FOETUS IN FOETU – A CASE REPORT

H Ng, K P Tan

ABSTRACT

A three-month old baby presented to the Paediatric Department with a problem of abdominal distension. Clinically, he had a large right abdominal mass which on investigations suggested a teratomatous lesion. At operation, a foetus-like tumour mass was located in the retroperitoneal space. A diagnosis of foetus in foetu was made. A close differential of a retroperitoneal teratoma is discussed and comparison with confirmed cases made.

Keywords: foetus in foetu, induced twin, vertebral axial skeleton, retroperitoneal teratoma

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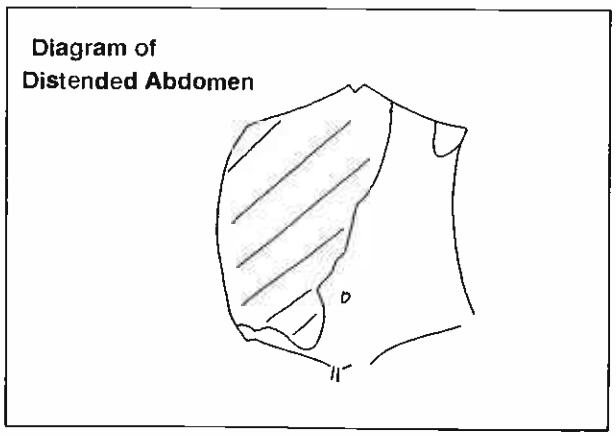
INTRODUCTION

Foetus in foetu as yet is an ill-defined class of which 42 alleged cases had been reported since 1956.

However, none were sufficiently studied to prove that it was in fact vertebrate. Hence a majority of reported "foetal inclusions" were invertebrate teratomas. Proof of the genuine "included twin" requires at least unequivocal radiographic or dissectional demonstration in part or whole of a vertebral axial skeleton, a demonstration which will be reinforced if other appropriately situated bones or organs are shown to be present (1).

CASE REPORT

MH is the second in the family of two. He had a full-term normal vaginal delivery with a birth weight of 4 lbs 7 ozs. His neonatal period was uneventful except for neonatal jaundice which was treated with phototherapy. At 3 months of age, his parents noticed that his abdomen was getting distended. He had no feeding problems or vomiting. His bowel habits were normal. There was no history of dysuria or jaundice. His milestones up to time first seen were within normal.



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Clinically he appeared healthy and was active with no evidence of pallor or oedema. His vital signs were normal. There were several small cervical nodes on both sides of the neck.

Abdomen was distended over the right hypochondrium with prominent superficial veins. There was a large mass approximately 9 × 12 cm arising from the RHC towards the right iliac fossa (RIF) just crossing the midline. The mass was firm and nodular over some areas, relatively mobile and non-tender. No bruit was heard over the lesion. There was another rounded mass just inferior to the main leison. The spleen was 1.5 cm palpable below the left costal margin. Both testes were felt within the scrotum. Initial impression was possibly Neuroblastoma or Hepatoblastoma. Blood investigations were unremarkable. Plain Abdominal radiograph (Fig. 1) showed fullness noted over the right abdomen with a large soft tissue mass extending from RHC to RIF, pushing the bowel loops to the left. It crossed the midline and had well defined margins. Frond-like irregu-

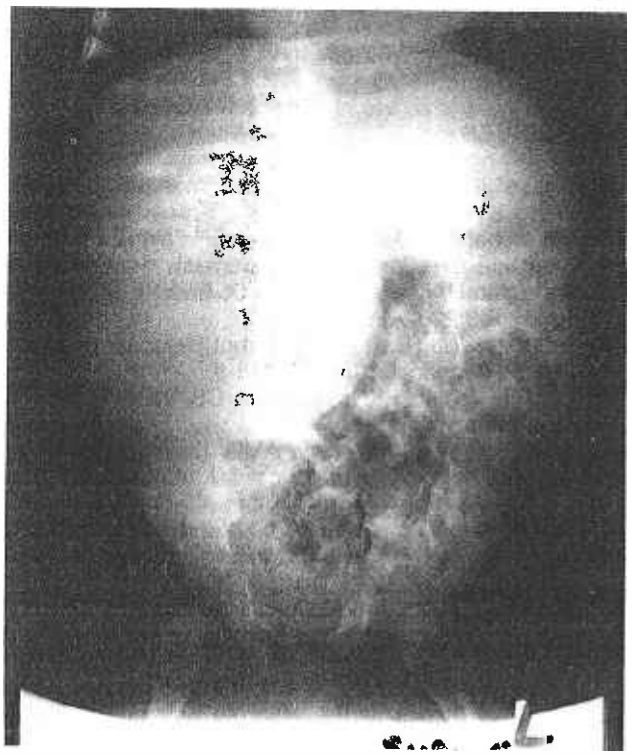

Figure 1 –
Plain radiograph showing a soft tissue mass (> >) pushing the bowel shadows to the left with calcifications within. (⟶)

