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Report on a Retroperitoneal Teratoma (Foetus *in foetu*)

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Abstract. Successful removal of a retroperitoneal teratoma from an infant is described. Various theories dealing with the origin of such tumors are discussed. Blood supply, hemopoiesis and circulation of the tumor have been investigated.

Teratomata have been defined as tumors which are composed of several tissues not usually encountered at the location where such growths arise, and which are capable of a certain amount of differentiation. The degree of differentiation and the primary site of the tumors form the basis of the somewhat arbitrary classification as dermoid cysts, solid teratoma, foetus *in foetu,* as well as gonadal, sacrococcygeal, retroperitoneal and other categories.

The most interesting of these tumors are the so-called retroperitoneal teratomata, since in some of these development has proceeded far enough to produce a nearly complete human foetus; hence the terms "enclosed twin", or "foetus *in foetu*". These tumors occur chiefly in the upper abdominal region and are located dorsal to the peritoneum, usually on the left side of the body, near the spleen, pancreas, or kidney.

Lord (1951) reported forty-two cases of retroperitoneal teratoma; Michalany *et al.* (1951) listed 58 cases and Charles (1953) eighty-two such tumors. Only nine specimens were recognized by Lord as true "foetus *in foetu*" on the basis of the presence of a body axis within the tumor.

Though these growths contain derivatives of all three germ layers and almost any type of tissue or even organ, they never possess all of them. To the embryologist they are of special interest because they represent a distorted picture of normal development; the student of pathology is interested in them because of their tendency to become malignant and because their further study may contribute greatly to the solution of the origin of cancerous growths.

The present paper contains a report of a retroperitoneal teratoma, which we have classified as foetus *in foetu* because it possesses a rudimentary body axis, a spinal cord with indistinct regional differentiation, and appendages.

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Presentation

On April 28, 1959, a twenty-five day old girl was admitted to the Wilden Hospital of Des Moines. The child had been losing weight since birth and had refused to eat. Examination pointed to the presence of a tumor in the area of the left kidney. The tumor and the kidney were successfully removed. The child is alive and growing normally.

The tumor weighed 363 grams and measured 9 by 9 by 6 centimeters. It consisted of a mass of tissue enclosed in a fibrous sac which was double-walled in some areas. The fibrous envelope was rough on the outside but smooth and shiny on the inside, resembling an early chorion-amnion. There was no indication of a true umbilical cord though the specimen bore a small appendage on its anterior surface near the site where the umbilical cord normally takes origin.

The blood supply came from the renal artery; the venous return was not determined. The cranial part of the specimen consisted of an irregular mass of cysts of various sizes and colors. Some of the cysts were filled with a gelatinous mass, others with a clear yellow fluid. Many of the cysts contained fibrous trabeculae spanning their lumina.

The caudal part of the tumor consisted of two malformed legs with feet and digits, and buttocks, as well as a lower anterior body wall. All of these structures were covered with normal appearing epidermis. Some harder substance could be felt within the legs and buttocks, giving the impression of cartilaginous rudiments of leg bones and pelvic girdle (Figures 1-4).[2]

A large number of tissue samples were taken from areas which suggested by their general appearance and location that they might be specific organs, such as brain, spinal cord, tongue, liver, etc.

A histological study of these samples revealed the presence of derivatives of all three germ layers. Some of the skin contained hair follicles and sebaceous glands; other skin samples were devoid of such structures. Most of the stratified squamous epithelium was heavily cornified and in many places horny epithelial pearl had formed. The cavities of the cysts as well as the lining of the fibrous envelope consisted of flat cells resembling mesothelium.

The body axis was represented by a caudal stump which contained a caudal spinal cord with meninges and a large fibrous sac filled with brain tissue. The cross section of the spinal cord showed some regional differentiation; ill-defined fibre tracts, neuroglia and a spinal canal with ependymal lining could be recognized.

[2]The drawings (Figures 1-4) were made by Mr. G. R. Weiner; the photographs were taken by Mr. L. Baldwin. We express our gratitude for their help and cooperation.

