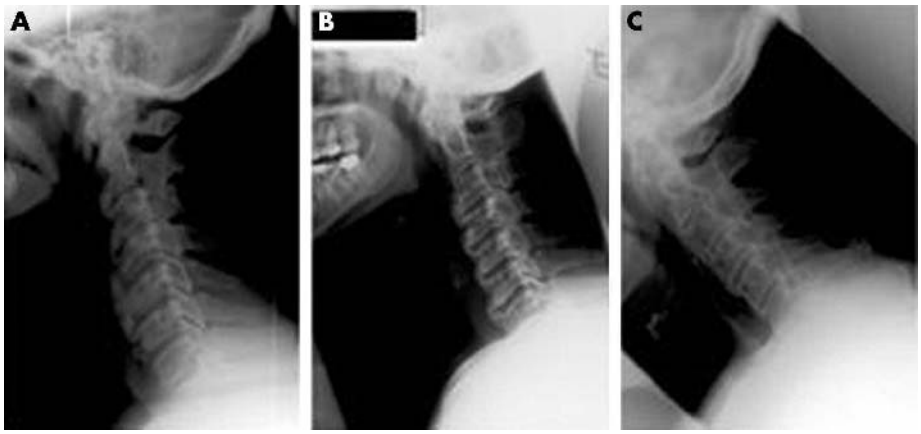

Figure 1 Lateral *x* ray findings of the cervical spine of the son (A), the daughter (B), and the father (C), showing severe anterior osteophytosis.

HLA link, as found between HLA-B27 and the spondyloarthropathies.

Although our patients were diagnosed as DISH, there are some atypical features. The most striking of these is severe cervical disease without extensive dorsal involvement. Forestier and Rotes-Querol in their classification criteria, considered involvement of at least three intervertebral bodies in the dorsal spine to be essential.[7] However, Utsinger's criteria do not include this as a necessary feature.[8] One similar case has been described: a 71 year old patient presented with similar cervical findings and sparing of the dorsal spine and sacroiliac joints. Difficulty was found classifying the condition as either DISH or ankylosing spondylitis.[9] Another unusual feature in our cases is the strong familial pattern. From the tissue typing results of our two sibling patients, it appears less likely that, if there is a hereditary component, it is linked to HLA status.

It thus remains to be seen whether this is a new disease entity or an unusual familial variant of DISH.

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Antiphospholipid antibodies in patients with scleroderma: prevalence and clinical significance

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Antiphospholipid antibodies (aPL) are detected in a variety of autoimmune disorders, most commonly systemic lupus erythematosus, but also in some infectious diseases, lymphoproliferative disorders, and even in apparently healthy people.

Although a wide prevalence of aPL in systemic sclerosis has been reported (between 0 and 41%), most studies have focused on anticardiolipin antibodies (aCL) and very little is known about other aPL in this disease. We determined the prevalence and clinical significance of aCL, antibodies to β_2-glycoprotein I (anti-β_2GPI), and antibodies to phosphatidylserine-prothrombin complex (aPS-PT) in 25 patients with scleroderma (18 with limited and 7 with diffuse scleroderma, as defined by LeRoy *et al*[1]) (table 1). Twenty four patients were female (median age 50 years (range 28–70), median disease duration 3 years (range 1–20)). One patient had a history of venous thrombosis. Of the 17 patients who had ever been pregnant, five had an adverse obstetric history. Two patients had miscarriages (before the 10th week of gestation), two patients had a fetal death (at the 10th week of

Table 1 Clinical characteristics of patients with scleroderma*

Characteristics	Limited (n = 18)	Diffuse (n = 7)	All (n = 25)
Calcinosis	2	1	3
Raynaud's phenomenon	17	6	23
Oesophageal dysmotility	9	4	13
Sclerodactyly	17	5	22
Telangiectasia	6	2	8
Pulmonary manifestations	11	5	16
Interstitial lung disease	10	5	15
Pulmonary hypertension	3	0	3
Cardiac manifestations	1	0	1
Renal manifestations	0	0	0
Bowel disease	0	0	0

*All patients were attending the 2nd Chair of Rheumatology, Rheumatology Division and Centre for Systemic Rheumatic Diseases, Cagliari University Hospital, Cagliari, Italy.

gestation or later), and one patient had a premature birth (before the 34th week of gestation) due to severe pre-eclampsia. Platelet count was normal in all patients. Only one patient had a prolonged activated partial thromboplastin time. One hundred healthy donors with no relevant medical history comprised the control group.

aCL, anti-β_2GPI, and aPS-PT were detected by enzyme linked immunosorbent assay (ELISA).[2–4]

aPL were present in 8/25 patients. Table 2 shows the distribution of aPL in patients and controls. aCL for IgG/IgM and aCL IgG were more frequently found in patients with scleroderma than in controls (24% *v* 5%, odds ratio = 6 (1.7–21.7), p = 0.008 and 16% *v* 3%, odds ratio = 6.1 (1.2–2.9), p = 0.03, respectively). The prevalence of anti-β_2GPI did not differ between patients and controls (8% *v* 3% for IgG/IgM, 4% *v* 2% for IgG, and 4% *v* 1% for IgM).

Interestingly, patients with telangiectasia and pulmonary hypertension had IgM aPS-PT more frequently than those without (37.5% *v* 0%, relative risk = 4.4 (2.0–9.5), p = 0.02 and 66.6% *v* 4.5%, relative risk = 14.6 (1.8–116.9), p = 0.03, respectively). No associations were found between the other aPL analysed and clinical manifestations of scleroderma.

One patient with scleroderma who had had venous thrombosis also had IgG aCL at low titres. Of the two patients with a history of miscarriages (<10th week of gestation), one had IgG anti-β_2GPI and the other IgM aPS-PT. None of the patients who had fetal death (n = 2) or prematurity (n = 1) had aPL.

Although the presence of all aPL was more common in patients with scleroderma than in healthy controls (32% *v* 5%), the clinical manifestations of antiphospholipid syndrome were not frequently seen in these patients.

The prevalence of aCL in scleroderma has been reported to range from 0%[5] to 41%.[6] In this study, only one patient had a history of venous thrombosis and aCL at low titres, suggesting that this manifestation may have been aCL related.

Parodi *et al* described anti-β_2GPI in 3/90 (3.3%) patients with scleroderma,[7] whereas Schoenroth *et al* reported a prevalence of 8% when studying 26 patients with this disease.[8] These studies are in agreement with our findings.

Although the prevalence of aPS-PT was low in our study, these antibodies were more frequently found in patients with

Table 2 Distribution of aPL in patients with scleroderma and controls

	Scleroderma (n = 25)	Limited (n = 18)	Diffuse (n = 7)	Controls (n = 100)
Any aPL	8	6	2	5
aCL	6	5	1	5
IgG	4	3	1	3
IgM	2	2	0	2
Anti-β_2GPI	2	2	0	3
IgG	1	1	0	2
IgM	1	1	0	1
aPS-PT	3	2	1	0
IgG	0	0	0	0
IgM	3	2	1	0

telangiectasia and pulmonary hypertension, supporting the data from Hasegawa *et al* in their cohort of 112 patients with scleroderma.[9] Overall, these findings suggest that aPS-PT may be a marker of vascular involvement in patients with scleroderma. However, as this is a very small study, further research is warranted to confirm or reject this hypothesis.

In summary, aPL are commonly found in patients with scleroderma but the ''typical'' clinical manifestations of antiphospholipid syndrome are not frequently seen in these patients.

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