lar calcifications were seen within the mass with areas which are slightly more radiolucent than the surrounding soft tissue of the mass. An ultrasound examination (Fig 2, 3) revealed a large mass measuring approximately 10 cm in diameter below the liver and separate from it. The mass was composed of highly echogenic parts with acoustic shadowing posteriorly and hypoechoic areas anteriorly. However several strong echoes were seen within these too. The highly echogenic region indicated presence of calcification while the rest probably contained large amounts of fatty tissues. Unfortunately, the right kidney was not identified with certainty. Features seem to suggest a benign lesion like a teratoma. However neuroblastoma was still possible. An Intravenous Urogram (Fig. 4) reported a normal left kidney. However, the right kidney was not visualized. Tomograms of the mass confirmed presence of Adipose tissue as there were several radiolucent patches within the mass. In view of the results, a CT scan (Fig 5, 6, 7) was then performed. A very large retroperitoneal lesion was seen pushing the liver towards the right and the right kidney caudally. It had various components like fat, bone and soft tissue. The right kidney appeared normal. A provisional diagnosis of a retroperitoneal teratoma was made.

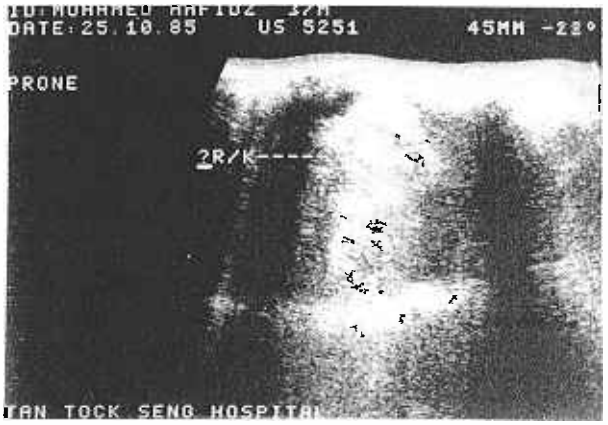

Figure 2 –
Ultrasound of the Right side posteriorly with the patient prone in the region of the renal angle showing echogenic as well as hypoechoic areas.

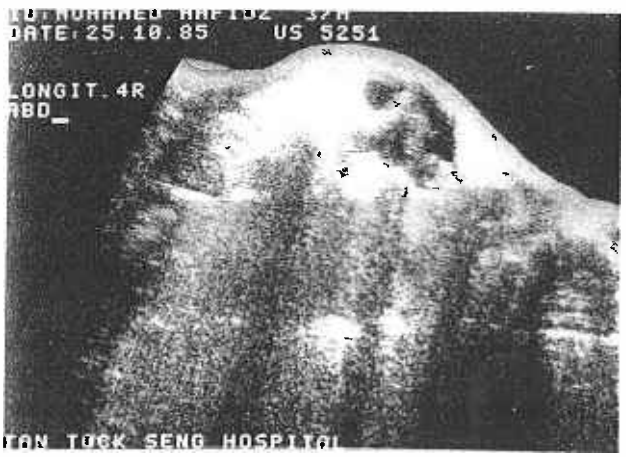

Figure 3 –
Ultrasound of the Right hypochondrium shows some densely echogenic foci with acoustic shadowing (> <) posteriorly.

At operation, a cystic mass 15 x 15 x 15 cm was found in the right abdomen, displacing the liver to the right and the kidney latero-inferiorly. The right adrenal and right renal vessels were enclosed. The adrenal had to be sacrificed, so was the genito-femoral nerve and right inferior phrenic artery. Both right kidney and adrenal were normal. Unfortunately, no mention of a membraneous sac or point of attachment was made in the operative notes.