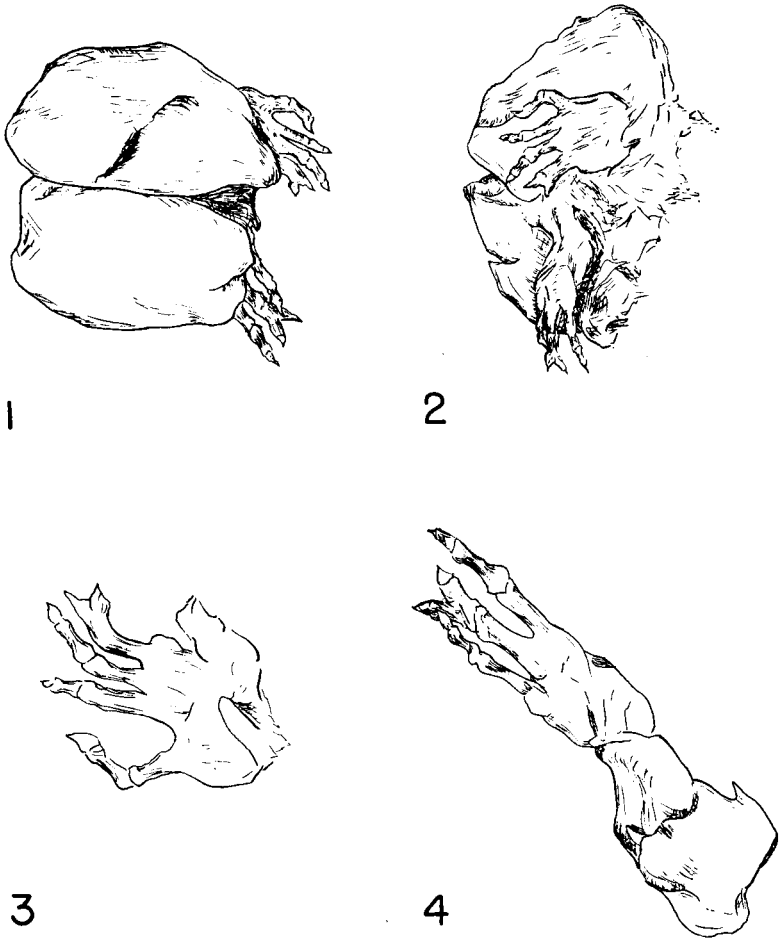

Figure 1. Undissected specimen of a "foetus *in foetu*" viewed in craniad direction, showing buttocks and feet.
Figure 2. The same specimen in dorsal view. The ragged edge on the right shows where some cysts have been dissected away.
Figure 3. Left foot, showing imperfect development of digits.
Figure 4. Right leg of specimen after partial dissection.

Blood vessels were present in great numbers, especially in the fibrous envelope of the tumor. There were veins, sinusoidal spaces, hemorrhagic areas (especially in the deeper areas of the tumor) and arteries with degenerate and hyalinized walls. The blood cells were of the same type as those of the host (Figs. 6, 7.)

The connective tissue consisted in many places of a loose arrangement of collagenous fibres and fibrocytes. Some connective tissue was mesenchymal in character with stellate cells. Degenerate striated muscle was present. The gut was represented by a

portion of large intestine with well developed smooth muscle, a myenteric plexus, normal appearing mucosa, and large and small submucosal lymph nodules. The epithelium contained large numbers of goblet cells.

The cross section of the umbilicus-like appendage (pedicel) was filled with loose connective tissue and had no traces of blood vessels. No bones or cartilages were present; the harder areas of the legs and pelvic part consisted of dense fibrous tissue.

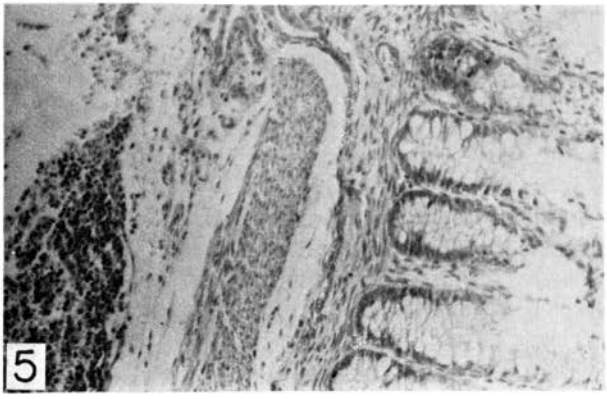

Figure 5. Section of the large intestine showing a large submucosal lymph nodule.

