

Table 1 Patients' data

Patients	Age	Pregnancy		Miscarriage*	Age last pregnancy	Transfusions*	SS*	M*
		Male	Female					
1	59	1	1	No	28	No	Yes	No
2	76	1	1	Yes	33	No	Yes	No
3	49	1	0	Yes	22	No	Yes	No
4	53	1	0	Yes	32	No	Yes	No
5	71	3	0	No	29	No	Yes	No
6	58	2	0	Yes	28	Yes	Yes	No
7	54	0	2	No	23	No	Yes	No
8	58	0	1	Yes	35	Yes	Yes	No
9	64	0	1	No	39	No	Yes	No
10	33	0	0	No	-	No	Yes	No
11	66	0	1	No	38	Yes	No	No
12	52	1	0	No	28	No	No	No

*Miscarriage = considered as a possible source of microchimerism; transfusion = considered as a possible source of microchimerism; SS = Sjögren's syndrome; M = microchimerism.

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showed finger clawing, sclerodactyly, and scleroderma affecting her arms to the mid-upper arm, the face, neck, upper chest, thighs, and calves. Pitting oedema, noted in the forearms, upper arms, chest, and thighs, had the following characteristics: it occurred in areas of affected skin—typically it occurred in the advancing front of skin involvement, was slow to induce (of bees' wax consistency), and it affected non-dependent areas. Her skin was also erythematous in parts. Livedoid patterning over the knees was also noted. The rest of the clinical examination, including musculoskeletal, cardiac, and respiratory systems, was unremarkable. A skin biopsy specimen from the upper right forearm and dorsum of the left fifth proximal phalanx showed dermal sclerosis and perivascular lymphocytic inflammation around superficial dermal vessels, consistent with scleroderma. Both skin biopsy sites healed with keloid scarring. Her antinuclear antibody titre was 1/640, speckled pattern, and extractable nuclear antigens were negative for RNP, antitopoisomerase, anticentromere, SSA, SSB, and Sm antibodies.

A simple bedside test (the skin pitting oedema time test or "SPOT" test) was devised to quantify the duration of skin pitting and see whether its measurement paralleled response to treatment. A small diameter coin was placed

over an area of oedematous skin on the arm, the site being recorded for future reference. A sphygmomanometer cuff was placed over the coin and around the arm, inflated to 100 mm Hg for 60 seconds, and then released. The sphygmomanometer cuff and coin were removed, leaving the coin's impression in the skin. From this time (time 0) the impression was palpated every minute by two independent observers (patient and author) until it was no longer palpable. This time was noted and recorded as the skin oedema time. Both interobserver and intraobserver variation were assessed over three consecutive days (coefficient of variation 6.8% and 6.8% respectively). The patient's skin oedema time was compared with that from a similar area of skin on a control matched for age, sex, and menopausal status (24 minutes).

At the patient's request, treatment with clindamycin over two consecutive days was started on 3 May 1999, at which time the skin oedema time measured 40 minutes. Clindamycin was stopped owing to marked diarrhoea and abdominal tenderness, which settled over five days. The patient was readmitted for pulse methylprednisolone on 13 May, by which time her skin had become increasingly sensitive and pruritic.

Figure 1 outlines her treatment with pulse steroids and cyclophosphamide. Cyclosporin

Pitting oedema in early diffuse systemic scleroderma

The term "puffy skin" is not infrequently noted at initial presentation of patients with systemic scleroderma. However, this is generally described as "non-pitting" oedema. The following case history challenges the latter widely held assumption, showing that its measurement is simple and may offer a more sensitive method of assessing response to treatment than the modified Rodnan skin score.¹

CASE REPORT

A premenopausal computer analyst first presented in May 1998 with a two month history of finger stiffness. The next 10 months were spent investigating and treating the cause of her iron deficiency anaemia—gastric ectasia (watermelon stomach). The diagnosis of diffuse systemic scleroderma was made in March 1999. At this time she presented with persisting symptoms of skin stiffness, burning and pruritis, especially in the early morning, affecting the skin of her arms and legs, face, and upper chest. Clinical examination

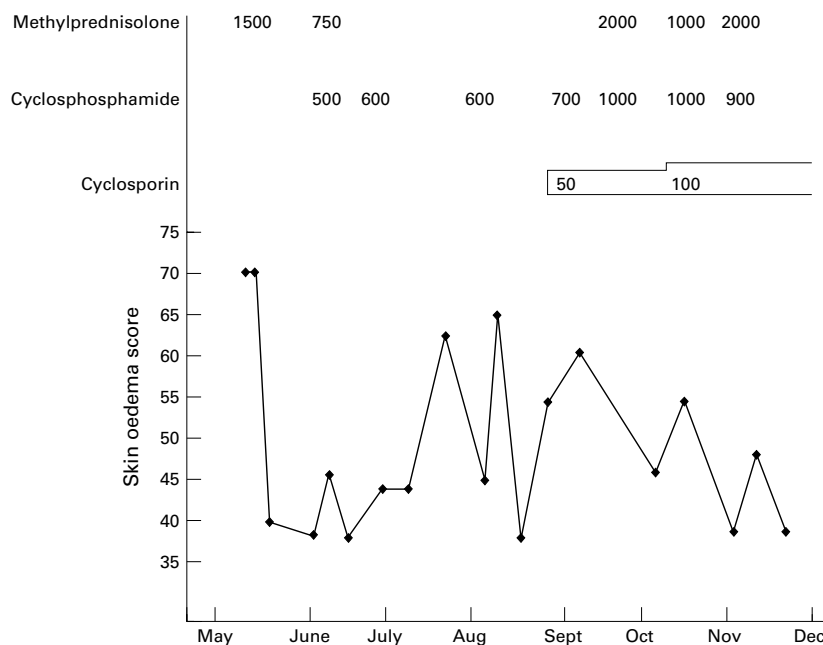


Figure 1 Patient's treatment with pulse steroids and cyclophosphamide.