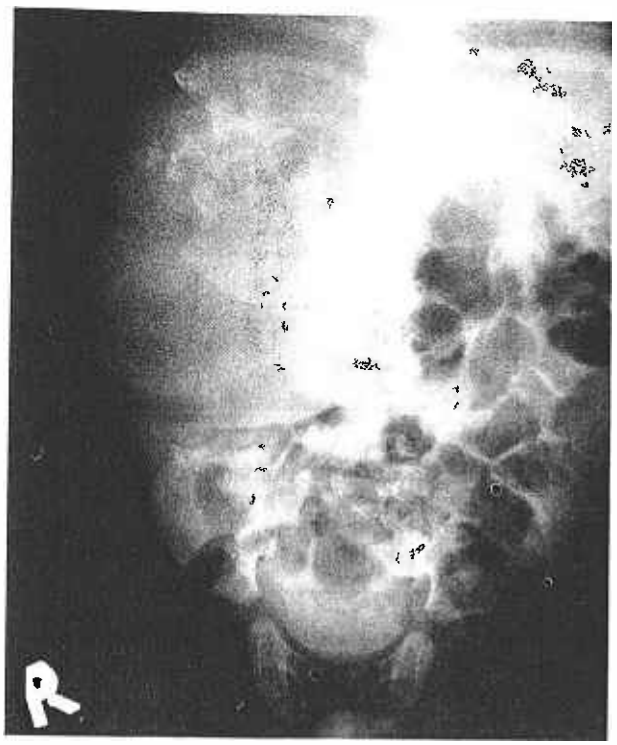

Figure 4 –
5 minute-radiograph of an intravenous urogram showing the calcification (> >) and the slightly lucent areas below and around it representing fat. The normal left collecting system (⟶) is seen.

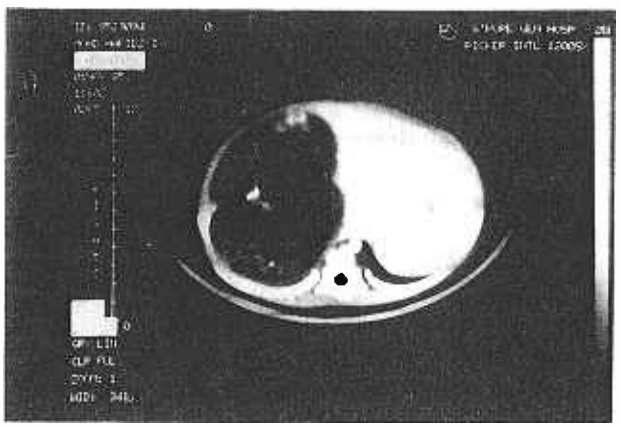

Figure 5 –
C T scan with intravenous contrast shows low attenuation contents which represents fat with some dense specks which appear bony.

Macroscopically the tumour mass had scalp with hair, limbs with fine hair follicles and even a breast. Unfortunately, no gross or microscopic section to study the detail anatomy was done and the specimen was potted whole.

The radiograph of fresh specimen (Fig. 8) revealed a distorted lump of nodular tissue with bones distributed in clusters all over. Radiographs and gross appearance of the Potted Specimen (Fig. 9-13) showed a suggestion of a limb bone probably a femur with part of the pelvic bone attached. There are 2 clusters of

bony densities which may resemble vertebral bodies though not too convincingly. The central ones seen both antero-posteriorly and in the lateral view appears slightly more convincing. Other smaller limb bones with digital appendages are seen.

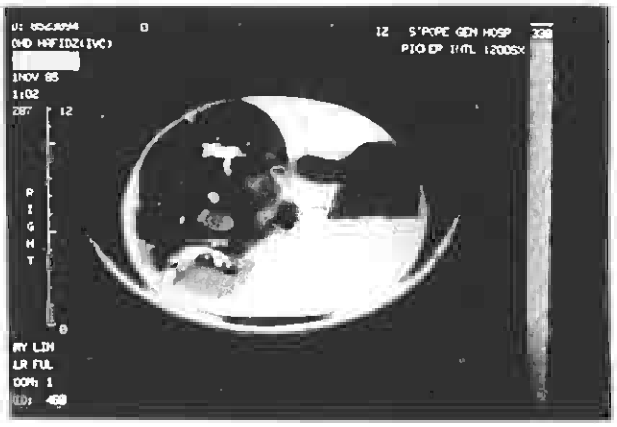

Figure 6 –
Sections lower down the abdomen shows calcification (⟶) which appear to be aligned and soft tissue components (> >) within.
The stomach with contrast is seen on the left.

