Knee contractures as the presenting manifestation of scleroderma

Gwen Kane-Wanger, Barbara E Ostrov, Bruce Freundlich

Abstract

Musculoskeletal disorders may be the primary manifestations of scleroderma. The case is reported of a patient admitted for an evaluation of severe knee flexion contractures who was diagnosed as having scleroderma but lacked the typical presenting features of the disease. Radiographic, serological, and histological support for this diagnosis are presented. Scleroderma should be considered in the diagnostic evaluation of joint contractures.

Musculoskeletal disorders are often the primary manifestations of scleroderma.¹⁻³ Patients may present with stiffness and polyarthralgias which can progress to limitation of movement. Flexion contractures of the small joints of the hands and elbows commonly occur, whereas contractures of the knees are far less common. We report the case of a patient diagnosed with scleroderma during the evaluation of severe bilateral knee contractures.

Case history

The patient was a 75 year old white woman who presented in December 1988 for evaluation and treatment of severe bilateral knee contractures. Fifteen years earlier bilateral leg lymphoedema was noted with no apparent aetiology. Eight years before admission the patient was diagnosed with and treated for hypothyroidism. Bilateral

leg cramping and knee stiffness were first noted two years before admission and progressive flexion contractures of the knees developed within one year. The patient eventually became confined to a wheelchair. In March 1988 physical therapy and traction resulted in a short lived improvement in knee motion. During the admission in December 1988 the patient had complained of intense muscle cramping and stiffness in the knees and legs. No other joint symptoms were noted and Raynaud's phenomenon, muscle pain elsewhere, or muscle weakness were not present. No dysphagia, symptoms of sicca syndrome, fevers, or weight change were noted.

On physical examination, the patient was an obese woman with normal vital signs. Examination of her face showed areas of equivocal induration without decreased mouth aperture or perioral wrinkling. No telangiectasies were noted. Thyromegaly was not detected. Examination of the hands did not show skin oedema or tightening. The hips had 15° flexion contractures bilaterally. The knees had 80° flexion contractures bilaterally and could be actively flexed to only 130° without pain. Marked non-pitting oedema was noted over the legs up to the knees. The skin overlying the lower legs was also hyperpigmented and thickened, but without nodularity. No tenderness, warmth, or obvious joint effusions were found. The neurological examination did not show muscle weakness or

Department of
Internal Medicine,
Beth Israel Hospital,
Boston, MA, USA
G Kane-Wanger

Department of Pediatrics
and Medicine,
Penn State University,
Milton S. Hershey
Medical Center,
Hershey, PA, USA
B E Ostrov

Rheumatology Section,
Department of
Internal Medicine,
Hospital of the
University of
Pennsylvania,
Philadelphia, PA, USA
B Freundlich

Correspondence to:
Bruce Freundlich,
Rheumatology Section,
Department of
Internal Medicine,
Hospital of the
University of Pennsylvania,
570 Maloney Building,
3400 Spruce Street,
Philadelphia, PA 19104, USA.

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Figure 1 Radiographs of the right knee showing subcutaneous calcinosis. (A) Anteroposterior and (B) lateral view.

Figure 2 Histological section of skin from the popliteal fossa showing marked dermal fibrosis with foci of lymphocytic infiltration.

fasciculations. The general examination was otherwise negative.

Laboratory studies showed an antinuclear antibody titre of 1/320 in a speckled pattern, centromere negative; antibodies to Scl-70, SS-A, and SS-B were not detected. Thyroid function tests showed a thyroxine concentration of 55.3 nmol/l (normal 64.4–128.7 nmol/l), and thyroid stimulating hormone of 57 arb. unit (normal <10 arb. unit).

Antibodies to thyroglobulin and microsomes were present. Creatine kinase was slightly increased at 190 U/ml (normal 30–150 U/ml). Other laboratory studies were negative. Radiographs showed mild degenerative arthritis of the knees with soft tissue calcifications overlying the right patella (fig 1). Hand films showed osteophytes but no calcinosis. The dose of thyroxine was increased and creatine kinase levels normalised. Three days after admission bilateral tendon releases were performed and an intraoperative skin biopsy sample was taken from behind the knee in an area that was not clinically oedematous. The biopsy specimen (fig 2) showed marked dermal fibrosis with foci of lymphocytic infiltration diagnostic of scleroderma. At the time of discharge, only 5° flexion contractures of the knees remained and repeat thyroid function tests were normal. At follow up three months after discharge, the patient continued to have leg cramping and stiffness, but she refused any further evaluation to determine the extent of scleroderma.

Discussion

Rheumatic symptoms are often the earliest manifestations of scleroderma. Rodnan and Medsger found that 41% of their patients with scleroderma initially had arthralgias or polyarthritis, typically in the hands, knees, and feet.^{2,3} As the disease progresses, increasing stiffness and limitation of movement due to cutaneous thickening is common, particularly in the hands and arms.² Decreased knee motion

is less common and usually occurs later in the course of severe skin disease.² Flexion contractures of the knees have occasionally been mentioned in reports of the articular manifestations of scleroderma, but little detail on the severity of these deformities has been reported.^{4,5} The patients described had classic scleroderma, all with finger contractures and limitation of motion of the knees with occasional knee effusions.⁵ Pathologically, cutaneous thickening probably accounts for most flexion contractures in scleroderma, though the tendons and synovium may also be affected.^{4,6} Specimens of the synovium and tendons were not obtained from this patient.

This patient is unique in that she not only had isolated leg flexion contractures that were particularly severe in the knees, but that these deformities were also the presenting manifestation of scleroderma. There were none of the more typical presenting features of scleroderma, such as Raynaud's phenomenon, skin thickening, dysphagia, or hand involvement. Subcutaneous calcinosis detected by radiographs, a speckled antinuclear antibody pattern, and the characteristic skin biopsy sample established the diagnosis, however. There is no clear explanation for this unusual presentation of scleroderma.

This patient had coexisting hypothyroidism which appears to be increased in occurrence in scleroderma.⁷ Myalgias and cramping, and an increase in creatine kinase levels may occur in either disease.⁸ In this patient, the decrease in creatine kinase levels to normal after an increase in thyroid replacement drugs, so this was probably due to hypothyroidism⁹ rather than scleroderma myopathy. Scleroderma myopathy may however, have caused the patient's persistent muscle symptoms.

Conclusions

This patient illustrates that flexion contractures of the knees may be a presenting feature of scleroderma. Scleroderma should therefore be considered in the evaluation of joint contractures, even in the absence of other typical features of the disease.

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