was added because of continuing disease activity. Before her first course of intravenous cyclophosphamide bilateral basal crepitations were noted. By 30 June 1999 skin hyperpigmentation and the development of mild dysphagia was noted. Concurrently, she also noted increased hair regrowth on the dorsum of her hands and lateral aspect of the calves. After her final course of cyclophosphamide her hands were no longer clawed into fixed flexion, the skin on the dorsum of the hands was again so supple that the dorsal hand veins were both visible, palpable and protruding from the skin surface; the forearm skin biopsy site was no longer keloid, though that on her finger was still slightly raised and palpable; her skin was much less irritable or pruritic. The most marked areas of skin improvement were those which were still oedematous at the start of treatment and where the skin was affected for the least time—upper arms, forearms, hand, thighs, and anterior chest. The area of skin least responsive to treatment was over the distal phalanges, which remained tethered and immobile.

DISCUSSION

This case is presented to demonstrate a number of features. The skin of early diffuse disease is often characterised by pitting oedema. The skin pitting oedema time is easily measured and its measurement is reproducible. Measured as a multigrade quantitative scale (units of one minute), it is more sensitive to change than the current four grade semiquantitative modified Rodnan skin score¹. It is useful in determining responsiveness of the skin oedema to various disease modifying therapeutic regimens. Finally, finger clawing seen soon after scleroderma onset, if largely due to skin oedema, may be reversible using immunosuppressive treatment.

There are two potential limitations of the "skin pitting oedema time" test. The first relates to its duration, making it impractical to administer in an outpatient department. The second relates to the potential influence of already fibrotic skin on the skin oedema score measurement. The first limitation may be overcome by applying a less compressive force (for example, 50 mm Hg) for a shorter duration (for example, 30 seconds)—both of which independently appeared approximately to halve the skin oedema recovery time, having the patient and/or a relative complete the assessment before leaving the surgery, or devising a mechanical device to monitor skin oedema quantitatively. The role of excess collagen deposition in affecting skin oedema recovery time is uncertain.

The occurrence of pitting skin oedema in early scleroderma is a useful clinical observation, providing information about the underlying pathology and hence the speed of responsiveness of the disease to treatment. It also provides guidelines for the relative usefulness of anti-inflammatory/immunosuppressive drugs rather than the historical antifibrotic "gold standard", D-penicillamine.

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Parvovirus arthropathy masquerading as the arthritis of Behçet's disease

Parvovirus B19 is common, with 50-75% of UK adults having evidence of previous infection. Complications, such as arthritis, occur in a small minority. We report a case of parvovirus associated arthritis causing diagnostic confusion in a patient known to have Behçet's disease.

CASE REPORT

A 34 year old white woman presented with a small joint polyarthritides, low back discomfort, severe heel pain, and patchy sensory disturbance, after a febrile illness with a rash. She had had Behçet's disease since the age of 3 years, manifest by arthralgia, mouth, nasal and genital ulcers, conjunctivitis, facial swelling, and livedo reticularis. Before her presentation, her Behçet's disease had been relatively well controlled by 100 mg of azathioprine a day, the only residual symptoms being pain and morning stiffness of 45 minutes affecting her fingers, wrists, elbows, and shoulders, and livedo reticularis.

On presentation she was afebrile, with cervical lymphadenopathy. The liver and spleen tip were palpable. She had mild metacarpophalangeal and wrist synovitis, bilateral knee effusions, and was exquisitely tender bilaterally over the plantar fascia. Her sacroiliac joints were tender. There was subjective symmetrical sensory loss to pin prick and light touch on the dorsum of the feet and the lateral aspect of the hands. Neurological examination was otherwise unremarkable. An x ray examination of the hands, calcaneum, and sacroiliac joints was normal, as was the full blood count, and C reactive protein. Rheumatoid factor, antinuclear antibodies, antineutrophil cytoplasmic antibodies, and anticardiolipin antibodies were not detected. She was treated initially with ibuprofen and colchicine, without improvement. The addition of methylprednisolone acetate 120 mg intramuscularly and local steroid injections to the plantar fascia (12.5 mg hydrocortisone acetate) brought about rapid improvement. All her symptoms resolved over the following few weeks. Parvovirus B19 IgM was subsequently found to be strongly positive by both enzyme linked immunosorbent assay (ELISA) and radioimmunoassay, indicative of recent infection.

DISCUSSION

Sacroiliitis is an unusual feature of Behçet's disease,^{1,2} and plantar fasciitis has not been previously described in this disorder. Parvovirus arthritis is an uncommon sequela of a common viral exanthem. The usual presentation is of an acute onset polyarthritides, resembling rheumatoid disease, which may follow a flu-like illness and rash.^{3,4} Transient neurological disease may occur.⁵ Parvovirus associated relapse of spondylitis after a symptom-free period of 15 years has been described.⁶

The diagnosis in our case was complicated by superficial similarities to the arthritis of Behçet's disease. The possibility of neurological involvement was a new, and worrying, feature. However, there were no additional features of active Behçet's disease, such as mouth or genital ulcers, which had been associated with previous exacerbations. The unusual features of seronegative arthritis and prior febrile illness raised the possibility of a coincidental reactive arthritis. Despite the presence of some atypical features, we consider parvovirus B19 to be the most likely cause of this patient's illness, on both clinical and serological grounds.

This case illustrates the importance of considering the diagnosis of coincidental parvovirus in patients with pre-existing rheumatic diseases where there are unusual features. Because parvovirus associated arthritis is usually self limiting, this diagnosis may allow a more conservative approach to treatment.

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