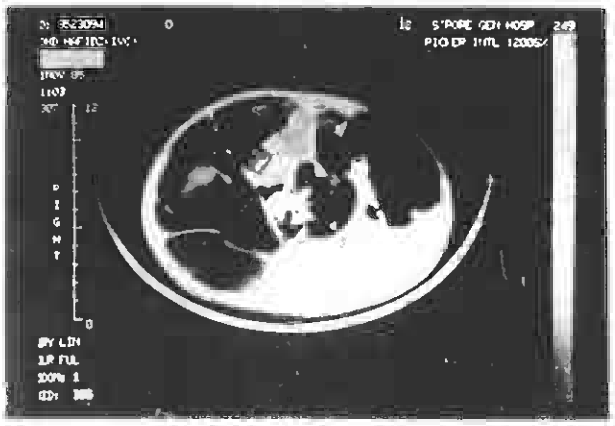

Figure 7 –
Lower sections showing locculation and loops of bowel.

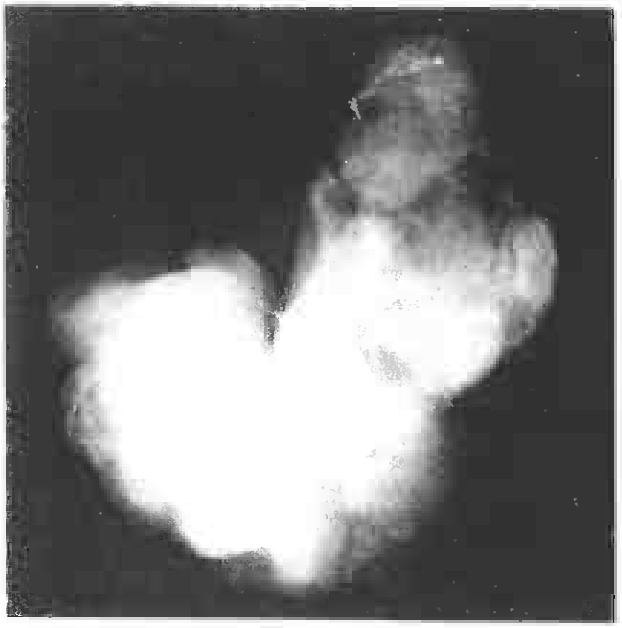

Figure 8 –
X-ray of fresh specimen from patient.

The patient recovered and was discharged. He did not return for follow-up since.

Figure 9 –
View of the same showing a limb with digits (⟶) and a breast nodule (>). Note the lanugo hair on the skin.

Figure 10 –
This view suggests a flexed lower limb with a thigh and foot.

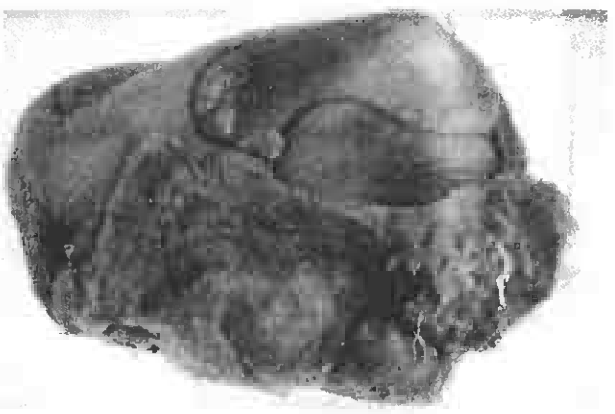

Figure 11 –
Scalp hair can be seen.

Figure 12 –
Radiograph of the specimen reveals bony elements which resemble a pelvis with the attached femur. (⟶) The collection of bones to the left (>) could represent a possible vertebrae.

Figure 13 –
A right angle view compared to Fig 12 showing some limb bones. The central structure (⟶) could be a sternum or vestigial spine.

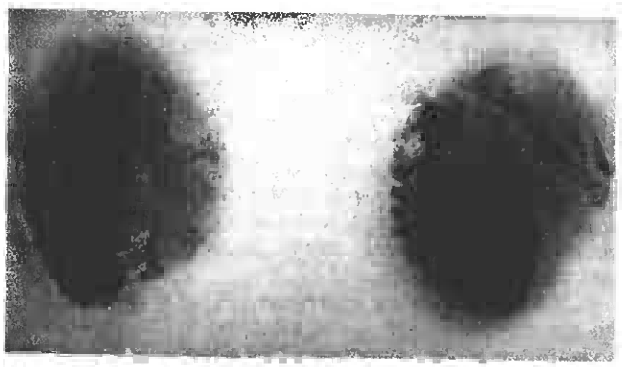

Figure 14 – Author's own case with radiograph of surgical specimen.

