Retroperitoneal Teratomata

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A TERATOMA is a true tumour or neoplasm composed of multiple tissues of kinds foreign to the part in which it arises (Willis 1962). It is an uncommon lesion and Palumbo et al (1949) could find only 58 "bona fide" cases of primary retroperitoneal teratomas in the literature up to 1949.

Case Report

A twelve day old male Indian infant was admitted to the General Hospital, Kuala Lumpur, Malaysia, on 14th March, 1972 with the complaint that the parents had noticed a swelling in the right side of the abdomen since the birth of the child. The swelling had remained of the same size. The birth was normal and full term. The child had an older sibling which also had a normal and full term birth, and was healthy. Apart from the abdominal swelling, the patient did not have any other complaints.

On examination, the child was normally developed for his age. There was an oval swelling in the right lumbar region, 8 by 10 centimetres in dimensions, firm and smooth. The swelling was easily balotable and did not move with respiration.

Plain radiograph of the abdomen showed a swelling in the right side of the abdomen with opacities which appeared to be rudimentary limb bones (Fig. 1 and 2). A diagnosis of abdominal teratoma was made.

Laparotomy was performed on 30th March, 1972 and this confirmed the presence of a large cystic right-sided retroperitoneal swelling which has pushed the liver upwards and medially (Fig. 3.) The swelling was easily excised. It was well encapsulated, cystic and had an umbilical-cord-like structure at one part (Fig. 4). When the tumour was excised it was found to contain a brownish clear fluid, hairs, well-formed bones, cartilage, muscle and other tissues (Fig. 5).

The histological examination of the specimen confirmed that it was a benign cystic teratoma with organoid formation.

Post-operatively, the child recovered well and was discharged on 11th April, 1972 in good condition.

Discussion

The main sites of teratoma (Willis 1962) are, in order of frequency, the ovaries, the testes, the anterior mediastinum, the retroperitoneum, the presacral region and the coccygeal region. Rarer sites are the base of the skull, the pineal gland, the brain and the neck. Teratomata of viscera (other than the gonads) and of the skull vault, posterior mediastinum, body walls and limbs are all very rare.

Teratomata have occured in the liver (KIRYABWIRE and MUGERWA 1967), the rectum (NIGAM, 1947; EL-KATIB 1972), the stomach (PAUL et al 1962) and the Fallopian tube (GRAY and HITCHCOCK, 1969). Teratomata are commoner in females and GROSS et al (1951) found that out of 40 cases, 32 were in females. It is thought that the female ovary differentiates later than the male testis so that females are more susceptible to developmental defects including teratomata.

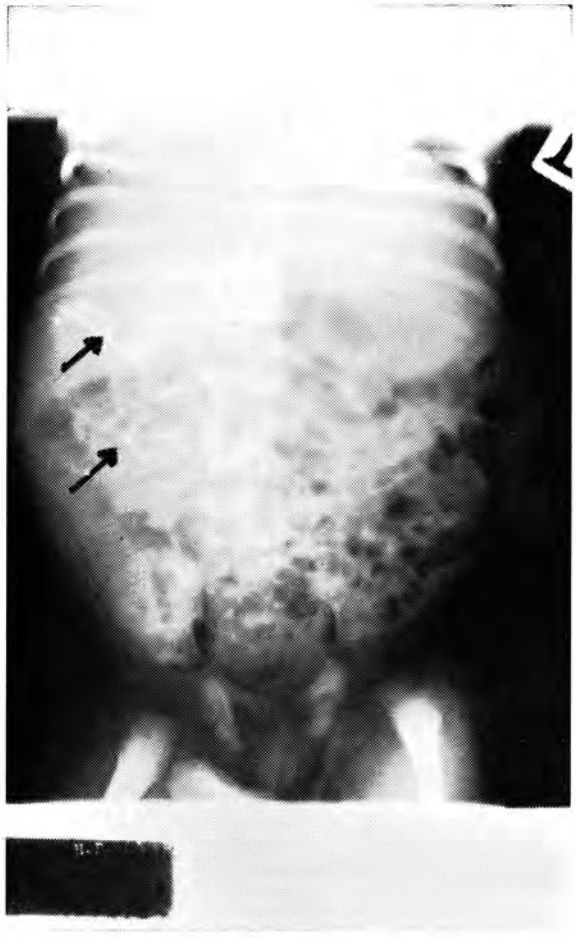


Figure 1
Plain postero-anterior abdominal radiograph. Arrows indicate bony clacification in rudimentary limbs.

