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Fetus *in fetu* and Teratoma

A CASE REPORT AND REVIEW

J. P. G. DU PLESSIS, W. S. WINSHIP, J. D. L. KIRSTEIN

SUMMARY

A case is reported in which a fetus *in fetu* and a malignant teratoma were present within the same intra-abdominal mass in a 6-month-old male infant. It is the first record of such an occurrence, and attention is drawn to the possible significance of this case, in view of the now rejected concept that a teratoma is the result of an abnormal process of twinning.

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A teratoma may be defined as a tumour made up of recognisable somatic tissues corresponding to either embryonic, fetal or adult stages of development foreign to the part in which it arises.[1] The term fetus *in fetu*, on the other hand, refers to the presence of a parasitic twin within its bearer. Its diagnosis depends on the clear-cut demonstration of an axial skeleton and an appropriate regional organogenesis—criteria first defined by Willis.[2,3]

CASE REPORT

We have recently encountered an infant who presented with an intra-abdominal mass arising from the upper retroperitoneal space, containing as adjacent but distinct entities, a fetus *in fetu* and a teratoma. The purpose of this report is to document the first recorded case of the coexistence of these two conditions.

History

A 6-month-old male infant was transferred from an outlying hospital with a history of vomiting and abdominal distension. The birthweight was 3,6 kg, following a normal pregnancy. Weight on admission was 5,5 kg.

The infant started vomiting bile-stained fluid at 2 weeks of age, and required intravenous rehydration. Vomiting had continued intermittently up to admission and the child failed to thrive. Three normal siblings were well, and there was no history of multiple pregnancies in the immediate family.

Department of Radiology, University of Natal, and Department of Paediatrics and Paediatric Surgery, Addington Hospital, Durban

J. P. G. DU PLESSIS, M.B. CH.B., D.M.R.D. (Present address: Eagle Bldg, Murchie's Passage, West Street, Durban).
W. S. WINSHIP, M.B. CH.B., M.MED. (PAED.)
J. D. L. KIRSTEIN, M.B. B.CH., F.R.F.P.S., F.R.C.S.

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Examination

The child was poorly nourished, with marked abdominal distension. Abdominal palpation revealed a large, irregular, craggy mass.

An emergency excretory urogram to exclude a Wilm's tumour revealed a normal urinary tract, but the plain film of the abdomen showed a central abdominal mass displacing the stomach upwards, and the bowel peripherally. An anencephalic fetus was present anteriorly in this mass (Fig. 1), displaying a well-formed vertebral column, and an appropriately arranged rib cage and femora. In addition, the mass contained calcific plaques and flecks, in a position above and posterior to the fetus.

Operative Findings

In view of persistent vomiting, removal of the mass was undertaken as an emergency procedure. Through a 15-cm, transverse, supra-umbilical incision, an enormous, partly cystic, partly solid tumour was delivered. The duodenum was stretched over and closely adherent to the tumour, and dissection of this region presented some difficulty. The tumour arose posteriorly from the upper retroperitoneal tissues.

Pathology

The tumour was removed *en masse*, but removal posteriorly and superiorly was incomplete. The specimen weighed 1 kg, i.e. 18% of the patient's weight.

The main mass contained an anencephalic fetus with rudimentary limbs and male external genitalia. Large areas of the fetus were covered by vernix-like material (Fig. 2), and it lay in fluid surrounded by an intact capsule, to which it was attached by a cord.

The vertebral axis of the parasitic fetus, with an appropriate arrangement of limbs, penis and scrotum, is demonstrated in Fig. 3, and proper organogenesis is shown histologically in a section of gut in Fig. 4.

The macroscopic appearance of this specimen conforms with the description of fetus *in fetu* given by Lewis.[4]

The smaller portion of the mass consisted of alternating solid and cystic areas (Fig. 5). Histology of these areas indicated a malignant teratoma (Fig. 6), the features of which extended up to the line of resection.

A detailed histological analysis is to form the subject of a later report.

