FOETUS IN FOETU AND THE RETROPERITONEAL TERATOMA

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Foetus in foetu, a descriptive term attributed to Meckel (*circa* 1800), should be applied only to those rare cases where a parasitic twin is found included within the abdomen of its partner. Lord (1954; 1956) reported two definite cases and reviewed other possible cases.

In contrast, the variety of teratoma which occurs in the retroperitoneal space is much less defined. Since the description of the first recognizable case by Dickinson (1871), an increasingly complex volume of literature has accumulated. Palumbo, Cross, Smith and Baronas (1949) documented many of the earliest cases of retroperitoneal teratoma and their work drew together some of the facts concerning a condition which they rightly say is seldom discussed in the medical reference volumes. Charles (1953) traced 82 case reports in the literature and since this time at least five further reports have appeared including some from Russia and Bulgaria. Many theories of origin have been suggested, with some authors contending that the teratoma represents a form of included twin. Although this is now unacceptable, it has resulted in the description of many retroperitoneal teratomas in confusing terms.

Willis (1958) pointed out the separate nature of the retroperitoneal teratoma and foetus in foetu, emphasizing that the former is a true tumour while the latter is not. However, as both conditions occur in the upper part of the retroperitoneal space, it is not surprising that terms such as 'foetus in foetu', 'parasitic twin' and 'suppressed twin' have been misapplied to teratomas having a marked degree of differentiation and some foetiform characteristic. Indeed, Willis concedes that final classification may be difficult, though he does not admit that the overlap of certain features implies that the two conditions are the same.

The purpose of this report is (1) to describe a further case where borderline pathological features are present; (2) to discuss and compare briefly the

pathology of foetus in foetu and retroperitoneal teratoma and (3) to discuss their symptoms, physical signs, diagnosis and treatment.

Case Report

K.J., a 4-week-old female infant, was admitted to the Department of Child Health, Llandough Hospital, Cardiff, on October 18, 1958. The provisional diagnosis by the referring practitioner was 'intussusception'. The mother stated that the infant, who hitherto had been quite normal, had had vomiting after feeds, apparent abdominal pain, and green loose motions for 24 hours.

Birth weight was 7 lb. 5 oz. and gain had been normal. She was breast fed initially, but changed to artificial dried milk after 10 days. Pregnancy and delivery of the mother had been normal. The family was otherwise healthy and there was no history of previous twin pregnancies.

On clinical examination, the infant appeared well nourished and healthy. Weight was 8 lb. 6 oz. There were no signs of dehydration, anaemia or jaundice. A 'strawberry' naevus was present on the lateral aspect of the right thigh. The abdomen appeared slightly distended in the right upper quadrant, and in this region, extending across the midline, a firm, immobile, circumscribed mass was palpable. The liver could be separately palpated a finger's breadth below the costal margin. Bowel sounds were increased. Rectal examination was negative.

In the early consideration of the diagnosis, a retroperitoneal tumour was thought likely. A barium enema examination performed immediately was found to be normal. An excretory pyelogram showed normally situated and functioning kidneys, with a bifid pelvis on the right side. During these examinations abnormal calcification was noted above the pole of the right kidney. Further postero-anterior and lateral views demonstrated a chain of solid calcification in this region. While exact interpretation was not possible at this time, the presence of this calcification appeared to favour the diagnosis of retroperitoneal teratoma.

Blood group was A Rh+; Hb 80% (11.8 g.); white blood cell count, 21,000/c.mm.; corrected erythrocyte sedimentation rate, 39 mm./hour. A urine specimen was found to contain pus cells and organisms identified as *Esch. coli*. A stool examination for pathogens was negative.

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The intercurrent urinary infection was treated with nitrofurantoin for five days with satisfactory response.