DISCUSSION

Foetus in foetu is a descriptive term attributed to Meckel (Circa 1800) limited only to those rare cases where a parasitic twin is found included within the abdomen of its partner. The first recognizable case was described by Dickinson in 1871. Since then 87 cases have been reported in literature up to the 1960's. Until the 1920's, teratomas were generally believed to result from the attempted formation of a new human and the term "foetus in foetu" was used to describe the apparent external foetiformity.

However, Nicholson (1934, 1937) [2] and Willis (1935, 1945) [3] reasoned that teratomas could not have passed through the primitive streak stage which endows the developing organism with its fundamental vertebrate pattern and axialization. This point has been ignored by every subsequent writer who has published a description of a specimen seemingly having a bony longitudinal axis. No attempt has been made to study the bony axis. This may prove to be so in this case as histological proof was not available. Hence foetus in foetu may be defined as a vertebrate foetus included within the abdomen of its partner. Whereas a teratoma is a true tumour or neoplasm composed of multiple tissues of kinds foreign to the part which it arises. Many theories have been suggested, ranging from a form of included twin to that of a very well differentiated retroperitoneal teratoma. However, Willis (1958) [3] has pointed out that these are two very separate entities. He emphasized that retroperitoneal teratoma is a true tumour while foetus in foetu is not, though terms such as "parasitic twin" and "suppressed twin" have been misapplied to teratomas having a marked degree of differentiation.

Josephine M Lord [1] suggests that proof of the genuinely foetal nature of presumed intra-abdominal foetus in foetu requires at least – the unequivocal radiographic or dissectional demonstration of part or whole of a vertebral axial skeleton. Hence by 1956, she found 31 reports of alleged foetus in foetu (before 1900 and since 1900, there were 11 more cases). There were only 4 deemed genuine cases by her. 2 were post-surgical while the other 2 were post-mortem specimens. These were, first the author's own case (see Fig. 14) removed from a 7-week old boy. The pedunculated tumour contained within a thin-walled sac in the region of the lesser sac was attached to the retroperitoneal tissues near the lower pole of the left kidney. X-rays showed a well developed spine-series of vertebrae with characteristic cranio-caudal differentiation. The thoracic part showed ribs on either side but limb bones, skull and the cervical part of the spine were absent.

The second is a surgeon's, H Morton Anderson's case (Fig 15) which was surgically removed in a girl of 5 weeks (personal communication, 1950; Davis (1939)) [1]. The specimen was later destroyed in the bombing of London. However, x-ray photograph proved that this was a vertebrate foetus possessing a skull with craniolacunae, a shoulder, pelvic girdle and both upper and lower limb bones.

The third case by Highmore (1815) (Fig. 16) [1] – An autopsy finding of a sac attached to the mesentery near the spine and pancreas. The patient was a boy of 15 who died of bleeding – massive haematemesis and had passed blood and fleshy material per rectum for 3 months before death. The duodenum and jejunum were incorporated as part of the sac wall with numerous

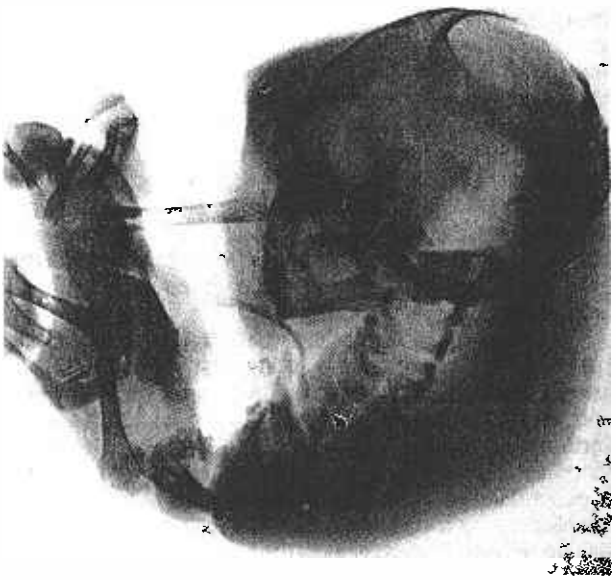

Figure 15 —
Mr. H Morton Anderson's case with x-ray of surgical specimen.

large blood vessels attached to the bowel. One of these had ruptured and most likely the cause of death. The specimen was a macerated foetus with no head and only 3 extremities. There was a much curved spine.