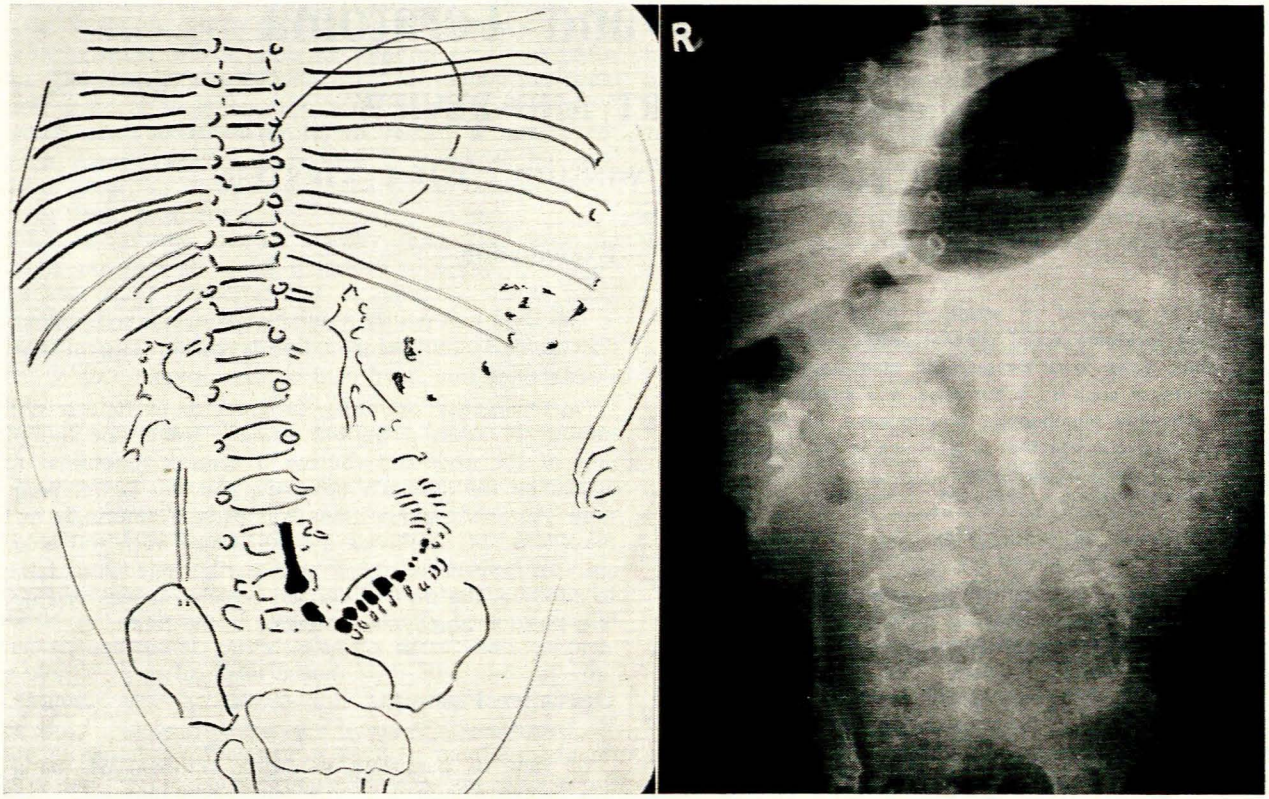

Fig. 1. Excretory urogram and line drawing, showing normal renal tracts, with an anencephalic fetal skeleton in the lower abdomen and calcific plaques in the left upper quadrant.