Twelve days after admission, laparotomy was performed through a right paramedian incision (Mr. D. B. E. Foster). The exploration revealed a retroperitoneal tumour approximately 2 in. (5 cm.) in diameter lying somewhat to the right of the midline, between the diaphragm and the third lumbar vertebra. The liver and its hilar structures, the duodenum and the pancreas were displaced forwards by the mass. After mobilization of the duodenum and transverse colon medially and downward, the capsule of the mass was opened with the release of a little reddish brown fluid. The contained mass was easily removed, and after careful dissection the capsule was also excised. The abdomen was closed in layers without drainage.

Postoperative course was uneventful and normal feeding was rapidly re-established. When discharged from hospital on the ninth postoperative day the infant weighed 9 lb. 12 oz. The patient has shown normal progress when seen in the follow-up clinic.

Pathological examination of the specimen was carried out by Dr. F. K. Storing. 'Macroscopically: The specimen consisted of a parasitic foetus showing small imperfectly developed extremities. A separate specimen labelled capsule was also examined. Microscopically: Sections taken from the interior of the trunk part of the specimen showed heterogeneous arrangement of a variety of tissues and cells, including muscle fibres, fat, cartilage, intestinal mucosa, glands and nerve fibres. The appearances were typical of an adult teratoma. The capsule consisted of very vascular connective tissue embedded in which were ganglion cells, nerve fibres and scattered lymph follicles.'

Further detailed examination carried out in conjunction with Dr. Storing, showed other interesting features. Apart from the rudimentary limbs (Figs. 1, 2 and 3), a head and hind end could be identified. The lighter coloured band which can be seen projecting from the ventral surface of the specimen was thought to represent an umbilical cord. Radiographs of the specimen (Fig. 4) demonstrated rudimentary development of a spinal column and rib cage. Some calcification could be seen in one of the limbs.

Pathology

The main features of foetus in foetu and retroperitoneal teratoma will be described and comparison drawn where necessary. Preliminary acknowledgment is made to Lord and Willis, since due to their efforts a clearer understanding of the two conditions has emerged from a confusing and conflicting mass of fact and theory.

Nomenclature, Definition and Classification. Foetus in foetu has become an accepted term, which well describes a fascinating condition. As has been suggested, the names parasitic twin and included

twin may equally well be used, provided the lingering association with teratoma is not propagated. The condition may be defined as a vertebrate foetus included within the abdomen of its partner. No further classification is necessary, though it can be mentioned that earlier writers described intra-abdominal and extra-abdominal types. No recent case of the latter, however, has been substantiated.

The retroperitoneal teratoma adopts the nomenclature, definition and classification of teratomas in general. A number of alternative names have been given to these tumours. These include 'teratoid tumour', 'terato-blastoma', 'teratocarcinoma' (when malignant) and 'embryoma'. Willis (1958) rejects these on various grounds and further condemns the use of the term 'dermoid cyst'. He states that apart from confusion which may occur with the sequestration dermoid cyst, even the simplest teratomatous 'dermoid cyst' contains a second tissue, and may be lined with tissue other than skin. A teratoma is defined as a true tumour or neoplasm composed of multiple tissues of kinds foreign to the part in which it arises. A wide range of classifications have been adopted, based on the physical or microscopic characteristics of the tumour. The use of the simple division, solid and cystic, may be criticized on the grounds that all teratomas exhibit a relative degree of these features. The names monodermoma, bidermoma and tridermoma have been used having relation to the proportion of the constituent tissues arising from the three germ layers. It is felt that this is a clumsy way to describe a tumour which almost always contains several types of tissues. Most commonly teratomas are classified as adult or embryonal, according to the maturity of the cells in the tumour. From the same evidence a tumour may be labelled benign or malignant. Owing to the varying characteristics of these tumours and even a single tumour, none of these classifications is entirely satisfactory. No further doubt should remain, however, that foetus in foetu and retroperitoneal teratoma refer to different conditions.

Incidence. Lord (1956) found 31 reports of alleged foetus in foetu before 1900 and 11 reports since that year. Of the 11 more recent cases only four had unequivocal evidence of the condition.