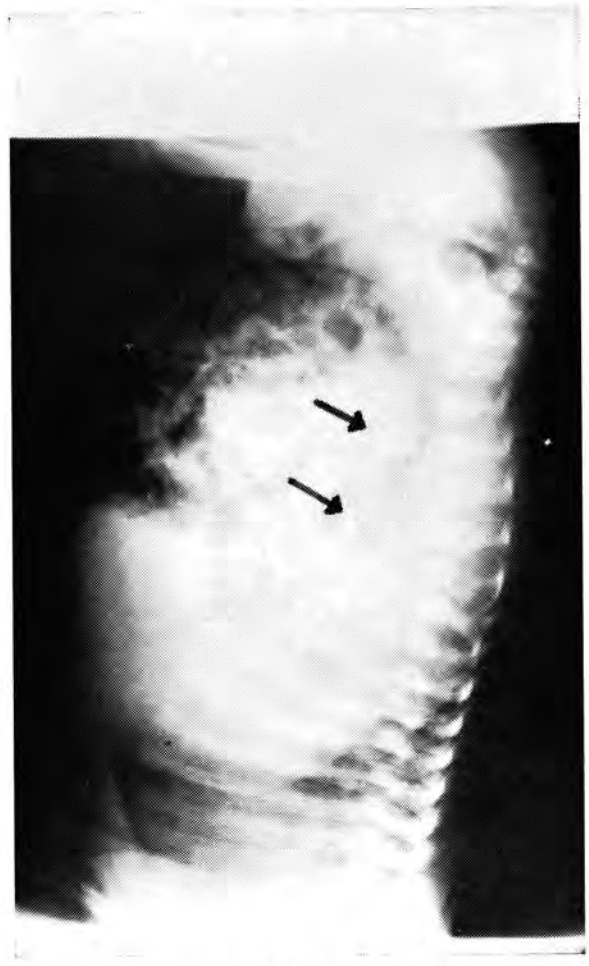
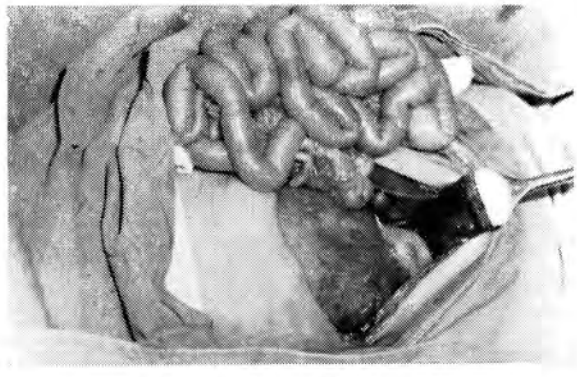


Figure 2
Plain right-lateral abdominal radiograph. Arrows indicate bony calcification in rudimentary limbs.



Fgirue 3
Operative findings. A large cystic right-sided retroperitoneal swelling is pushing the liver upwards and medially.

Figure 4
The uncut specimen. Note the umbilical-cord-like structure at one part.

