A case of primary fibrosarcoma of the liver

D. SMITH
M.R.C.Path.

S. R. RELE
M.B.B.S., D.C.H.(Bom.), D.C.H.(Lond.)

West Cumberland Hospital, Whitehaven, Cumberland

Less than a dozen cases of primary fibrosarcoma of the liver have been reported. Cavallio, Lichewitz & Rozov (1968) accept only five cases and add one of their own.

Case report

A labourer of 37 was admitted to hospital in August 1968 with pleuritic pain in the right chest and shoulder of 4-weeks' duration.

He was pyrexial (T 37·7° C) tender over the right lower ribs, with impaired percussion note, diminished air entry and a pleural friction rub at the right base. The liver edge, firm and tender, was palpable just below the costal margin. The spleen was not felt. Appendicectomy had been performed 6 weeks previously. No abnormality was found at operation and the appendix was histologically normal.

Investigations

He had a normocytic, normochromic anaemia, (Hb = 55%) a leucopenia (3700/mm³) with a normal differential, and an ESR of 109 mm (Wester-gren). Serum bilirubin, flocculation tests and SGPT were normal. Alkaline phosphatase was elevated, (55 KA units) serum albumen was 2·4 and globulin 3·8 g/100 ml, with slight increase in α₂ and γ-globulins. Blood culture was sterile. Chest X-ray showed an elevated right diaphragm with patchy collapse at the right lung base and shift of the mediastinum and heart to the left. Screening revealed a completely immobile right hemidiaphragm. No subphrenic gas was seen, but in view of recent appendicectomy, the abdomen was explored to exclude a subphrenic abscess. No fluid or pus was found, but the liver was large and tense. A large bore needle was inserted into it but no cyst or abscess was found. Liver biopsy showed only sinusoidal congestion and patchy lymphocytic infiltrate in portal tracts. Bronchial carcinoma with phrenic paralysis was considered, but bronchoscopy revealed no evidence of tumour.

He deteriorated steadily, with increasing cachexia, dyspnoea, intermittent pyrexia and massive right chest dullness, the heart being displaced into the left axilla. No pleural fluid could be aspirated. The liver remained palpable three fingers below the right costal margin. X-ray showed complete opacity of the lower two-thirds of the right lung field, with a convex upper margin due to gross elevation of the right diaphragm.

He died on 20 April 1969.

Necropsy

The liver was enormously enlarged, (7325 g) extending high into the thorax with the right hemidiaphragm stretched over its surface. The lower and middle lobes of the right lung were collapsed. No tumour was found in the bronchi or lungs. A massive, single, soft, fleshy, tan to cream-coloured tumour enlarged and replaced the right lobe of the liver. Its edge was irregular, but well-demarcated from the surviving liver tissue. Yellowish areas of necrosis and friable haemorrhagic areas were present and lobulated cream-coloured tumour penetrated through the capsule on the inferior surface of the liver, extending to surround the upper pole of the right kidney and the adrenal. Areas of liver free from tumour appeared normal and were not cirrhotic. The portal and hepatic veins and spleen were normal and no tumour was found in any of the other abdominal viscera. The brain was normal.

Histology (Fig. 1)

The tumour was a poorly-differentiated fibrosarcoma with myxomatous areas. Sections from different areas showed similar appearances, elongated spindle-shaped cells with basophilic nuclei, lying in a loose connective tissue stroma containing extracellular mucin in areas. Connective tissue stains demonstrated mature and immature collagen associated with the tumour cells, and the close pericellular reticulin network indicated a mesenchymal rather than epithelial origin. The tumour cells showed hyperchromatism, frequent mitoses and some pleomorphism. Special stains failed to demonstrate cross-striations, myofibrils, or fat within tumour cells. Extensive areas of necrosis and haemorrhage were present. Areas of liver uninvolved by tumour were normal.
Discussion

The study of large series of primary liver tumours shows that fibrosarcoma is very rare.

Simpson, Baggenstoss & Stauffer (1955) found only one case amongst the primary malignant neoplasms of the liver seen at the Mayo Clinic in over 24,000 necropsies. Steiner (1960) found only one fibrosarcoma amongst 860 primary malignancies of the liver, and Balouet & Destombes (1967) describe a single case in a study of 216 primary liver tumours. Two cases were seen in the Department of Pathology at Kyoto University during a 5-year period (Ojima et al., 1964).

Occasional reports of spindle-cell sarcoma of the liver appear in the earlier literature, but their true identity is doubtful owing to the histological similarity between sarcoma and highly anaplastic carcinoma. Evans & Hoxie (1937) reported a case of myxosarcoma of the liver, the naked-eye and histological appearances of which resemble the present one.

In adults, the symptoms of primary liver sarcoma are ill-defined. Weakness, anaemia, anorexia, dyspepsia, an enlarging abdominal mass, and pain in the right upper quadrant with radiation to the right posterior chest and shoulder occur commonly. Jaundice is frequent, due to obstruction of the extrahepatic bile ducts. Most cases are rapidly fatal, but Snapper, Schraft & Ginsberg (1964) describe recurrent hypoglycaemic attacks in a man who survived for 4 years after biopsy of a liver tumour had shown it to be fibrosarcoma. Small lung metastases were present at necropsy. Lymph node metastases have been described (Totzke & Hutcheson, 1965; Simpson et al., 1955) and in the latter case the tumour had invaded the wall of the jejunum. In the only patient treated by surgical resection (Shallow & Wagner, 1947) the patient collapsed and died on the eleventh postoperative day, presumably from pulmonary embolism, but permission for necropsy was not given.

The site of origin of liver sarcoma is ill-defined and seems to be variable. Origin has been suggested from the perivascular connective tissue, cellular elements of vessels, perilymphatic tissue, connective tissue of the portal tracts, regeneration nodules in cirrhosis, or from the wall of a cyst or abscess.

The association of fibrosarcoma of the liver with cirrhosis is variable. It was absent in the present case.

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References


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