Idiopathic palmar fasciitis and polyarthrititis syndrome

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DESCRIPTION

A 79-year-old man presented to our hospital with 2-month history of difficulty in flexing and extending all fingers of both hands. Three years previously, he was diagnosed as having monoclonal gammopathy of undetermined significance (MGUS) and was followed-up without progression to multiple myeloma. On physical examination, the patient had thick and tight flexion contractures of both hands (woody hands) with mild oedema (figure 1A,B). The remainder of the physical examination was unremarkable. He had no history of Raynaud's phenomenon. C reactive protein was 0.07 mg/dL. Serum Ig levels were as follows: IgG 2087 mg/dL, IgA 264 mg/dL and IgM 32 mg/dL. Immunoelectrophoresis revealed IgG- κ type M-protein. Serum-free light chain analysis showed a mild raised κ/λ ratio of 1.76 (normal range: 0.26–1.65). A bone marrow aspiration revealed 1.6% of plasma cells with atypia such as flame cells, grape cells and giant plasma cells. These findings are compatible with the diagnosis of MGUS. We suspected the likelihood of amyloidosis. However, biopsy of the duodenum was negative for Congo red stain. A test for antinuclear antibodies was positive at a titer of 1:40, with a speckled pattern. However, test for Scl-70 antibody was negative and Raynaud's phenomenon was absent. Therefore, the patient did not meet the criteria for the diagnosis of scleroderma. At this point, we suspected the likelihood of palmar fasciitis and polyarthrititis syndrome (PPFAS), and a deep skin biopsy from the dorsum of the right hand was done, which revealed severe thickness of the fascia (figure 1C double-headed

arrow) with increase of fibroblast-like spindle cells and collagen fibres (figure 1D), and scarce infiltration of inflammatory cells. A stain for Congo red stain was negative. Thus, a diagnosis of PPFAS was made. PPFAS is regarded as paraneoplastic syndrome, characterised by flexion contractures of both hands and thickening of palmar fascia.¹ Therefore, we performed whole body screening for malignancies such as whole body CT, upper and lower gastrointestinal endoscopy and measurement of tumour markers. However, thorough examinations of the entire body could not identify any evidence of malignancy. Thus, PPFAS was currently considered idiopathic. Until now, only two patients with idiopathic PPFAS have been reported.^{2,3} However, PPFAS can occur months to years before the detection of malignancy and appears unrelated to the tumour stage. Therefore, he is currently under close follow-up for development of malignancy in the future.

Learning points

- ▶ Palmar fasciitis and polyarthrititis syndrome (PPFAS) is an uncommon disorder characterised by progressive flexion contractures of both hands and regarded as paraneoplastic syndrome.
- ▶ Our image captures the macroscopic characteristics of PPFAS, that is, tight flexion contractures of both hands, also known as 'woody hands'.
- ▶ We believe that this report will increase awareness about PPFAS among physicians.

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Figure 1 The patient had thick and tight flexion contractures of both hands (woody hands) with mild oedema (A,B). Skin biopsy from the dorsum of the right hand revealed severe thickness of the fascia ((C) double-headed arrow) with increase of fibroblast-like spindle cells and collagen fibres (D).

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