Cardiac Abnormalities in a Patient with Localized Scleroderma

SIR—Localized scleroderma is a benign disease characterized by spontaneous resolution of skin lesions over several years. However, various mild and self-limited systemic features may co-exist, including arthritis, myopathic changes, abnormal oesophageal peristalsis and decreased carbon monoxide-diffusing capacity [1, 2]. Here, we describe a 34-yr-old woman affected by localized scleroderma who presented with a number of cardiac abnormalities as the only signs of systemic involvement.

The patient reported the presence of sclerotic skin areas during the last 10 yr. She also complained of sporadic episodes of palpitations. There was no history of dysphagia, dyspnoea, joint pain or swelling, fatigue, weight loss or Raynaud’s phenomenon. She was a non-smoker, and no typical symptoms or signs of coronary artery disease were present. On physical examination, she had a lesion on her face in a typical linear distribution described as en coup de sabre, as well as the presence of circumscribed hyperpigmented sclerotic plaques on her right upper arm, right leg and right chest. All these findings were consistent with the diagnosis of linear scleroderma plus morphea. The remainder of her physical examination was normal. A complete blood cell count, ESR, thyroid hormone levels, as well as other routine laboratory tests, were all within the normal range. ANA were positive in low titre (1:64) with a homogeneous pattern, whereas antibodies against double-stranded (ds) DNA, Sm, RNP, SS-A/Ro, SS-B/la, Jo-1, centromere or SCL-70 were absent.

Further investigation to detect possible visceral involvement was carried out as normally done for all patients with suspected systemic sclerosis (SSc). Chest X-ray and high-resolution computed tomography (HRCT) scan of the lungs were normal. Pulmonary function testing showed normal respiratory volumes and normal carbon monoxide-diffusing capacity. Oesophageal manometry revealed normal oesophageal motility. Capillaroscopy and surface ECG were also normal.

Cardiac function was further assessed by sensitive diagnostic tools. These included 24 h Holter monitoring, two-dimensional and Doppler echocardiography, first-pass radionuclide angioventriculography, 99mTc methoxy-isobutyl-isonitrile (SESTAMIBI) myocardial planar scan and single-photon emission computed tomography (SPECT) performed both at rest and after exercise.

Holter monitoring showed the presence of frequent asymptomatic ventricular ectopic beats, ventricular couplets and short runs of ventricular tachycardia. Two-dimensional echocardiography and myocardial SPECT showed hypokinesis of left ventricular anterior wall and fixed perfusion defect, respectively. First-pass radionuclide angiography showed significant depression of right ventricular ejection fraction (RVEF). Considering the absence of typical symptoms and conventional risk factors for coronary artery disease, the normal ECG stress testing and the absence of reversible exercise-induced myocardial perfusion defects, we considered that our patient should not undergo coronary angiography.
Ventricular arrhythmias have never been described previously in patients with localized scleroderma. However, these are commonly found in SSC and they are associated with sudden death [4]. For this reason, it could be desirable that patients affected by localized scleroderma with cardiac rhythm disturbances be further investigated to reveal possible underlying heart disease. Our patient had impaired RVEF in the absence of pulmonary hypertension. This suggests that the reduced RVEF could be due to right cardiomyopathy. In SSC, reduced RVEF has been related to pulmonary hypertension, as previously observed in 33% of cases by right heart catheterization [3]. However, right ventricular dilatation and hypocontractility with normal pulmonary artery pressure in SSC have been recently described [4].

Cardiac perfusion defects observed in our patient in the absence of coronary artery disease are probably due to myocardial fibrosis. Various degrees of myocardial fibrosis in patients with SSC have been shown at necropsy [5]. Moreover, myocardial fixed perfusion defects in the presence of angiographically normal coronary arteries have been reported in these patients [6]. Although the pathogenesis of fibrosis in SSC is still unknown, it has been suggested that the initial tissue derangement may be related to ischaemic damage due to either anatomical or functional impairment of the coronary microcirculation [5].

We report here on a case of localized scleroderma with well-documented cardiac manifestations that should be attributed to an unusual systemic involvement of the disease. To the best of our knowledge, similar cases have never been previously reported. This observation suggests that the screening for occult cardiac abnormalities, commonly performed in SSC, could also be extended to selected patients with localized scleroderma.

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