The fourth case was Seymour Taylor's (Fig. 17) [1]. At necropsy, a tumour was found behind the stomach over the first and second lumbar vertebrae. He found the tumour to be covered by loose fatty membrane. A vertical section revealed a rudimentary vertebral column with a longitudinal groove containing what represented a spinal cord. Rudimentary limbs were recognised too.

There has been no mention of the incidence so far. The retroperitoneal teratoma, its close mimic, is recognised in only 10% of all retroperitoneal primary neoplasms. All reported cases of foetus in foetu were noted in the first 12 months of life or early infancy as in

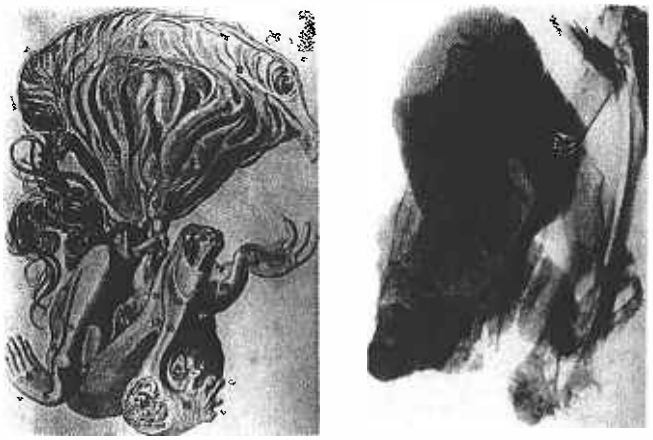

Figure 16 –
A. Highmore's case. Specimen removed at necropsy from a boy aged 15.5 years. Photograph of drawing by W Clift.
B. Same case with recent x-ray of the museum specimen.

our case. Compared to retroperitoneal tumours where only 50% manifest in early life, with an average time of diagnosis at thirteen years (Palumbo et al 1949). The upper part of the retroperitoneal space is the only site of occurrence of foetus in foetu. No case has been reported in the pelvis or in any abdominal organ. Previous writers have described the occurrence of foetal structures within the brain, abdominal wall and the uterus. A particular case is that of Kimmel, et al in 1950 [4] who reported a cerebal tumour allegedly containing five human foetuses. However, doubt exists whether these fully meet the requirements to classify as included quintuplets.

The foetus in foetu is usually suspended by a peduncle within a capsule containing a little fluid. The capsular wall is often thickened at the point of attachment of the peduncle with an associated plexus of vessels. The thickened part may be orientated towards the base of the mesentery and the origin of the superior mesenteric artery. Unfortunately for our patient, no mention was made of the attachment of the mass in operative notes. In all the cases studied, no direct connection of peduncular vessels with those of the host has been shown. Retroperitoneal teratomas are also commonly found in the upper retroperitoneal space, but twice more frequently on the left than right. It typically does not have a capsule or pedicle and the attachment to the posterior abdominal wall is broad and intimate.

The outward appearance of the foetus in foetu may range from an anencephalic globular or reniform mass with rudimentary limb buds, through all stages of development to the well-formed foetus. More than one part of a foetus should always be recognisable and the suspending cord or peduncle should also be identifiable.

Internally, a vertebral column at some stage of development will be found, together with other bones. A complete foetal skeleton may exist. Of the soft tissue structures, the intestinal tract is usually best represented. Various other organs may be recognisable including brain, spinal cord, gonads and adrenals. Although the demonstration of a vertebral column is important, in those cases where it is very dysplastic and underdeveloped, it may not be identifiable on a gross radiologic basis. A rudimentary cardiovascular system may be present but is not functional. Hence, the foetus in foetu is acardiac deriving its blood supply from its host. No trophoblastic or placental tissue has yet been described in the capsule.

