Case report

Pancreatic necrosis in progressive systemic sclerosis

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SUMMARY Fatal pancreatic necrosis, secondary to extensive acute arteritic changes, is reported in a case of progressive systemic sclerosis. The patient presented first with hypertension and renal involvement, with active vascular lesions demonstrated by biopsy. The renal lesion at necropsy was inactive, showing the characteristic concentric fibrosis only, while the pancreatic vascular lesions were both chronic proliferative and acute in type.

Progressive systemic sclerosis (PSS), or scleroderma, is regarded as a complex disease of vascular, connective tissue, and inflammatory reactions. According to some, scleroderma is essentially a vascular disease involving the arterioles and the capillary bed in many tissues, and the pathological findings are sequelae of the vascular lesions. Vascular complications occur in scleroderma, but are less common than in periarteritis nodosa or lupus erythematosus. The vascular lesions involve both skin and internal organs, most commonly heart, gastrointestinal tract, kidney, spleen, liver, and skeletal muscle. This is a report of fatal pancreatic infarction and acute hemorrhagic pancreatitis secondary to occlusion of medium-sized pancreatic arteries, a finding so far unreported in this disease.

Case report

A 55-year old white female with skin features typical of PSS and a long history of mild hypertension (180/80 mmHg) was admitted to hospital on 15 December 1974. Her disease had become manifest about a year before, with prominent involvement of the kidneys, leading to renal failure shortly after the onset of clinical symptoms and requiring haemodialysis. She had no Raynaud's phenomenon. She had repeated episodes of fibrinous pericarditis, requiring partial pericardectomy. Her last admission (14 March 1975) was precipitated by subarachnoid haemorrhage. Despite therapy her condition gradually deteriorated and was characterised by progressive weakness and obtundation. She died of cardiac failure on 4 April 1975.

PATHOLOGICAL OBSERVATIONS

A renal biopsy specimen showed the presence of advanced vascular changes, namely, concentric thickening of intima with fibroblast-like cells present (Fig. 1) together with active disease (endarteritis) in small arteries.

At necropsy the subarachnoid haemorrhage was found to be slight and localised to 1 side of the cerebellum. Massive pancreatic necrosis, clinically unexpected, was a cause of death.

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Histologically, all sections of the pancreas showed vascular changes of scleroderma to a varying extent, involving arteries of various calibres. Recent thrombotic endarteritis was found with extensive secondary infarction of the body of the pancreas (Fig. 2). Probably as an earlier change acute panarteritis in some of the medium-sized arteries was present (Fig. 3). Other muscular arteries showed segmental necrosis of the wall, with fibrosis and in some recanalised thrombi. Many arteries showed only bland concentric fibrosis of intima (Fig. 4) or onion-skinning of the wall. There was mild atrophy of exocrine pancreas with focal fibrosis of the interstitium.

Cardiomegaly was present (weight 550 g), with scattered foci of expanded interstitium with loose amphophilic-staining ground material. Some of the smaller arteries showed marked adventitial onion-skin cufing; others showed intimal fibrosis. Fibrinous pericarditis was present. Lung changes included left lower lobe atelectasis with focal embolic acute pneumonitis. In the kidney the disease was inactive at necropsy; no thrombotic or active arteritic changes were present. The characteristic endarteritic changes involved many of the moderate-calibre arteries, interlobular and arcuate. The glomeruli showed a corresponding degree of ischaemic changes, with only rarely sclerosis of glomeruli. Atrophy of tubules was not marked, and there were only focal areas of interstitial early fibrosis. The examined segments of oesophagus and the gastro-intestinal tract were uninvolved. Sections of skin showed focal dermal fibrosis consistent with scleroderma. There was no myositis. An increased amount of haemosiderin was present in the liver, spleen, and lymph nodes, a consequence of microangiopathic haemolytic anaemia and continued haemodialysis.

Discussion

The fundamental manifestations of the patient’s disease were cardiovascular. Scleroderma was diagnosed by the typical clinical features of the disease, and by the histological skin, renal, myocardial, pericardial, and lung alterations. The diagnosis of scleroderma on renal histology alone, however, is difficult, or tenuous in the presence of longstanding hypertension (especially in the malignant
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References
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