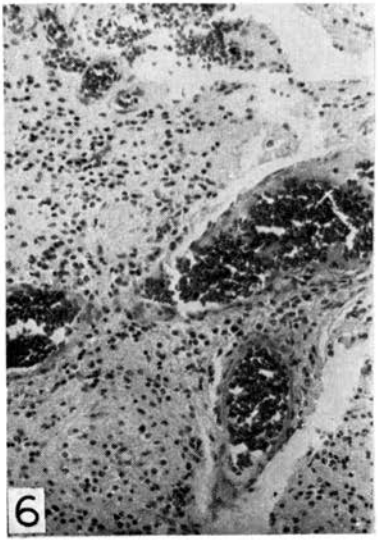

Figure 6. Three arterioles with degenerate walls from the interior area of the tumor.
Figure 7. Sinusoid spaces, venules and extravasations of blood from the lower abdominal area.

Discussion

Numerous theories have been put forward to account for the origin of teratoid tumors. The most widely accepted are the following:

(1) Double fertilization, of either two ova or an ovum and one of its polar bodies, with subsequent migration to an unusual site.

(2) A normally fertilized egg falling apart in the blastomere stage, the fragments developing somewhere outside the uterus.

(3) Ovarian or tubal pregnancy.

(4) Development of a totipotent cell at the original site of the primary gonads or at the actively growing ends of the body axis.

The fertilization of a polar body, while theoretically possible, is not a likely explanation of the origin of a teratoma. *In vitro* experiments by one of us never showed penetration of polar bodies by sperm, though ova were fertilized with ease. It is also hard to imagine how a fertilized egg or a fertilized polar body could accomplish long distance migrations and form a teratoma in such unlikely places as the orbit of the eye, the nasal cavity, or the brain. (*Vide* Kimmel, 1950.)

Recent development of the technique of determining sex by examination of various types of body cells makes it possible to recognize the sex of a tumor. This method of sex determination could be used to decide between some of the theories listed above. One would expct, for instance, that individuals produced from a divided blastula or those originating by parthenogenesis (or ephebogenesis, the male analogue of parthonogenesis) would always provide offspring of the same sex (as in identical twins). Myers (1959) carried out extensive studies on sex chromatin in teratomas and reported the following findings: In sixty-four teratomas from females, typical female sex chromatin was present; in forty-one terotomas of males the nuclear sex chromatin was male in thirteen cases, uniformly female in ten cases and mixed in others. Myers assumed that the occurrence of mosaic tumors rules out parthenogenesis in tumors of male hosts.

Duerken (1932) explained the development of chorion epithelioma or of dermoid cysts in testes and epididymis by parthenogenetic development of spermatozoa. Cowen *et al.* (1958) indicated that teratoid tumors of the epididymis may arise from the Muellerian ducts, remnants of the appendix, testis, or paradidymis.

Intraovarian development without fertilization may furnish a plausible explanation for some ovarian teratomas. Loeb (1932)

demonstrated that in the guinea pig about ten per cent of all cases show such development. The senior author has observed repeatedly spurious development of Graafian follicles within the ovaries of rats and mice. Aften perfect spindles and equatorial plates could be seen. The stimulus to this development seemed to be the onset of atresia.

Since the method of sex recognition by means of sex chromatin still leaves a large area of doubt (*vide* discussion in Myers' paper, p. 52), the present authors have compiled from the literature those cases of retroperitoneal teratoma in which the sexes of both the host and the tumor were stated. In the case of the foetus *in foetu* cited by Lord (1956), the tumor found in a seven-week old boy contained testes. In the Young teratoma (1806-1809) the tumor found in a nine-month-old boy had male genitalia. The Brunkow teratoma (1942) of a fourteen-year-old girl contained a rudimentary ovary. In these few instances the sex of the tumor determined by gross or by microscopic study was established with sex of the host.

Several of the theories given above assume extensive migration. It is well known that teratomas occur in preferred sites such as the gonads, the growing ends of the body axis (Colcock *et al,* 1955). Since the region just below the diaphragm is also the seat of the primitive gonads in the embryo, it has been assumed by some writers that not all of the primitive sex cells have been included in the descending gonads and that those left behind could well give rise to teratomas under certain conditions, including genetic tendencies of the bearer of the tumor. The preferred sites of various localities differ in their growth capacity and metabolic activity; hence, we may expect that the frequency distribution of tumors at various sites differs. This is indeed the case as was shown by Colcock *et al.* (1955), Hoge and Kay (1958), Peterson (1956), and others.