The retroperitoneal teratoma is also a rare condition, being recognized in only 10% of all retroperitoneal primary neoplasms. It is also a fairly uncommon member of the teratoma family; in order of frequency of occurrence the various sites are ovary, testis, anterior mediastinum, retroperitoneum, sacrum, coccyx, cranium and neck.

FIG. 1.

FIG. 2.

FIG. 3.

FIG. 4.

FIGS. 1 and 2.—General views of the specimen.

FIG. 3.—Close up view of digits of one lower limb.

FIG. 4.—Radiograph of specimen.

Age and Sex Distribution. More than 50% of all retroperitoneal tumours manifest themselves in early life.

Practically all the reported cases of foetus in foetu were noted in the first 12 months of life, usually in early infancy.

In the case of retroperitoneal teratoma, Palumbo *et al.* (1949) stated that the average time of diagnosis was at 13 years; 55% were said to have been diagnosed in the first decade of life, while 30% were found in the first year of life. Cole and Gerrish (1951) reported removing a retroperitoneal teratoma

15 hours after birth, while at the other extreme a similar tumour has been removed from a patient aged 53 years.

There is no indication of any particular sex incidence in either condition. Gross and Clatworthy (1951) made the observation that while the sex incidence in retroperitoneal teratoma is nearly equal, in sacrococcygeal teratomas there is a four to one preponderance toward occurrence in females.

Site of Occurrence. 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, Moyer, Peale, Winborne and Gotwals (1950). In this instance, there was a cerebral tumour allegedly containing five human foetuses. However, doubt exists whether these fully meet the requirements to classify as included quintuplets, and discussion still continues.

The foetus in foetu is usually suspended by a peduncle within a capsule containing a little fluid. The wall of the capsule is often thickened at the point of attachment of the peduncle with an associated plexus of vessels. The thickened part may be orientated toward the base of the mesentery and the origin of the superior mesenteric artery. No direct connexion of peduncular vessels with those of the host has been shown, in the cases where this examination has been possible.

The retroperitoneal teratoma also more commonly occurs in the upper part of the retroperitoneal space. Arbitrarily dividing the space into left and right compartments, with the spinal column forming the median boundaries, this type of teratoma is said to occur twice as frequently on the left side. Most are located in an immediate paramedian pre-axial position.

The retroperitoneal teratoma is typically without a capsule or pedicle, and attachment to the posterior abdominal wall is broad and intimate. The area of attachment may be in close relation to the upper pole of the kidney, the pancreas or the coeliac axis of vessels. Growth of the tumour may displace any related organ with resulting distortion of the anatomical situation.

Macroscopic and Microscopic Appearance. The outward appearance of the foetus in foetu may vary 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 recognizable 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 recognizable including brain, spinal cord, lungs, gonads and adrenals. A rudimentary cardiovascular system may be present, but this is not functional and the foetus is acardiac, deriving its blood supply from its host. The enclosing capsule usually consists of connective tissue with a lining endothelium which may be stratified squamous in type. No trophoblastic or placental tissue has yet been described in the capsule.

The gross and histological features of the teratoma are well known. The tumour may reach a very large size and one weighing 26 lb. has been reported. This is in contrast to the foetus in foetu which remains quite small. Occasionally a well-formed foetal part may be found within the teratoma, such as a single digit. An instance of this was described by Krug (1956). The formation of a vertebral column and the occurrence of segmentation is unknown.

Microscopically, multiple different mature and immature tissues will be observed, with no evidence of organ-like arrangement. It is interesting, however, that some tissue correlation does occur. For example, teeth may be set in bony sockets and may even erupt into cystic cavities, alimentary mucosa may be related to smooth muscle, and respiratory tissue to cartilage. In explanation, Willis (1958) suggests that contiguous tissues may exert an inductive effect on one another. In malignant teratomas abundant immature embryonic cells will be seen, having marked proliferative and invasive properties.