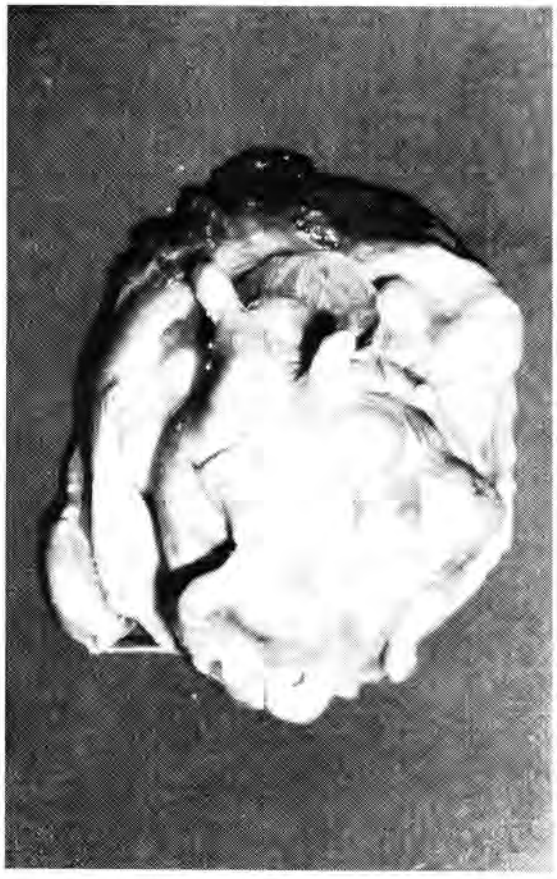


Figure 5
The cut specimen. Note the bony and cartilaginous projections amidst the soft tissue.

Most, and probably all, of the retroperitoneal teratoma are present at birth (Willis 1962). Most teratomata — gonadal, retroperitoneal, sacrococcygeal and cranial — arise in tissues which developmentally occupy pre-axial median or closely paramedian positions. This distribution strongly suggests disturbances occurring in the embryonic axial structures — the primitive streak, the head process and the notochord — as causing teratomata (Budde 1926).

The most widely accepted theory of origin of teratomata is that of WILLIS (1951) who thought that they arose from foci of toti-potent cells which had escaped the influence of the primary organiser during embryonic development.

Teratomata may be mainly crystic or completely solid and both types may be intermixed. The cysts are usually multiple and contain either sebaceous matter or clear fluid. Teratomata are tridermal structures and often contain derivatives of ectoderm (skin, teeth, nerve tissue), endoderm (alimentary and respiratory epithelia), and mesoderm (vascular and connective tissue).

A very important group of neoplasms in infancy and childhood are those in the retroperitoneal space. The two commonest ones are Wilm's tumour (embryoma of the kidney) and neuroblastoma. Retroperitoneal tumours are the third commonest of this group of retroperitoneal tumours although they are much rarer than the Wilm's tumour and neuroblastoma (Gross, 1953). Before operation it is often difficult to tell which of these three tumours is present. However, certain features are helpful in the differential diagnosis. Wilm's tumour and neuroblastoma are much commoner than retroperitoneal teratoma. The first two tumours are more liable to present at the end of the first year of life or after this time, with a peak incidence at between the second and fourth years of life. Retroperitoneal teratoma is discovered most commonly in the first

year of life, especially the early months after birth. A neuroblastoma usually has a finely nodular surface and its borders are ill-defined — such features are less often present in a teratoma. Plain abdominal radiographs rarely show calcification in Wilm's tumour but this is often present in teratoma and neuroblastoma. If the radiograph shows tooth or bone, then the lesion is a teratoma. A pyelogram of the urinary tract which shows great distortion of the renal pelvis and calyces is more suggestive of Wilm's tumour. If metastases occur, secondaries limited largely to the lungs suggest Wilm's tumour whilst multiple destructive deposits in the long bones and skull suggest a neuroblastoma.

Teratoma range from entirely benign structures consisting of mature tissue with no signs of proliferative activity to malignant growths which involve all or only a segment of the specimen. Because of the danger of malignancy and of other complications such as haemorrhage, pressure effects on adjacent vital structures and infection, the treatment of teratomata is surgical removal. The approach should always be transperitoneal by way of an abdominal incision. This will allow of maximum access to the lesion and enable it to be dissected away from surrounding important structures such as duodenum, kidney, aorta and inferior vena cava. If malignancy is present in the specimen, post-operative irradiation is given to the tumour site and its surrounds. The prognosis depends on the degree of malignancy of the tumour and if benign lesions are completely excised, complete cure of the patient results.

Summary

Teratomata are rare developmental tumours and amongst the rarest of these are the retroperitoneal teratomata. They vary from benign to highly malignant lesions. Treatment is by surgical excision followed by post-operative irradiation if the tumour is malignant.

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