A CASE OF DOUBLE TERATOMA.

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PLATE XXXV.

The following case of teratoma of the testicle followed by teratoma of the head is interesting both from a clinical and a pathological point of view.

Clinical history.—The case was first seen by Dr. Lovett in consultation with Dr. John B. Curtis of Somerville on June 6, 1894. At this time the child was nearly three weeks old, having been born on May 18 after a normal pregnancy and labor. The other child in the family was well developed and strong. The patient was normal with the exception of an enlargement of the scrotum, which was noticed at birth. This swelling, in size about as large as a hen's egg, was situated on the right side and had the appearance of a hydrocele. The diagnosis of hydrocele having been made, Dr. Curtis did nothing in the way of treatment until it had become obvious that the tumor was increasing in size. On June 2 an exploratory puncture was made, and a light yellow serous-looking fluid, amounting to about a gramme was obtained and was followed by some blood. When seen by Dr. Lovett on June 6 the child appeared to be a healthy, well-developed boy, normal in every respect save for the scrotal tumor. The scrotum was very much swollen, dense and shiny, the appearance presented being that of an enormous hydrocele. The circumference of the tumor over the middle portion was 8 inches. Aspiration with a hypodermic needle was attempted under the impression that the tumor was a large hydrocele; a gramme of fluid was obtained from a puncture at one point and was immediately followed by blood. In no other place could fluid be obtained by puncture.

A provisional diagnosis of malignant tumor of the testicle was made, and three days later the testis was removed. The very much thickened cord was dissected out as far as the external abdominal ring, where it was tied and cut off. Union took place by first intention, and on June 14, 6 days after the operation, the child was sent home, and on the 28th was dismissed from medical care.
A Case of Double Teratoma

The specimen was examined both in the fresh state and after hardening in Müller's fluid. The outside of the tumor was smooth. On one side of it the epididymis could be distinctly made out. The tumor occupied the position of the testicle. It was oval in shape and measured 7 x 5 cm. On section it was found to consist of a solid stroma, in which appeared small islands of cartilage enclosing numerous cysts of various sizes. The largest of the cysts measured 1 cm. in diameter. The smallest were just visible to the naked eye.

Microscopic sections were made from the tumor in various places. Examination of these shows that the tumor is composed of cysts and solid tissue. In the latter there is a large amount of striated muscular tissue, the fibres not having any definite arrangement. They appear both singly and in masses; though found in all parts of the tumor, they are more abundant in some parts than in others. The muscular fibres themselves are small and contain a relatively greater number of nuclei than normal. No nerves were found in connection with these fibres. Scattered in the tissue are small areas of cartilage, the largest measuring 2 mm. in diameter. Most of the cartilage is hyaline, although in a few of the areas fibrous tissue is found between the cells. Bone is found in various places, both in connection with cartilage and free. It shows the characteristics of newly forming bone. It contains numerous typical bone corpuscles lying in large wide spaces and does not possess a definite system of Haversian canals. A large portion of the bone is covered with osteoblasts; in places there are large multinucleated giant cells lying in depressions in the bone. The soft tissue of the stroma contains a considerable number of round and spindle-shaped cells. In places these are so numerous as to give to the section a distinctly sarcomatous appearance, and in other places there is a considerable amount of fibrous or mucoid tissue between the cells. Cysts are found in great abundance in the tumor; they are all lined with epithelium. In some the wall is smooth; in others there are numerous papillary projections extending into the cavity. The character of the epithelium lining the cysts varies. In some there is a single layer of the low columnar type; in others the epithelium consists of numerous layers and has the characters of skin epithelium with
a well-marked stratum corneum, while in others it partakes more of the character of the epithelium of a mucous membrane. Some of the cysts are lined with a single layer of ciliated epithelium; or the epithelium is ciliated over some and not over other cells. In one cyst the epithelium is ciliated on the surface, while below this there are two or three layers of cylindrical cells, the arrangement being similar to that found in the lining of the trachea and the large bronchi. Most of the epithelium contains a considerable number of dark pigment granules in and between the cells. In one of the sections there is found a structure which closely resembles parts of the eye. At this place there is a flattened cyst which is partly covered with flat cells containing masses of black pigment. On the wall opposite to this pigmented epithelium, and in places in connection with it, there is a formation of epithelium closely resembling the retina. On the surface of this, close to the pigment layer, there is a smooth lining of epithelium with projections not unlike the cones of the retina. Back of this there are two or three layers of oval nuclei similar to those of the granular layer. A diagnosis of teratoma was made.

