in our patient.

At present, the treatment guideline of panniculitis in dermatomyositis and polymyositis has not been established yet. Most of the cases are treated with prednisolone with variable outcome. Other treatments are intravenous methyl prednisolone, azathioprine, methotrexate, intravenous immunoglobulin, hydroxychloroquine, or cyclosporine.<sup>3,10,11</sup> It has been suggested that panniculitis may be a positive prognostic sign with a good response to treatment and rare underlying neoplasm.<sup>2,4,12,13</sup>

In conclusion, we report the first case of polymyositis with antisynthetase syndrome, which has panniculitis preceding the onset of proximal muscle weakness. Although panniculitis is a very rare clinical finding in polymyositis and antisynthetase syndrome, this condition should be included in the differential diagnosis of patients presenting with idiopathic panniculitis.<sup>10</sup>

## AUTHOR CONTRIBUTION

AR: conceived and designed the report, collected the data, analyzed the data, wrote and reviewed the manuscript. NS: conceived and designed the report, analyzed the data, wrote, reviewed the manuscript and approved the final draft. Both authors were involved in patient care.

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