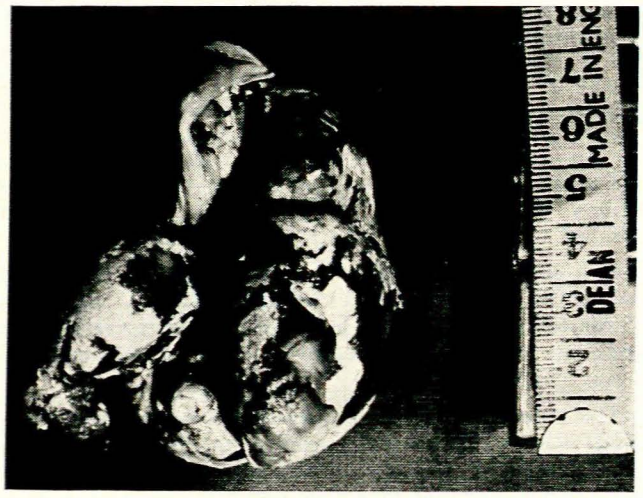

Fig. 2. The parasitic fetus, illustrating anencephaly, vernix, rudimentary limbs and toes, and male external genitalia.

DISCUSSION

Lord,[5] in 1956, accepted 11 previously recorded cases for her authoritative review of fetus *in fetu*, including 1 of her own and 3 from the previous century. Even so, she accepted only 4 as 'proven', but some of her reservations about the remaining cases appear to be on technical grounds only (e.g. inadequate description of the specimen, imperfect X-ray films). Since the 11 cases reviewed by Lord, a further 6 cases[4,6–10] have been recorded in the English literature, and 2 in Hanoi.[11] We have not had access to the latter descriptions.

Teratomas, of course, are encountered far more frequently in man. Palumbo[12] reviewed some 58 cases of retroperitoneal teratomas in 1949, and Arnheim[13] in 1951 analysed 39 cases in children. Engel *et al.*[1] traced a subsequent 29 case reports, and added 1 of their own.

Kimmel *et al.*[14] described a case of a hydrocephalic infant, delivered by Caesarean section, whose brain contained a teratoma, with 5 fetiform structures. The latter have not been accepted as examples of fetus *in fetu*.

Previous writers have envisaged teratomas as resulting from a modified process of twinning, and have traced a natural progression from normal twins to conjoined symmetrical twins, through parasitic fetuses and fetal inclusions, and leading, finally, to teratomas.[15]

Recent authors, following Willis's publications, have emphatically denied a relationship between the two conditions, and have regarded fetus *in fetu* as a monozygotic twin of its bearer, and a teratoma as a true, *de novo*, tumour.

Willis's experience was based on the meticulous analysis of multiple thin sections of 13 personally encountered

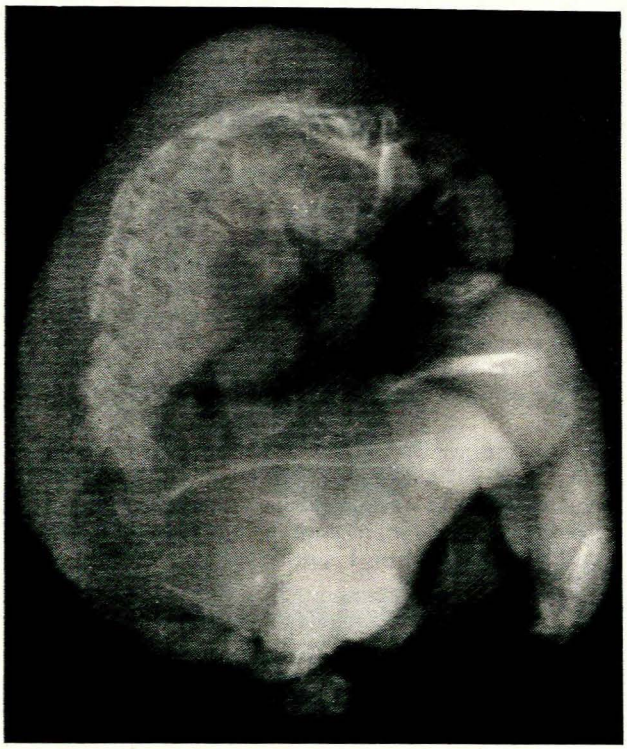

Fig. 3. Radiograph of the fetus, illustrating an appropriate regional organogenesis (limbs and genitalia), in relation to the axial skeleton.

