lined by squamous, columnar or respiratory epithelium with areas of muscle, glial tissue, cartilage and glandular tissue. These morphological findings were similar to those seen in our case. Malignant variants of this tumour are extremely rare, and the importance of early and correct diagnosis is stressed, since in uncomplicated cases surgery is curative.

REFERENCES


Renal Echinococcosis in Children

A Report of 2 Cases

P. D. GAJJAR, C. C. SINCLAIR-SMITH

SUMMARY

Two cases of renal echinococcosis in children are described. In one of the children the lungs and liver were involved as well. The pathology, diagnosis and treatment, as well as the usefulness and limitations of the Casoni skin test and serological tests, are discussed.


Echinococcosis presents a serious threat to man in many parts of the world. The disease has frequently been recorded in Asia, Australasia, South America, Iran, Iraq, southern Africa and the Mediterranean countries, while isolated cases have occurred in North America. Although the liver is reputed to be the organ most frequently involved, Slim et al. found the lungs to be the commonest site, 22 of their series of 34 children having been so affected. This was confirmed by Auldist and Myers. The brain, kidneys, spleen and bone are also commonly involved, as are other sites. In a series of 22 cases of echinococcosis, Amir-Jahed et al. reported that the disease had affected the stomach, the small bowel and its mesentery, the large bowel, and the omentum, peritoneum, gallbladder, pancreas, heart, femur and urinary bladder. The kidneys are involved in approximately 3% of all cases of hydatid disease, and the cysts are almost invariably unilateral and single. Neither Deliveliotis et al., who reported 12 cases of renal echinococcosis, nor Makki, who reported 5, mention involvement of other organs.

CASE REPORTS

Case 1

A 9-year-old Coloured girl from the Eastern Cape presented at Groote Schuur Hospital in April 1961 with a history of intermittent left chest pain, and a progressive cough associated with haemoptysis. A general examination, which included the chest, revealed no abnormality, but a large mass in the left loin and a 2-cm hepatomegaly were palpated.

Her haemoglobin concentration was 11,0 g/100 ml, the white cell count was 5,000/µl and the erythrocyte sedimentation rate was 52 mm/1st h (Westergren). A chest radiograph revealed radio-dense lesions in the left lower lobe and right middle lobe which are compatible with hydatid cysts, and calcification in the walls of the abdominal mass. The results of the Casoni skin test were positive.

At operation a large cyst, which virtually replaced the left kidney, was found, and a nephrectomy was performed. A second cyst found in the liver was treated by needle aspiration of the cyst contents, followed by injection of formalin into the cyst and an endocystectomy. Two cysts were removed from the left lobe of the lung.

During a subsequent admission a month later, a
bronchogram showed that the right middle lobe cyst was communicating with the right main bronchus, which necessitated a right middle and right lower lobectomy. The patient was declared well and discharged.

Pathological examination confirmed the diagnosis of hydatid cysts. In the kidney the cyst measured 14 cm in diameter, and occupied the lower pole, and there were scolices.

Case 2

An 11-year-old Coloured boy presented at a large hospital in South West Africa with a 3-month history of a swelling in the left hypochondrium, and a provisional diagnosis of Wilms' tumour was made. He was transferred to the Red Cross War Memorial Children's Hospital, where a large firm mass in the left upper quadrant, extending to 7 cm below the costal margin, was felt. No abnormality was detected in the other systems.

The haemoglobin concentration was 13.8 g/100 ml, the white cell count was 13 000/μl and the eosinophil count was 520/μl. There was no haematuria and the results of renal function tests were normal. A Casoni skin test was not done, but the result of a hydatid complement fixation test was negative. A radiograph of the abdomen (Fig. 1) showed an enlarged left kidney. No calcification was seen. An intravenous pyelogram (Fig. 2) showed a large intrarenal mass. This was confirmed by arteriography, which also showed that the mass was avascular. Ultrasound evaluation suggested that the mass was probably a hydatid cyst.

At operation a very large cyst, measuring 12 × 10 cm, was seen to involve almost the entire left kidney, and a nephrectomy was performed. The postoperative course was uncomplicated, and the patient was discharged in good health.

**Fig. 1.** A plain radiograph of the abdomen, showing an enlarged left kidney. No calcification is shown.

**Fig. 2.** An intravenous pyelogram, showing marked upward displacement of the upper calyx of the left kidney, as well as the upper portion of the ureter.

