A Case of Polycystic Disease of the Liver and Kidneys

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SUMMARY

A case of polycystic disease of the liver and kidneys in a middle-aged Black woman is reported. The condition presented as an abdominal mass, and the diagnosis was established at laparotomy. Surgical treatment of the cysts was not indicated.

The patient also suffered from pellagra.

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Congenital cysts are common enough in the kidneys, but are rare in the liver.[1] Polycystic disease of the liver has been found mostly at necropsy, and clinical diagnosis has proved difficult. Investigation of some cases of renal polycystic disease has shown associated hepatic cysts,[2] but very few cases have presented with symptoms attributable to the hepatic lesion.[3-6]

CASE REPORT

A Black woman aged 40 years was recently admitted to the H. F. Verwoerd Hospital. In addition to a skin rash, she complained of a painless epigastric mass which had been present for about 4 years. She had experienced no symptoms other than occasional attacks of vomiting after heavy meals, and some nocturia. Her diet consisted largely of maize, milk and vegetables. The family history was not contributory and her 5 children were healthy.

Examination showed the cutaneous eruption of pellagra on the sun-exposed areas. A large epigastric tumour which moved on deep inspiration was visible. On palpation this tumour proved to be part of the liver which was enlarged to at least 16 cm below the right costal margin. The surface of the liver was very irregular and soft in places. The kidneys were not palpable and no other masses were present. The blood pressure was 140/90 mmHg.

Investigations showed a normal blood count, but the erythrocyte sedimentation rate was 31 mm in the 1st hour (Westergren). The usual tests of liver and kidney function, including an intravenous pyelogram, showed no abnormalities. Radiological findings included bilateral elevation of the diaphragm, more marked on the right side (straight X-ray film); a barium meal showed marked displacement of the stomach to the left and posteriorly (Fig. 1), and slight distortion and rotation of 1 calyx in the right kidney (retrograde pyelogram).

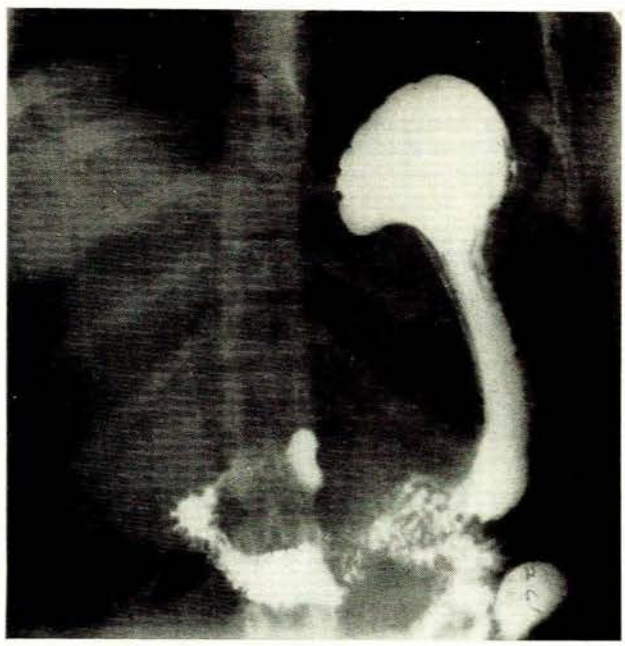

Fig. 1. Barium meal showing the displacement of the stomach to the left and posteriorly.

A scintigram of the liver (using selenium-75) confirmed the gross hepatomegaly, more marked on the right, and showed parenchymal loss with a number of large areas where the isotope was poorly concentrated; the diagnosis of a hepatoma was considered.

The true nature of the hepatic lesion was only revealed at laparotomy. The liver was found to be grossly enlarged, particularly the right lobe, and covered with round or oval cysts, measuring 1 - 15 cm in diameter.

Several specimens were taken from the cyst walls for biopsy, and the fluid content of the cysts was found to be clear and watery. The intervening hepatic tissue was pale and firm. About a dozen cysts of similar appearance were noted in each kidney, but the kidneys were otherwise normal. Surgical ablation of the cysts was not attempted.