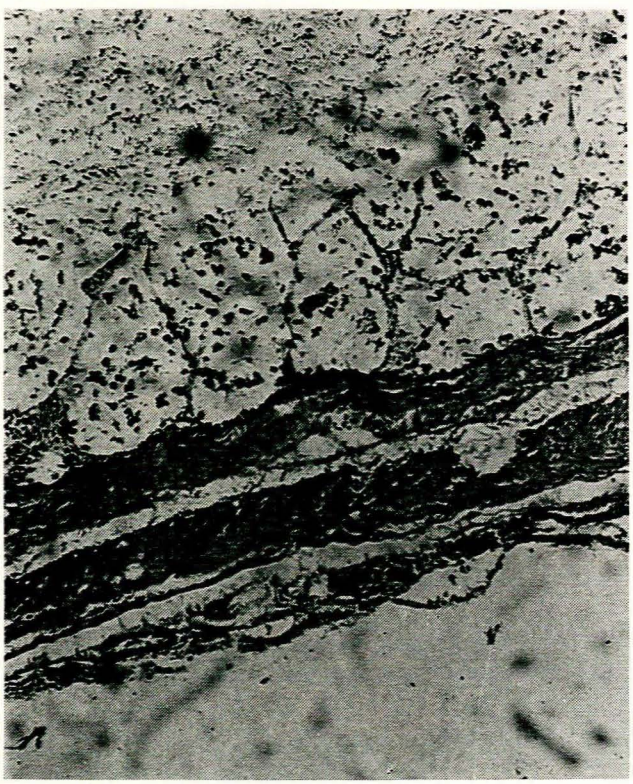

Fig. 4. Histological section of gut. From above downwards: autolysed mucosa, submucosa, muscularis externa and serosa.

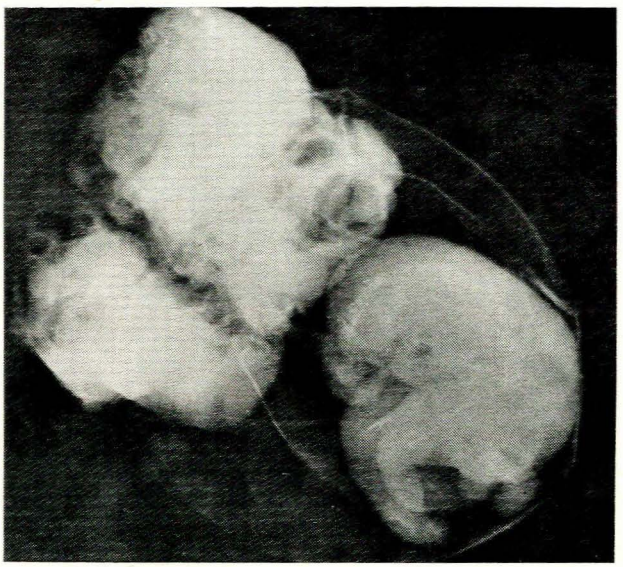

Fig. 5. Radiograph of the specimen, after drainage of the amniotic fluid, showing the solid and cystic nature of the teratoma, with areas of calcification on the left. On the right, the fetus is seen enveloped in its membranes.

teratomas. He describes these tumours as 'containing masses of central nervous tissue, but no brain, masses of bone, but no bones, respiratory or intestinal cysts or canals, but no proper trachea or intestinal tract, renal tissue, but no kidney'.[2]

He cites the following in rejecting the concept that a teratoma is a distorted fetus:

1. Teratomas exhibit no signs of axiation, metameric segmentation or delamination of germ layers, i.e. the earliest and most fundamental features of the formation of the soma are lacking.
2. Teratomas possess no organs or true somatic regions.
3. The multiplicity of certain constituents in teratomas would imply that, if fetal in origin, the entodermal or dental rudiments of the fetus have suffered some unparalleled kind of developmental subdivision and separation. He finds it hard to imagine a process in a fetus so disruptive as to produce the picture of a teratoma.
4. There is an anomalous absence of particular tissue in certain teratomas, e.g. skin.
5. Abnormal tissue relationship and mixtures of tissue types are frequent in teratomas.

