Large vessel disease in CREST

Sir: We read with interest the report by Youssef et al of four cases of large vessel occlusive disease associated with the CREST syndrome and scleroderma. The authors described three cases of limited and one case of diffuse scleroderma in which the patients developed symptoms, signs, and angiographic evidence of large vessel occlusive disease. Furthermore, the authors suggested in the discussion that as these four patients are Caucasian, the patients, their affected population that this represented a significant association of macrovascular disease with scleroderma.
Ischaemia in scleroderma may occur owing to vasospasm,\(^2\) vascular damage,\(^1\) or abnormalities of haemostasis.\(^3\) The process may affect a wide variety of internal organs, such as the oesophagus\(^4\) and the heart,\(^5\) as well as the peripheries. Furthermore, their initial pathological studies, as the authors pointed out, did not recognise significant macrovascular disease.

The department of medicine at Ninewells Hospital has a specific interest in both scleroderma and thrombotic disorders, and we, therefore, see a great deal of small and large vessel disease associated with a variety of causes. Forty-three patients with scleroderma have attended the unit over the past five years. In view of the digital ischaemia all of these patients were screened for cardiovascular risk factors, including plasma glucose, fasting cholesterol, and triglycerides. Furthermore, all patients with digital and peripheral limb ischaemia had ankle:brachial blood pressure ratios measured. The prevalence of intermittent claudication in the general population of Lothian, a neighbouring health region to Dundee, is estimated at 4.5% and major asymptomatic disease with significant impairment of blood flow at 8.0%. In our experience three patients with progressive systemic sclerosis or limited scleroderma had significant large vessel occlusive disease—only two had symptoms—representing 6-9% of our scleroderma population. This suggests that there is no difference in the rate of large vessel occlusive disease, symptomatic or asymptomatic, between the general population and those with scleroderma.

In addition, the patients in this report were aged 49, 67, 69, and 75 years; the youngest patient had two risk factors—smoking and hypertension. It is not unreasonable, even without positive risk factors, to expect some moderate atherosclerosis to be present in their vasculature at these ages. Finally, the authors suggest that the pathological findings showed 'severe atherosclerosis rather than the expected age related atherosclerosis'. We believe this is really very subjective and should not be overinterpreted.

In conclusion, our experience contrasts with that of the authors in that we have not seen a higher rate of large vessel occlusive disease associated with the large cohort of patients attending our unit with scleroderma.

A longitudinal, prospective study of the rate of macrovascular disease examining all risk factors, including family history and atherogenic factors, is required to answer this question satisfactorily.

References: