ISOLATED NON-PARASITIC CYSTS OF THE LIVER IN NEWBORN INFANTS

REVIEW AND CASE REPORT

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Reports in the literature have suggested that isolated non-parasitic cysts of the liver, polycystic disease of the liver and kidney excluded, are uncommon. Detailed reviews have been presented by Munroe[10] and Geist,[5] who collected 193 cases from the literature. Since then numerous reports have appeared, including that of Desser and Smith,[4] who found 8 more cases in hospital records. Warren and Polk,[11] Morgenstern[9] and others have also added to the literature. The records of the Department of Pathology, University of Cape Town, show at least 5 more examples in adults found incidentally on postmortem examination. It would seem that the overall incidence of these cysts is higher than would appear from the numbers reported in earlier years.

These cysts have been reported less commonly in the paediatric age-group. Desser and Smith[4] found 31 examples in the literature in children under 13 years of age, and the 8 cysts they themselves reported were all in children. Of these, 3 were in neonates. Of Geist's series[5] only 2 were in neonates. One of these was that reported by Bagot (1892).[1] It was a large cyst of the liver in an infant with a papular eruption of syphilitic character and it caused dystocia. Muller, quoted by Bagot, noted in an infant a 'lymphatic tumour of the liver' weighing 4 lb., which also obstructed labour. Moll[8] (quoted by Geist) described a cyst of the right lobe of the liver containing 110 ml. of fluid in a neonate. Thus it can be seen that these cysts have been reported very rarely in neonates, only 6 being on record. The following additional case is therefore of some interest, presenting with the unusual combination of a liver cyst with anaplastic giant cells of the foetal adrenal cortex.

CASE REPORT

History

The mother, a coloured patient (i.e. of mixed White and

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non-White ancestry) was admitted in labour at 36 weeks with pre-eclamptic toxaemia. Soon after delivery postpartum haemorrhage necessitated manual removal of the placenta; the mother died half-an-hour later.

The infant was limp and cyanosed at birth, and was given oxygen. A large flabby abdomen was noted. The infant died two-and-a-half hours after birth.

Findings at Autopsy of Infant

The infant's weight was 3,044 g. and the length 49 cm. The abdomen was enlarged and flabby. There was a large caput succedaneum, partial bilateral talipes equinovarus and short metatarsals of the little toes.

Liver cyst. The cause of the enlarged abdomen was a huge laxly-filled, reddish-brown cyst, 13 cm. in diameter, hanging from the posterior surface of the right lobe of the liver (Fig. 1). It lay anterior to the intestine, its wall being

Fig. 1. Liver with the attached cyst.

contiguous with that of the fundus of the gallbladder. It weighed 300 g., its bulk being nearly twice that of the liver, which weighed 154 g. A leash of blood vessels ran over the surface of the cyst from its hepatic attachment. Its base was a triangular area of the posterior surface of the liver above the fundus of the gallbladder. A small amount of liver tissue was present in that part of the wall of the cyst which was in contact with the liver. The cyst, which was unilocular, contained 285 ml. of yellow-brown fluid as well as an off-white coagulum which had been visible through the wall of the cyst. After the fluid had been removed, the cyst-wall was seen to be smooth, reddish and translucent.

Cyst fluid. Chemical estimation of the fluid was as follows: Van den Bergh, negative; bilirubin, 1·2 mg. per 100 ml. (mainly indirect); cholesterol, 26 mg. per 100 ml.; alkaline phosphatase, 2·7 Bodansky units; proteins, albumin, 2·0 g. per 100 ml., globulin 4·1 g. per 100 ml. No crystals were seen in the fluid on microscopy.

Other organs. The liver, apart from the cyst, was normal, as were the gallbladder and bile ducts. The adrenals (9 g.) had a normal foetal appearance. The thyroid showed a marked generalized enlargement — 2 - 3 times normal, but unfortunately was not weighed. The other organs showed no special features.

Histology

At the cyst's origin the liver substance tailed off gradually into the cyst wall with no epithelial lining. The wall consisted of 3 ill-defined layers. In the middle there was loose connective tissue containing scanty bile ducts and many sinusoids. The outer and inner layers consisted of scanty flattened cells in connective tissue. The wall of the fundus of the gallbladder was contiguous microscopically with that of the cyst in one small area. The liver elsewhere showed very active extramedullary haemopoiesis, with some increase in portal connective tissue.