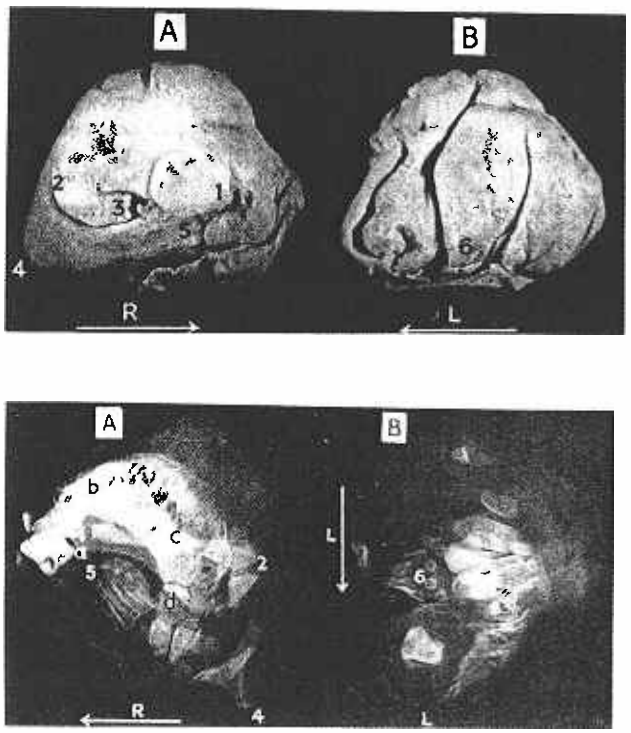

Figure 17 –
Taylor's case. Photograph of external appearances with accompanying radiographs.
A – Right portion
B – Left portion

GROWTH AND MALIGNANCY

Foetus in foetu commencing existence, as a true twin, grows initially in parallel with its fellow. Soon however, because of its anatomical confinement and perhaps some other factors which determine its parasitic role, it lags behind its host.

Although limbs of the included foetus may appear to be well developed, their ratio to the trunk lags that of a similar normal foetus. After attaining a variable size, further growth ceases and actual retrogression can occur as the host twin progresses. Willis (1958) [3] described in detail how reduction of a parasitic twin may be brought about, with loss of head, limbs and internal organs. It is uncertain whether the factor involved is the vascular dominance of the host twin or some inherent defect in its parasitic partner.

There is no evidence that foetus in foetu has neoplastic or maglignant propensities. Oppositely, retroperitoneal teratomas may be malignant.

PATHOGENESIS

Foetus in foetu is a monozygotic twin. Developing from a single ovum, it would, if not for the limitation in growth, which occurs later, be of the same sex and closely resemble its host.

Early cleavage of the dividing cell mass may so occur that each may be shared. Then it is likely that each foetus will have its own amniotic cavity. However, occasionally, twin embryos appear at a later stage and a common amniotic cavity is utilised. Willis (1958) [3] states that most if not all conjoined twins and double monsters are in this group. Hence it is possible to theorise that an even later cleavage into twins might result in inclusion of the second embryo within the body layers of its partner. However, no supporting evidence for this has been found.

TREATMENT

Surgical removal of the tumour should be carried out without undue delay. Cole and Gerrish (1951) [5] commented that procrastination in small infants results in no gain, as time goes on, the newborn infant becomes a poorer surgical risk. However, if the diagnosis is reasonably certain, removal of a foetus in foetu does not normally call for such urgency, in the absence of infective complications which might induce peritonitis or local adhesive reactions. Whereas, the potentially malignant nature of a teratoma or other similarly situated tumours when the diagnosis is doubtful calls for early operation.

CONCLUSION

Hence it can be seen that a diagnosis of foetus in foetu can be very difficult because some cases may not have well defined limb skeleton or the ossification of the vertebrae was not complete or dense enough to be seen. Another reason is that it could be all that remained of the interior of a markedly reduced foetus.

Hence it may be difficult to identify correctly borderline instances of these conditions unless very minute and painstaking examination of the fresh specimen is carried out. Most often, as Knox and Webb [6] pointed out, foetus in foetu is not diagnosed preoperatively, even though radiographic features retrospectively are usually quite typical. In our case, it has been assumed to be a foetus in foetu by the surgeon as well as pathologist based on outward appearance. Like in Nocera et al's case [7], although no formed bony elements were seen, such as identifiable radius, ulna or fermur, definite bony structures are present. The vertebrae which we do not see may suggest otherwise. The spinal column may be so vestigial that it cannot be recognised with certainty.

The disagreement regarding how to classify foetus in foetu and teratoma will persist, but since there is a complete difference in malignant potential, there does seem reason to try to differentiate the two. At the moment we feel our patient did have an included twin.

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