Clinical history continued.—The patient, who had suffered much pain before the operation, began steadily to improve, and on September 1 was reported as being strong and well in every way.

Early in January the child was brought to Dr. Lovett's office on account of a swelling in the right parietal region, which had been noticed for over a month. The tumor was connected with the skull. The skin was movable over it. It was hard, about the size of half an apricot, and gave the impression of having been pressed through the skull. The head appeared to be distinctly hydrocephalic, being out of proportion to the size of the child.

Dr. Burrell saw the case with Dr. Lovett and a provisional diagnosis of sarcoma of the skull was made, and in the middle of January, 1895, the child was operated upon at the City Hospital. A skin flap having been made and dissected back, a soft tumor containing and partly covered with bone was found attached to the skull. It was detached by means of a periosteal elevator and a sharp scoop, leaving the skull rough and bare under it. The growth was soft and contained spicules of bone. The wound was stitched up. The child suffered a good deal of shock from the loss of blood during the operation, as the tumor was quite vascular.
A Case of Double Teratoma

The tumor was sent to the pathological laboratory and a diagnosis of teratoma was made. The head at this time was watched very carefully and was found to be growing in size. It increased as rapidly as 1 cm. a day in circumference, and the left parietal region was seen to be bulging. Two weeks later at a private hospital a large flap was made, near the site of the former incision, exposing the skull. An opening was made with a 1½-inch trephine, and at the bottom a large vascular mass containing cysts was found. The hole was extended with rongeur forceps upwards and backwards, and an exploration of the tumor mass was made. The growth was so large in extent that its removal was manifestly impossible, and since it was thought to be a tumor of the brain substance, extensive destruction of it was avoided. The wound was closed and sutured. It united by first intention, and the child seemed somewhat relieved by the operation.

Neither at this nor at any other time in the history of the case was there any tendency toward convulsions nor were there any symptoms indicating implication of the motor area. A fatal prognosis was made, and the child kept under observation from this time on. The circumference of the head continued to increase at about the rate of 2 cm. a week. In March the child began to emaciate rapidly. The skin over the tumor broke and suppuration took place. Toward the end of March there was gangrene of some parts of the skin over the tumor and a fetid discharge from several sinuses. The child became cachectic and at times refused to nurse, almost always suffering extreme pain. Opiates were given in moderation. The child gradually sank and died, April 23, 1895.

Plate XXXV, Fig. 1, is a photograph taken during life. Microscopic examination of the portion of the tumor removed showed numerous cysts, not a few of which were filled with sebaceous matter and hair. Many of the cysts were lined with mucous epithelium and mucous glands were found deep in the tissue. The tumor extended into the button removed by the trephine, but the bone itself appeared to have taken no part in the growth.

The autopsy was made on April 24, 1895.

The body was large for the age of the child; somewhat emaciated, pale and anaemic. On the top of the head, extending more towards the left side, is an enormous tumor mass. The surface of this is irregular; at various places there are knobby projections, the largest being 6 cm. in diameter. Below the tumor the skull was pressed out on the right side.
Over its anterior surface the tumor was covered with skin which was not adherent to it. Posteriorly, the skin in several places was absent over the surface of the tumor. It was everywhere extremely thin and in places necrotic and infiltrated with pus. The necrosis had extended into the tumor. The largest projection of the tumor was over the anterior fontanel; there was also a second of considerable size on the left side at about the middle of the coronal suture. An incision was made completely around the tumor and the skin dissected off from it. The skin was in no place directly adherent to the tumor and was not involved in its growth. Beneath, the tumor was everywhere adherent to the dura. There are no adhesions between the dura and the pia. On removal of the tumor the left side of the brain was found greatly compressed; the convolutions were flattened and the mass of the brain was pressed over to the right side. The projection on the right side of the head was not occupied by the tumor, but by the displaced brain substance. The convolutions, though greatly flattened, were otherwise normal. The weight of the brain was 950 grammes. The pia at the base of the skull was smooth and normal, and the vessels were of normal size. Microscopic examination of the brain after hardening showed no evidence of degeneration.