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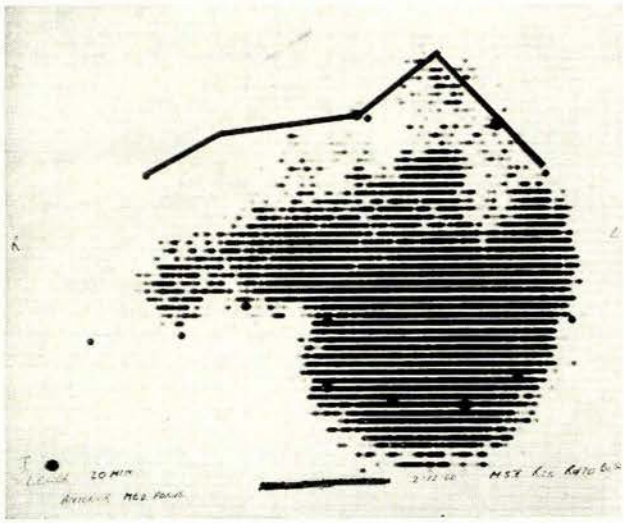

Fig. 2. Liver scintigram (anterior focus) showing the hepatomegaly, loss of parenchyma, and areas of poor isotope concentration.

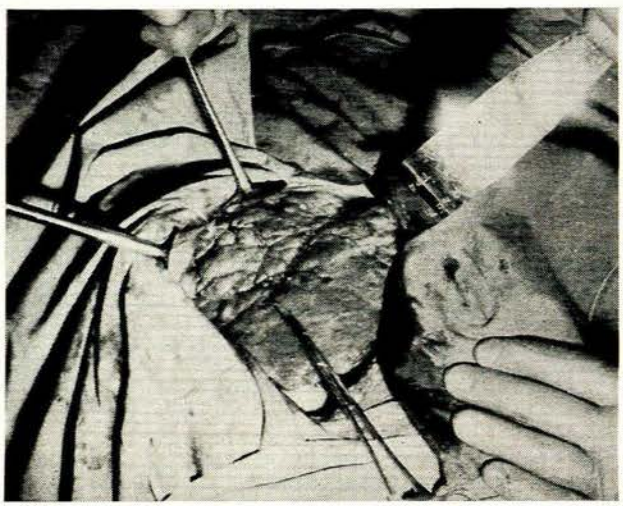

Fig. 3. The liver at laparotomy, with the round or oval cysts on the anterior surface measuring 1 - 15 cm in diameter.

The hepatic biopsies showed multiple hamartomatous cysts, lined by a layer of columnar epithelium of bile-duct type.

The patient made an uneventful recovery. Her pellagra had cleared in the meantime on an ordinary ward diet with vitamin supplements.

DISCUSSION

Polycystic disease of both liver and kidneys is rare, and has seldom been reported during the past 20 years. Ackman and Rhea[7] found only 6 cases of non-parasitic hepatic cyst in 6 141 autopsies. Melnick[1] reported 1 case occurring in a 36-year series of 687 necropsies. The disease is seen more often in older patients as the cysts evolve gradually with age.[1]

About 50% of cases of hepatic polycystic disease are associated with cysts in the kidney,[1] although only 30% of the adult cases of the latter have related hepatic cysts.[2] Infantile, polycystic renal disease is invariably accompanied by multiple. epithelial-lined cysts in the liver, or by bile-duct proliferation.[2] Associated cysts occur more rarely elsewhere, but have been found in pancreas, parathyroid, pineal gland, peritoneum, and renal pelvis.[1] Poutasse *et al.*[8] have reported a case of hepatic cysts with associated intracranial aneurysms.

The hepatic cysts are lined by columnar epithelium of the bile-duct type, and are surrounded by fibrous connective tissue. Various theories have been proposed concerning the origin of these cysts. Melnick[1] described 2 cases in which newly-formed intralobular ducts arising from hepatic cells were found in adult livers. It does seem probable that the cysts may arise from the dilatation of persisting Meyerburg's complexes (the intralobar bile-duct islands that normally involute in the foetus).

The type of treatment required for this condition is determined by the extent of the liver involvement and the presence of complications. Surgical procedures include simple puncture, drainage, marsupialisation, or resection,[9] but normally only symptomatic treatment is necessary.

The prognosis depends on the severity of the involvement of the liver, and especially of the kidneys, if renal cysts are present. The hepatic affection is usually mild, but death from an *E. coli* infection of the hepatic cysts has been reported.[6]

Notwithstanding her age and her 5 pregnancies, the patient's renal and hepatic functions were normal. As the prognosis appeared relatively favourable, surgical treatment was not considered necessary. The pellagra was clearly an unassociated condition. Regular follow-up will be required to treat complications, such as urinary tract infections, if and when they arise.

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