Growth and Malignancy. The 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 factor which determined its parasitic role, it lags behind its host. From the latter it becomes dependent for its blood supply, and does not develop a functioning heart and circulation of its own. Although the limbs of the included foetus may appear to be well developed, their ratio to trunk is behind that of a similar age normal foetus. After attaining a variable size, further growth ceases, and actual retrogression can occur as the host twin progresses. Willis (1958) describes in detail

how reduction of a parasitic twin may be brought about, with loss of head, limbs and internal organs. It is not, however, certain whether the factor involved is the vascular dominance of the host twin, or some inherent defect in its parasitic partner which causes these changes. It thus becomes evident that the extensively reduced foetus may be distinguishable only with difficulty from a teratoma. There is no evidence, however, to suggest that the foetus in foetu can become anything but a quiescent malformation, and therefore has no neoplastic or malignant propensities.

Quite oppositely, the retroperitoneal teratoma is characterized by progressive growth. It probably arises at an early stage in prenatal development and will have been present for a long period before discovery in infancy, childhood or later life. Maturation of tissues in the tumour may continue apace with those of the host. In fact maturation may become complete and the tumour become quiescent. The presence, therefore, of relatively immature tissues in the tumour of a small infant may not be a sign of malignancy. The observation of such cells or tissues in the teratoma of an adult, however, would have serious implications.

Immediate recognition of malignancy may be difficult. It is probable that more than the accepted 10% of retroperitoneal teratomas are malignant at time of operation. Metastases are frequent and may occur in the lungs, liver and vertebrae. If the parent tumour is actively proliferating, for example in a young patient, then the metastases will be composed of mixed cells and tissues. If, however, in an adult type tumour one element becomes malignant, then the metastases will be of this tissue.

Pathogenesis. The foetus in foetu is a monozygotic twin. Developing from a single ovum, it would, but for the limitations in growth which occur later, be of the same sex and closely resemble its host. It is not proposed to discuss the phenomenon of twinning, apart from recalling the strong hereditary tendency, and noting that the membranes may be shared to a varying degree by the developing foetuses. Early cleavage of the dividing cell mass may so occur that each may have its own chorion, or that this may be shared. It is then likely that each foetus will have its own amniotic cavity. Occasionally the twin embryos appear at a later stage and a common amniotic cavity is utilized. Willis (1958) states that most if not all conjoined twins and double monsters are in this group. While it would be possible to theorize that even later cleavage into twins might result in the inclusion

of the second embryo within the body layers of its partner, there is no supporting evidence for this, and for that matter no other reason for foetal inclusion has been suggested.

In contrast to the foetus in foetu, very much has been written about the pathogenesis of the retroperitoneal teratoma and teratomas in general. Many theories have been put forward, both complex and conflicting. Most attempt to relate the development of a teratoma to some form or gradation of twin development, or the parthogenetic stimulation of sex or dislocated germ cells. These views should now be discarded. Teratomas should not be considered as incomplete or distorted foetuses within a host, nor the result of fertilization of an ovum within a foetus or infant.

Spemann (1938) first postulated that a 'primary organizer' determined the orderly development and growth of embryonic tissues into the normal structure of the human body. Askanazy (1908) had already indicated that teratomas must arise from abnormal tissue primordia in early embryonic life, while Budde (1926) first suggested the theory that teratomas result from disturbances in the primitive streak. Needham (1936; 1942), Nicholson (1935) and especially Willis (1935; 1937; 1953) endorsed, expanded and nurtured the theory that teratomas were derived from cells related to the invaginating primitive streak, which escaped organizer influence.

In summary, it may be concluded that the retroperitoneal teratoma (or any teratoma) is congenital, and arises in a median or paramedian location from embryonic pleuripotential cells associated with the primitive streak. These cells, uninfluenced by the primary organizer, form a true neoplasm, which may later exhibit benign or malignant characteristics.