The average sizes of the tumors also differ in various locations as shown in the following table based on measurements by various authors.

Table 1

Average Size of Tumors From Various Sites

Location of tumor	Diameter in cm.
scalp	2.5
nose	2.3
submental	3.0
neck	10.0
head	1.5
mediastinum	9.0
presacral	17.0
epididymis	2.0
retroperitoneal	19.1

Most of the theories which have been put forward to explain the lack of proper order in the arrangement of organs of teratomas (or absence of organs) emphasize the role of organizers. Little weight has been assigned to the nutritional supply, the vascular system of the tumors. An adequate blood supply is essential in fast growing embryos; the development of the circulatory system is given priority during the early stages of embryonic development.

All teratomas described so far lack a placenta and placental circulation. Some of the specimens studied by others were surrounded by a fibrous capsule in which (or just outside of which) a network of blood vessels was found. Such a vascular plexus could hardly be called a vitelline circulation, as the presence of such a circulation would presume the presence of a yolk sac.

On the other hand nearly all described specimens of retroperitoneal teratomas possessed an appendage, usually named "pedicel", which has features reminiscent of a body stalk. The specimen described by us had a stalk originating from the lower anterior body wall, at a site where the primitive body stalk usually arises and where the umbilical cord is inserted before the lower body wall grows, with a resultant cephalad shift of the insertion. The histological examination of this pedicel of our specimen did not show any blood vessels. On the other hand, the teratoma described by Young (1809) contained two blood vessels; the Schoenfeld specimen (1841) (*vide* Lord, 1951) had a stalk which extended to the host's liver and was connected to the umbilical vein. The Davis (1939) teratoma resembled that described by the preceding author.

Most of the other described specimens resembled ours and received their blood supply from one of the systemic vessels of the host. Apparently the nearest artery of the bearer supplies the tumor. Hoeven (1952) found that the tumor studied by him was nourished through the renal artery of the host, whereas Gross and Clathworthy (1951) reported the blood supply as coming from the superior mesenteric. Michalany and others (1951) gave spinal arteries as the source. Our specimen was supplied from the renal artery.

Lord held (1951, p. 637) that there was no direct connection between the vascular system of the host and that of the tumor. If this were true it would raise the question of the origin of the blood cells of the tumor.

Since many of the tumors reported in the literature contained well-developed bones, erythropoiesis might conceivably take place in them. Hemopoiesis occurs in foetal bones as early as the second month of life. The literature on teratomata generally fails to state

whether bone marrow was present in the bones of most tumors. The only paper dealing with bone marrow was that by Fujikura *et al.* (1959), who found red marrow within a femur containing eosinophil granulocytes, myelocytes, and rare normoblasts. No megakaryocytes were found.

Our specimen did not contain bones and there was no evidence of erythropoiesis. We assume that the blood supply came directly from the host, as it most likely did in most of the described specimens. (It is of interest to note that the description of the Tokyo teratoma in the 1960 March issue of *Newsweek* mentioned the presence of whiskers on the face of the enclosed twin. It is possible that such hair growth was induced by blood borne hormones of its host.)

Some hemopoiesis does take place within the tumor. Our specimen contained lymph nodules within the mucosa of its intestine (Fig. 7). Numerous mitoses could be seen within these structures. Fujikura (cited above) also mentioned the presence of lymphoid tissue and lymph nodes. Several other writers have noted the presence within the tumor of thymus and of spleen tissue.

It is questionable whether the blood of the foetus *in foetu* is set in motion by a propelling system within the tumor. Several writers note the presence of a heart (Lord, 1950; Klebs, 1876) but the hearts were described as rudimentary. The Klebs specimen even contained an aorta.

Our specimen contained a considerable number of blood vessels of various sizes, including arteries, sinus-like spaces, capillaries and veins (Fig. 8). It is of interest to note that all arterial walls showed extreme degeneration. In most cases all components except the endothelium were hyalinized. We have observed similar vessel degeneration in an embryo with an ectopic heart combined with incomplete aorta. It can be inferred that the blood flow in such vessels is very slow, if not stagnant.

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