The thyroid showed absent colloid and increased collagenous tissue intralobularly and between the acini. In the pancreas the islets of Langerhans were prominent and some extramedullary haemopoiesis was present.

The adrenal gland (only 1 was sectioned for histology) contained striking anaplastic giant cells (Fig. 2). There were

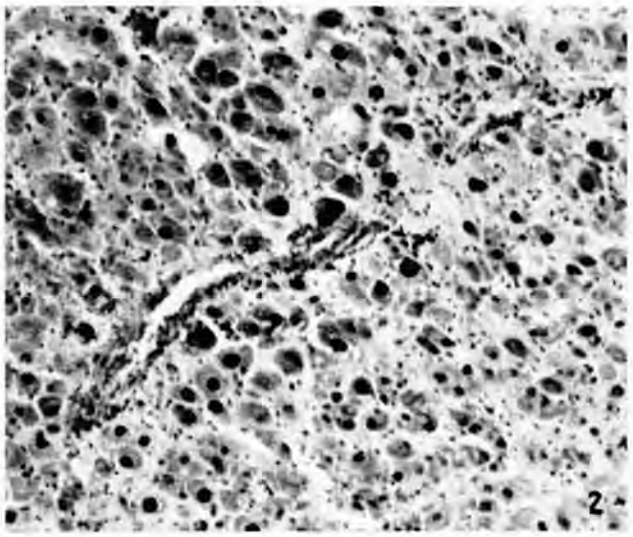

Fig. 2. High-power view of the foetal adrenal cortex showing numerous giant cells.

numerous giant cells in most of the foetal cortex though none was seen in the definitive cortex. They were mostly in the inner areas of the foetal cortex and showed pronounced nuclear and cytoplasmic anaplasia. In these areas there were also numerous normal cells. The giant cells were large, up to 6 times the diameter of normal cells, and were distributed in sheets, clumps and isolated individual cells. The nuclei were hyperchromatic, with well-defined borders, and their chromatin was distributed in a granular and linear network. Their shape was usually round or oval, often irregular. The nuclei were large in relation to the cytoplasm. A few cells had up to 5 nuclei, and many of the nuclei contained slightly eosinophilic inclusions up to half the size of the nucleus. The cytoplasm was similar to that of the normal cells, but in some there were numerous clear vacuoles, most marked round the nucleus. The small, thin, definitive cortex contained no lipoid. Some extramedullary haemopoiesis was noted in the innermost portion of the gland.

The other organs were normal on histological examination.

DISCUSSION

Origin of the Cyst

The site of origin of the cyst is speculative. It may have originated in the liver capsule, or in hepatic tissue beneath the capsule, growth being in the line of least resistance outwards, the huge size of the cyst causing great distension and thinning of the wall. This may explain the scanty bile ducts and absence of liver cells in the wall compared with those noted in other cases. The liver capsule surrounded the cyst and therefore, though the fundus of the gallbladder was contiguous with the cyst wall, the gallbladder could not have given rise to the cyst.

The histogenesis of the cyst may have been from an intrahepatic bile duct, the bilirubin in the cyst fluid being compatible with this possibility. Other structures from which it could have arisen are lymphatic, venous, arterial or capillary vascular channels, but the simple structure of

the cyst does not favour this origin. The first suggestion seems the most likely.

The absence of any epithelial lining is worthy of comment. Very little active secretion could have occurred, yet the laxly-distended cyst contained 285 ml. of fluid.

These cysts so far have been of no clinical importance in neonates except when of great size, when they have caused obstructed labour.

Association with Giant Cells in the Foetal Adrenal Cortex

Giant cells were first noted by Kampmeier[6] in the adrenal cortex of foetuses aged 2 - 4 months. In recent years they have been studied by several authors.[2,3,7] They appear to be related to cells found in the adrenal glands of normal foetuses. Under certain circumstances these cells remain conspicuous until a few months after birth, and very rarely may be present many years later.[7] No effect can yet be ascribed to them, and there is no evidence in favour of a neoplastic or viral origin of these cells. Their fate is a mystery.

They have not hitherto been associated with a cyst of the liver. The occurrence of both these conditions in this case is thought to be purely coincidental.

SUMMARY

The literature on isolated non-parasitic cysts of the liver is briefly reviewed, the rarity of reports of these cysts in neonates being noted. An additional case is described in a neonate, where the unusual combination of a liver cyst with anaplastic giant cells in the foetal adrenal cortex was found. The combination is thought to be coincidental.

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