Elsewhere[3] he supports his arguments by stating that no case of a parasitic twin showing powers of progressive neoplastic growth has ever been recorded.

Spemann[16] first postulated that the influence of a 'primary organiser' determined the orderly growth of fetal tissue, and Willis takes the view that teratomas arise from foci of multipotential embryonic tissue which, for some unknown reason, have escaped this influence.

As a result of the work of Willis, almost all writers[4-10,17] over the past 25 years or so, have echoed his rejection of the concept of a relationship between teratoma and fetus *in fetu*.

The only exceptions appear to be Gross and Clatworthy,[18] who, in reporting a case of twin fetus *in fetu*, regard the latter as a highly developed or complex form of teratoma.

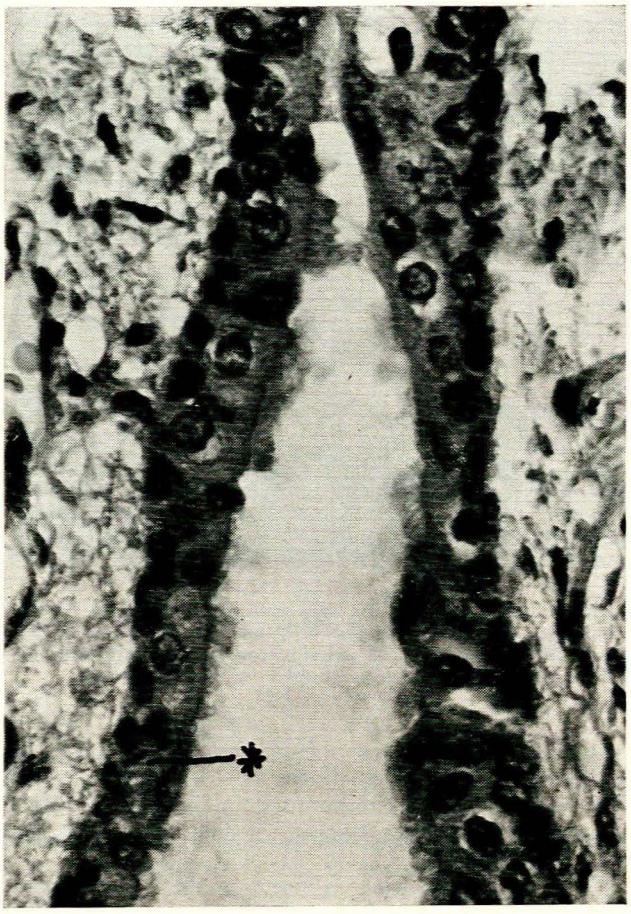

Fig 6. Histological section of an epithelial-lined structure in the malignant teratoma. The epithelium is ciliated, and one mitotic figure is visible towards the bottom left of the photograph.

Interestingly, their first list of 18 references does not include the name of Willis, who had published his opinions 15 years earlier. They also state that 'a fetus *in fetu* is a form of teratoma, and should always be removed, along with its membranes, to ensure a cure'. Three years later, Lord[19] contradicted this by stating that 'removal of a fetus *in fetu* from within its sac is a safe procedure, and heroic surgery is not necessary'.

CONCLUSION

In presenting this case history, we suggest that the older theory, that a teratoma represents a disrupted fetus, may have been discarded prematurely. As difficult as Willis found it to imagine 'so disruptive a process in a fetus as to produce the picture of a teratoma', we submit that it is as difficult to imagine that the teratoma in this case had an aetiology which differed basically from that of the fetus *in fetu*.

It therefore seems reasonable to suggest the intriguing possibility in this case of a potential triplet situation gone awry, resulting in the host, his parasitic twin, and a teratoma arising from a third embryo which may have escaped the influence of a 'primary organiser'.

We wish to thank Dr M. Suskind for the pathological studies.

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