CASE REPORTS

Malignant epithelioma of the liver

A. K. BURROUGHS
M.B., Ch.B., M.R.C.P.

S. J. BARTER
M.B. B.S.

W. J. JENKINS
M.Sc., M.R.C.P.

Academic Department of Medicine, Royal Free Hospital, London NW3 2QG

Summary
Carcinoma complicating congenital cystic conditions of the liver is rare. The authors report a primary malignant epithelioma of the liver in association with multiple intrahepatic biliary cysts, which are not usually considered to predispose to malignancy.

Introduction
There are several congenital cystic conditions affecting the liver and bile ducts. The first group includes the intrahepatic cysts, which may be solitary or multiple, and their precursors, the von Meyenburg complexes. These are microscopic areas of duct ectasia which may enlarge gradually to form macroscopic cysts. The second group includes those lesions affecting the biliary drainage system itself, congenital hepatic fibrosis, cystic dilatation of the intrahepatic ducts, and choledochal cysts. The risk of malignant tumours complicating this second group of conditions has been recognized for some time. Bloustein (1977) assessed the incidence of malignant change as 1% in congenital hepatic fibrosis, 4% in choledochal cysts, and 7% in congenital cystic dilatation of the intrahepatic ducts. In contrast, malignancy in solitary non-parasitic cysts of the liver, or in polycystic liver disease has been described relatively rarely in single case reports by Willis (1943), Richmond (1956), Edmondson (1958), Ameriks, Appleman and Frey (1972), Greenwood and Orr (1972), an unpublished case but cited by Bloustein (1977), and 2 cases by Cruikshank (1961) in a series of primary liver cell carcinomas. A large series reported by Melnick (1955) includes 2 cases of benign neoplastic transformation of von Meyenburg complexes in 70 cases of polycystic liver disease, but no malignancies. Homer, White and Read (1968) have also described benign neoplastic change of von Meyenburg complexes in 2 cases, and an adenocarcinoma arising in a third.

Case report
In February 1979 a 75-year-old woman was admitted to hospital complaining of pain in the right upper abdominal quadrant, abdominal swelling and increasing breathlessness. On examination, a firm irregular liver was palpable 12 cm below the right costal margin, and the spleen was palpable 2 cm below the left costal margin. There were no liver bruits.

The patient had presented with similar findings in 1950, when multiple non-parasitic liver cysts containing brown acellular fluid were found at laparotomy. Histology of the cysts showed that they were lined with biliary tract epithelium. The cysts became infected 5 times between 1950 and 1964, and on each occasion the infection was treated with antibiotics and surgical drainage.

On admission in 1979, the Hb (13·4 g/dl) and WBC (5·7 x 10⁹/l) were normal, but the alkaline phosphatase 30·4 u./l (normal 3–13) and aspartate aminotransferase 38 u./l (normal 4–15) were raised. The serum bilirubin, albumin, prothrombin time and immunoglobulins were normal, and hepatitis B surface antigen and alphafoetoprotein tests were negative. A ⁹⁹mTc liver scan showed a large defect in uptake in the right lobe. A grey-scale ultrasound examination showed not only multiple cysts but a solid mass infiltrating the liver. A CT scan confirmed this mass in the right lobe, and demonstrated small cysts in the left lobe. No abnormality was seen in the pancreas, kidneys or intestine. A needle biopsy of the mass in the liver showed a malignant epithelial tumour, growing in closely packed islands and nests resembling bile duct epithelium (Fig. 1). The patient,
who was given regular analgesia to control her abdominal pain, died at home 6 weeks later.

Discussion

Lowenfels (1978) has suggested that bile stasis or increased bacterial growth in bile may be responsible for the increased incidence of biliary tract carcinoma seen in cholelithiasis, choledochal cyst and ulcerative colitis. In ulcerative colitis, biliary tract carcinoma may be related to the high incidence of sclerosing cholangitis reported in that condition (Chapman et al., 1980). It certainly appears that malignant change in congenital cystic conditions of the liver is more common where the bile is in direct contact with the epithelium lining the cysts, such as in cystic dilatation of the intrahepatic bile ducts and in choledochal cysts. In contrast, in solitary cysts and in polycystic liver disease there is usually no contact with bile (Gallagher, Millis and Mitchinson, 1972), and neoplastic change is rarer. In addition, infection is also less common in the latter conditions. It may be that the fatal biliary malignancy reported here could have been related to the multiple infections of the cysts. Reports of cases such as the present one are few but other similar cases may go unreported in the belief that they are extreme rarities, so that the true incidence of neoplastic change in congenital cystic disease of the liver and biliary tract may have been incorrectly assessed.

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References


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A. K. Burroughs, S. J. Barter and W. J. Jenkins

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