Chronic Disease of the Liver Associated with Systemic Scleroderma

LLOYD G. BARTHOLOMEW, M.D., JAMES C. CAIN, M.D., R. K. WINKELMANN, M.D., and ARCHIE H. BAGGENSTOSS, M.D.

Systemic scleroderma is an uncommon disease that is most easily recognized through its expression of cutaneous sclerosis. More recently, it has become possible to recognize involvement of other organs, and it is now accepted that the esophagus, small intestine, lungs, heart, and kidneys are frequently involved in the course of a severe case.1

The liver has rarely been reported as involved in scleroderma, and the evidence supporting significant hepatic disease is not well documented. Goetz and Berne,2 coining “progressive systemic sclerosis,” discussed the disease in detail, while mentioning biliary cirrhosis only as an uncommon and probably an incidental finding. Goldgraber and Kirsner3 in 1957, in a comprehensive review of scleroderma of the gastrointestinal tract, did not mention hepatic involvement. Calvert and coworkers4 in 1958 described two cases of scleroderma in which hepatic fibrosis with portal hypertension developed. Tissue was not available for study in either case. Batsakis and Johnson5 in 1960 reviewed the infrequent references in the literature to the hepatic involvement associated with scleroderma and referred especially to the marked periportal fibrosis and chronic inflammatory changes occurring within the liver. Treacy6 described in detail the changes occurring in the gastrointestinal tract in 15 necropsies of scleroderma and mentioned one in which death was due to hepatic failure.

Of 727 patients with systemic scleroderma,7 8 had evidence of serious hepatic disease. Because tissue was available in most of these, we attempted to determine the types of hepatic damage that might be seen in scleroderma and to see whether the scleroderma might be an etiologic factor in the hepatic disease.

From the Section of Medicine, Section of Dermatology, and the Section of Experimental and Anatomic Pathology, Mayo Clinic and Mayo Foundation, Rochester, Minn.

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REVIEW OF CASES

Case 1

A 57-year-old white woman was seen in August 1958, because of recurrent gastrointestinal bleeding. She denied any history of previous hepatic disease. Symptoms suggestive of scleroderma began at age 20 when she noted typical Raynaud's phenomenon. For 4 years she had noticed progressive dysphagia, exertional dyspnea, ankle edema, intermittent fever, recurrent thrombophlebitis, and a gradual loss of 65 lb.

Physical examination revealed a chronically ill, jaundiced patient with sclerodematous changes involving her hands, feet, and face. The abdomen contained ascitic fluid and the liver was irregular and extended to the umbilical level. The spleen was felt 4 cm. below the left costal margin.

The laboratory findings are given in Table 1. Of particular interest are those related to the marked hepatic dysfunction and the positive lupus erythematosus clot test. X-ray examination revealed cardiac enlargement and varices in the lower part of the esophagus. Esophageal-motility studies revealed small, weak, and frequently simultaneous contractions and at times no contractile responses to swallowing. The electrocardiogram showed low amplitude of the QRS complex throughout and inversion of the T wave in leads II, III, V-4, V-5, and V-6. Pulmonary function studies demonstrated evidence of a diffuse pulmonary fibrosis.

Needle biopsy of the liver revealed small regenerative nodules separated by inter nodular zones broadened by large numbers of fibroblasts and lymphocytes and some collagen fibers (Fig. 1). There was a mild degree of atypical ductular proliferation, and septal formation was prominent. Activity of the process was indicated by necrosis of the limiting lamina of liver cells of the regenerative nodules with a polymorphonuclear leukocytic and fibroblastic reaction. There was also focal necrosis of the regenerative nodules with a Kupffer cell reaction. A diagnosis of postnecrotic cirrhosis was made.

Fig. 1. Biopsy (Case 1) material showing small regenerative nodules with necrosis of cells of limiting plate and leukocytic reaction. (H&E, × 200)