**Fig. 3.** The cut surface of the kidney and the very large cyst are shown. The contents of the cyst, the ecto- and endocyst, are seen on the right.

**DISCUSSION**

*Echinococcus* is a genus of cestodes, two species of which, *Echinococcus granulosus* and *E. multilocularis* are known to affect man as an intermediate host. The former is
responsible for the classic unilocular hydatid cyst. Man becomes infested by the ingestion of the ova of the adult worm which is harboured by the dog, the definitive host. Most of the larvae migrate to the liver by way of the portal system, and only approximately 20% reach the left side of the heart, and thence the various organs in the body.

The wall of a hydatid cyst is composed of three layers. The inner layer, called the endocyst, is the germinal layer, and from it brood capsules develop. The ectocyst is a non-nucleated laminated layer, and is surrounded by an adventitial layer which represents the host's response to the cyst. A renal hydatid cyst is 'closed' if it is covered by all three layers, 'exposed' when it is no longer covered by the adventitial layer, and 'communicating' when there is rupture of all the layers, allowing free communication between the cyst and the calyces or pelvis of the kidney. Renal echinococcosis usually presents late, because the cyst is slow-growing and produces few symptoms. The most common modes of presentation are a loin mass, haematuria, renal colic and hydatiduria. Of the 12 patients reported by Deliveliotis et al., only 2 patients presented with renal colic, which was associated with haematuria in the one patient and hydatiduria in the other. One other patient presented with an unexplained urticaria. Of the 5 patients reported by Makki, 4 presented with an abdominal mass, and the fifth presented with intermittent renal colic. Only 1 of our 2 patients was aware of an abdominal mass.

The investigations which may aid in the diagnosis of renal echinococcosis include an eosinophil count, the skin test, various serological tests, radiological investigations and ultrasonography. A significant eosinophilia is uncommon, and only 1 of the 12 patients reported by Deliveliotis et al. had an eosinophilia of more than 10%. Auldist and Myers, who documented hydatid disease involving various organs in 114 children, recorded an eosinophil count of more than 300/µl in 71% of their patients. The Casoni skin test, which has been in use for over 60 years, has been shown to have several limitations. A positive skin reaction does not necessarily indicate active infection, since the skin sensitivity may persist for life in some patients. The incidence of positivity has varied in the reports of different authors. Kagan suggests that this may be attributable to the lack of standardization of the test. False-positive and false-negative results have been described as well. Deliveliotis et al. found a 100% incidence of positivity in their patients with renal echinococcosis. Of the serological methods described, the complement fixation and indirect haemagglutination tests are the most widely used. Arabatzis and Papapanagiotou studied sera from 120 patients with echinococcosis and 150 control patients, and found a high titre (more than 1:640) in 90% of their patients with the indirect haemagglutination test and in 60% with the complement fixation test. Three of the control sera (2%) produced a measurable titre (1:160 to 1:320) with the indirect haemagglutination test and 4 (2.7%) with the complement fixation test. The same authors found that when the indirect haemagglutination test was used in combination with the Casoni skin test, 92% of their patients had positive reactions. However, Slim et al. performed the skin test and both serological tests on 8 of their patients (all under the age of 15 years), and found that only 2 patients had positive reactions to all three tests. They suggest that the humoral reaction to echinococcosis appears late, and that the antibody mechanism may be different in children.

In any patient with suspected echinococcosis, the skin test and the serological test should be performed. A positive reaction will be significant, but a negative reaction does not exclude the disease. Radiological investigations which may aid in confirming the diagnosis of renal echinococcosis include a straight radiograph of the abdomen. If calcification is present the cyst will produce a characteristic picture, but if it is absent, the cyst may be indistinguishable from other renal tumours. An intravenous pyelogram and angiographic studies may also prove useful in making the diagnosis. With the latter one may be able to determine the extent of the disease as well, which is important when there is more than one cyst. Ultrasound, which proved useful in our second patient, is another investigation which may be performed.

Treatment consists of nephrectomy in most cases. Cystectomy may be impossible, because of difficulty in finding a plane of cleavage between the cyst and the kidney. Before removal is attempted, it is suggested that the cyst be sterilized by the injection of a 10% solution of formalin (i.e. 4% formaldehyde in water).

Renal echinococcosis is not a common disease. We feel that awareness of the condition is important, because the symptomatology, clinical features and radiological appearance may closely resemble those of malignant tumours of the kidney on the one hand, and benign lesions on the other.

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REFERENCES