

Scleroderma and portal hypertension

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Relationships between scleroderma and the gastrointestinal tract are well documented but involvement of the liver in this disease is less well recognized. The present communication describes four patients with scleroderma, each of whom had coincident liver disease.

Case reports

Case 1, a 53-year-old woman was diagnosed as having hypertension in 1963. The blood pressure was controlled with a diuretic. Later that year she developed Raynaud's phenomenon in both hands and dysphagia became a prominent symptom.

In 1968 she was admitted to hospital with a haematemesis and melaena and after preliminary investigations was transferred to the Bristol Royal Infirmary.

Examination

There was anaemia, and widespread telangiectases were particularly evident on the lips. Calcific nodules were prominent over the fingers and extended into the forearms. There were numerous spider naevi. There was no jaundice and no hepatic foetus. The spleen was palpable 4 cm. below the left costal margin and the liver edge extended to 8 cm. below the right costal margin. The blood pressure was 140/90 mm. Hg.

Investigations

Haemoglobin 13.3 g. per cent.; serum bilirubin 1.2 mg per cent.; serum proteins 8.1 g. per cent.; serum albumin 4.4 g./100 ml.; serum alkaline phosphatase 69 K.A. units (normal < 13 units); SGOT 125 I.U. (normal 5-46 units); smooth muscle antibody—negative; antinuclear factor—positive 1/20; mitochondrial antibodies—negative.

A barium swallow examination showed oesophageal varices and splenoportography showed the portal and splenic veins to be patent. A skin biopsy showed changes of scleroderma.

Treatment and progress

At operation an end-to-side porto-caval anastomosis was performed. The gall bladder and extrahepatic biliary tracts were normal. The liver architecture was normal. There was some accumulation of lymphocytes and epithelioid cells near bile ducts. These 'granulomata' probably represented extravasated bile.

The postoperative course was uneventful and the patient is alive and well 12 months later.

Case 2, 49-year-old woman, came to the outpatient department in September 1969. One month previously she had taken aspirin for a headache after which she had had melaena. She had no haematemesis. In the previous month she had gained one stone in weight, and her abdomen had become distended.

After a pregnancy complicated by toxæmia, 20 years earlier, the patient had been hypertensive and was treated with α -methyl dopa. She had suffered from migraine for many years and had noticed that her fingers became numb and changed colour in cold weather and on immersion in cold water.

Examination

She was clinically anaemic. The blood pressure was 150/90 mm. Hg. The skin over the middle phalanges was shiny and tight. The fingers were cold. Apart from ascites there was no extrahepatic evidence of liver disease.

Investigations

Haemoglobin 7.4 g. per cent.; serum bilirubin 1.2 mg. per cent.; serum proteins 5.8 g./100 ml.; serum albumin 3.5 g./100 ml.; LE-cell preparation negative; serum alkaline phosphatase 24 K.A. units; RA latex-fixation test—positive; smooth muscle antibody—negative; mitochondrial factor—negative; antinuclear factor—positive (1/12 dil.); bromsulphthalein retention—21 per cent at 45 min.

A barium swallow examination revealed oesophageal varices; splenoportography showed the portal and splenic veins to be patent; hepatic wedge pressure 16 mm. Hg; splenic pressure 31 mm.; ¹¹³I rose Bengal scan within normal limits.

Liver biopsy (operative) revealed that the liver architecture was grossly disturbed. The pattern was nodular, the nodules being surrounded by reticulin fibres and collagen. The portal areas were infiltrated by lymphoid cells. Twin cell plates were identified in some of the nodules. Skin biopsy revealed histological features typical of scleroderma.

Treatment and progress

After a period of rest in bed and diuretic therapy the ascites gradually disappeared. Anaemia was corrected by a blood transfusion. The portal hypertension was relieved operatively by an end-to-side porto-caval anastomosis. Since operation the patient has remained well and has needed no hypotensive therapy.

Case 3, a 63-year-old woman, was admitted to the Bristol Royal Infirmary in 1966. She had been in another hospital because of a haematemesis due to bleeding oesophageal varices. In 1955 she had developed Raynaud's phenomenon for which she had been treated, unsuccessfully, by bilateral cervical sympathectomy. In 1965, telangiectases developed over the face, and together with calcified subcutaneous nodules and tight skin over the fingers suggested a diagnosis of scleroderma which was confirmed by skin biopsy.