Symptoms, Signs, Diagnosis and Treatment

Foetus in foetu and retroperitoneal teratoma may present in essentially the same fashion. Certain variations do occur and these will be indicated. In a similar way diagnosis and treatment may be considered concurrently.

Symptoms and Signs. These are due to the pressure effects of the tumour, and while initially vague, become increasingly persistent as the tumour enlarges. This is especially so in the case of retroperitoneal teratoma where pressure effects tend to be more severe.

The infant may present with feeding difficulties, vomiting, constipation, weight loss, dyspnoea and pain. Alternatively, abdominal distension with an accompanying abdominal mass may be noticed first.

This is more likely to be the case in older patients having a retroperitoneal teratoma. The mass is usually circumscribed, sometimes mobile and may feel lobulated or cystic. Dilated veins may be seen beneath the skin of the abdominal wall and the extremities may exhibit secondary oedema. Sepsis in, or avascular necrosis of, a foetus in foetu may cause rapid increase in size of the tumour, with pyrexia and possibly signs of peritoneal irritation.

Diagnosis. By a process of elimination the presence of a retroperitoneal tumour may be deduced and indeed it may be possible to specify foetus in foetu, or teratoma.

In infants, other conditions will require urgent exclusion. These include intussusception, Wilms' tumour and neuroblastoma. In older patients extensive investigation may be required to exclude gastrointestinal tract, pancreatic and renal tract lesions. Secondary abdominal lymph node deposits from a malignant testicular tumour may give rise to an abdominal mass, 'the epigastric tumour of Osler'. An associated pyrexia may influence the diagnosis toward a malignant tumour, primary retroperitoneal or otherwise, except in the circumstances already mentioned.

Plain radiographs of the abdomen may provide conclusive evidence. Apart from the position of the soft tissue mass on postero-anterior films and lateral films, and indentation or displacement of gas shadows, irregular calcification or actual bones may be seen. The presence of a formed foetal skeleton, or bones with vertebrate organization is strongly in favour of a foetus in foetu. It must be remembered, however, that calcification may occur in other degenerating tumours. Barium studies of the upper and lower intestinal tract may be necessary if the diagnosis is in doubt. An excretory pyelogram will serve to exclude a renal lesion. Normally functioning kidneys may be outlined, but the close proximity of a retroperitoneal tumour may cause one or the other kidney to be displaced forward and laterally.

Treatment. Surgical removal of the tumour should be carried out without undue delay. Cole and Gerrish (1951) commented that nothing was to be gained in small infants by procrastination, and reiterated that as time goes on the newborn infant becomes a poorer surgical risk.

If the diagnosis is reasonably certain, however, the removal of a foetus in foetu does not normally call for such urgency. In the absence of infective complications, which might induce peritonitis or a local adhesive reaction, a reasonable time for

investigation and preparation may be taken. Depending on the age and condition of the patient and the anatomical relations of the tumour, it may be considered advisable to remove the foetus only, from within its sac.

On the other hand, the potentially malignant nature of a teratoma, and of other similarly situated tumours when the diagnosis is doubtful, calls for early operation. The patient and surgeon should be prepared for a lengthy and difficult procedure. Complete removal of all the abnormal tissue must be aimed at; otherwise local recurrence with possible subsequent malignant degeneration can occur. Preoperative preparation should include a chest radiograph and skeletal films, since distant metastases may have occurred already. Lord (1954) in her review of previous cases of foetus in foetu, observed that the prognosis was far better in this condition, than in teratoma, with or without surgical intervention.

Conclusion

An attempt has been made to separate the characteristics of two interesting conditions, often confused in the past. A further illustrative case has been described, which in the author's opinion was probably a foetus in foetu. Certain contradictions in the pathological description are evident. The internal histology of the specimen was manifestly teratomatous. Yet could this represent all that remained of the interior of a markedly reduced foetus? It is suggested that it may still be difficult to identify correctly borderline instances of these conditions, unless very minute and painstaking examination of the fresh specimen is carried out.

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