Patellar tendon ectopic calcification in a patient with undiagnosed limited cutaneous systemic sclerosis

Sirs,

A 62-year-old woman with a 30-year history of Raynaud's phenomenon and a 2-year history of epigastralgia presented with swelling of the left knee. Physical examination revealed facial telangiectasia, sclerodactyly of both hands, and subcutaneous induration of the right knee (Fig. 1A, arrow). Antinuclear antibody was positive (1240×; reference <40×), and anti-centromere antibody level was >240 U/mL (reference, <7.0 U/mL). A right knee x-ray revealed ectopic calcification of the patellar tendon (Fig. 1B, arrows). She was subsequently diagnosed with limited cutaneous systemic sclerosis. After treatment with beraprost sodium (60 μg/day), Raynaud's phenomenon improved. Ectopic calcification was present; however, no subsequent deterioration was observed during the 1-year follow-up period in the outpatient clinic.

Systemic sclerosis (SSc) is a complex disorder characterised by the involvement of small arteries, microvessels, and connective tissue, with deposition of fibrotic tissue and microvascular obliteration in the skin and internal organs (1). Depending on the extent of skin involvement, SSc is classified into limited cutaneous SSc and diffuse cutaneous SSc (1). Given the heterogeneity of clinical symptoms and signs, the American College of Rheumatology/the European League against Rheumatism developed new classification criteria in 2013 (2). In patients with SSc, while Raynaud's phenomenon is the initial finding, calcinosis is common and refers to the sub-epidermal deposition of calcium salts in the skin. Calcinosis cutis is the deposition of insoluble calcium in the skin and subcutaneous tissues and is a manifestation of several autoimmune connective tissue diseases, most frequently with systemic sclerosis (3). Autoimmune connective tissue disease-associated calcinosis is of the dystrophic subtype, which occurs at sites of damaged tissue in the setting of normal serum calcium and phosphate levels (3). It has been reported that in SSc, calcinosis is associated with long disease duration and sclerosis autoantibodies and that vascular hypoxia could play a more important role in SSc-associated calcinosis (3). Limited cutaneous systemic sclerosis primarily involves the fingers (sclerodactyly), distal knee, and face, where ectopic calcification may occur (4). Since SSc-related calcinosis is associated with significant morbidity, including through cutaneous ulceration and predisposition to become infected, and the only treatment for skin calcification that progresses and interferes with daily life is surgical excision, it is vital to prevent scleroderma progression through early diagnosis and early intervention. SSc-related calcinosis is an area of significant unmet clinical need and has been a neglected area of research for too long. Safe, effective treatments are badly needed to improve patient quality of life and outcomes. To facilitate future clinical trials, we require an increased understanding of pathogenesis to inform the selection of potential targeted therapies and reliable outcome measures, including those which will measure the impact and severity of calcinosis from the patient perspective.

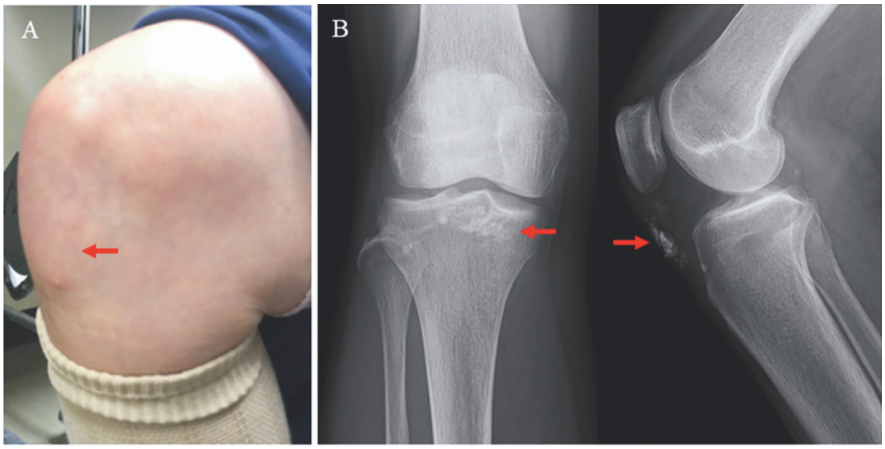

Fig 1. Macroscopic and radiologic findings of the right knee.
A: Subcutaneous induration of the right knee (arrow).
B: Radiography of the right knee showing ectopic calcification of the patellar tendon (arrows).

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