Examination

In the Bristol Royal Infirmary in 1966 clinical examination confirmed the diagnosis of advanced scleroderma. Abdominal examination revealed marked abdominal distension with ascites.

Investigations

Haemoglobin 12 g. per cent.; plasma proteins 7.8 g. per cent.; serum albumin 3 g. per cent.; serum alkaline phosphatase 29 K.A. units.

Liver biopsy (operative) showed active chronic hepatitis. A barium swallow examination showed evidence of oesophageal varices.

Treatment and progress

Porto-caval anastomosis was not performed. The oesophageal varices were ligated through a transthoracic approach.

Termination

For 3 years after the operation the patient was well, but in 1969 she died in hepatic failure.

Case 4, a 65-year-old woman, first became ill in 1953, when she presented with bilateral dropped wrist and dropped foot. There were few sensory signs. At the start of the illness a diagnosis of a lead neuropathy was made and she was treated with chelating agents. The neurological symptoms and signs persisted and became more severe, so that she was considerably disabled. The clinical picture became that of motor neurone disease.

For most of her life the patient had complained of symptoms of Raynaud's phenomenon. Recently she had complained of dysphagia.

Examination

In 1969 she had a severe haematemesis. In hospital she was found to have subcutaneous calcinosis together with typical changes of scleroderma in the terminal phalanges. Neurological examination confirmed the bilateral wrist and foot drop. Abdominal examination revealed marked hepatosplenomegaly.

Investigations

Hb 11.9 g. per cent.; liver function test; bilirubin 0.7 mg. per cent.; albumin 3.4 g. per cent.; globulin 5.4 g. per cent. (electrophoretic strip—raised γ globulin); mitochondrial antibodies—negative; antibodies to smooth muscle—negative; antinuclear factor—positive (1/80).

A barium swallow showed oesophageal varices.

Treatment and progress

The patient underwent oesophageal transection. She had no further episode of gastrointestinal bleeding.

Termination

In March, 1971, the patient died. At post mortem the liver appeared cirrhotic. Histologically there was a well-marked lymphocytic infiltration of the portal tracts, with 'aggressive' fibrosis.

Discussion

Liver disorder was a prominent feature in four patients with scleroderma. Whilst involvement of the gastrointestinal tract in scleroderma is well recognized, hepatic involvement is less common and is seldom a dominant feature of the condition. A recent American post mortem study of 58 patients with scleroderma revealed cirrhosis in five of 57 of the patients in whom liver tissue was available for examination, which was less than the eleven cases of liver disorder found in a carefully matched control group (D'Angelo, Friels, Masi, and Shulman, 1969). Bartholomew, Cain, Winkelmann, and Buggenstoss, (1964) were able to find only eight instances of liver involvement in 727 patients with scleroderma, whilst another large series of 150 cases does not mention liver disease but comments on arterial lesions which were found in the gall bladder (Leinwand, Duryee, and Richter, 1954). In Great Britain the situation appears to be different, for four patients with scleroderma and liver disorder have been seen in Bristol in the period 1965-1969 in a hospital in which an average of fifty new cases of cirrhosis are seen each year.

Current opinion suggests that scleroderma is an autoimmune disorder and as such occasionally occurs with other autoimmune diseases such as rheumatoid arthritis and systemic lupus erythematosus (Rodnan, 1966). Although overt liver disorder is rare in autoimmune conditions, the detection of antibody markers has shown that sub-clinical liver involvement may occur (Whaley, Goudie, Williamson, Nuki, Dick, and Buchanan, 1970; Walker, Doniach, and Doniach, 1970). Such relationships strengthen the possibility of a connection between scleroderma and liver disease and suggest that the connection may be autoimmune.

In the present patients liver involvement was reflected by gastrointestinal haemorrhage, indicating in each case, portal hypertension. Calvert, Barling, Sopher, and Feiwel (1958) described two patients with hepatic cirrhosis and portal hypertension in whom alimentary haemorrhage was an important feature. Gastrointestinal haemorrhage in scleroderma, however, may be associated with other manifestations of the disease. One patient has been described in whom haematemesis and melaena were associated with bleeding telangiectases of the stomach (Kolodny and Baker, 1968) and yet another patient had oesophageal haemorrhage arising from ulcerative oesophagitis (Berliner and Burson, 1966).

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