The tumor weighed 1320 grammes. Its outer surface, especially on the left side, is partly covered with thin bone. In some places the bone is eroded, the tumor protruding through it in round masses. Laterally and anteriorly there are numerous projections apparently composed of cysts with soft fluctuating contents. Long spicules given off from the larger bones extend over the surface. The largest portion of the tumor corresponds in position to the posterior part of the anterior fontanel. A section made directly through it shows that it contains numerous cysts. In places there are small areas of calcification, which lie in dense masses of connective tissue extending in various directions through the tumor. The general appearance of the tumor on section is that of a greyish, soft, medullary mass. The contents of the cysts vary; some are filled with a clear thin fluid, others with a thick, more or less gelatinous material similar to the contents of multilocular ovarian cysts, while in others, again, a brownish material appears. In many of the cysts hairs are found among the contents. In the posterior part of the tumor, corresponding to the areas of necrosis and suppuration, some of the cysts contain almost clear pus. The lower surface of the tumor is covered by the dura and in no place projects below it. The tumor is adherent to the upper surface of the dura and can be separated from it by breaking up the adhesions. Nowhere is there an integral connection between
A Case of Double Teratoma

the dura and the tumor, and nowhere is the skin adherent to the tumor except by accidental adhesions. The bone over the posterior portion of the tumor is in places necrotic and eroded.

The pus in the area of purulent infiltration shows numerous streptococci, and cultures made from the tumor and the organs show the same organism in great abundance.

The remainder of the autopsy showed nothing abnormal with the exception of emaciation.

Plate XXXV, Fig. 2, shows a section through the tumor of the head, about two-thirds of the natural size.

Microscopic examination.—The tumor of the brain was hardened in corrosive sublimate and in formalin. Examination of sections shows the presence of numerous cysts differing in size and lined with various sorts of epithelium. In the solid part of the tumor several varieties of tissue can be made out. The stroma, generally, is rich in cells and in places it has a definitely sarcomatous aspect. There are large numbers of nerves in the tissue. Some of these are arranged in bundles and have the character of medullated fibres. In connection with these nerves, there are groups of ganglion cells in all respects similar to the sympathetic ganglia. In other parts of the tumor a good deal of bone is found, the character of which varies. In places it resembles normal adult bone with corpuscles and Haversian canals. In the neighborhood of the bone there are numerous areas of hyaline cartilage which have no relation to the bone. In addition to the bundles of nerves in the tumor, single nerve fibres are found in considerable numbers. Very many striated muscular fibres are present. In some places these are collected in well-defined masses with a more or less evident connective-tissue fascia around them. In others they appear as single fibres. The medullary nerve fibres are found in considerable numbers in the masses of muscle. In places aggregations of nuclei, apparently corresponding to muscle plates, are found, but no connection between these and the nerves can be made out. A small amount of adipose tissue is found in the tumor.

In several of the sections the solid portion of the tumor over large areas has the general characteristics of central nervous tissue. In this there is an intercellular reticulum consisting of a loose network of
fibres, which are frequently in connection with the processes of spindle and stellate cells. There are numerous nerve cells in this tissue, both scattered irregularly and in small groups. Many of them are triangular and similar to those of the cerebral cortex. The general aspect of the tissue here is similar to that of cerebral tissue, but no separation into white and grey matter can be made out. In several places there is an almost complete reproduction of the cortex of the cerebellum, the tissue here being characterized by the presence of large branching cells like the cells of Purkinje. The main processes from these cells extend up into the reticular tissue, which contains scattered round cells. On the other side the tissue is closely packed with round cells, but no nerve fibres can be made out in it. On the surface which corresponds to the dura there are numerous round cells and much fibrous tissue. In this part of the tumor there is a cyst, lined with low cuboidal epithelium with numerous papillary ingrowths at various places and numerous secondary cysts. A large number of vessels and a small amount of loose connective tissue are found in these papillary ingrowths. In places the epithelium covering the papillae and that of the cyst are ciliated. This structure corresponds so closely to the choroid plexus that it could easily be demonstrated as being identical with it. The portions of the tumor, which so closely resemble the cerebrum, are generally found adjoining spaces which appear in the specimen more as fissures than as true cysts.

The genuine cysts in the specimen are lined with epithelium of various sorts, and many of them contain masses of desquamated epithelial cells. At various places in the wall of some of the cysts hair bulbs are found, and between these lie numerous sebaceous glands. The adipose tissue spoken of appears in places normal; in other places the oil globules are contained in large spindle cells. There are numerous fissures in the specimen, lined with low, flat endothelium. These apparently correspond to lymphatics. In several places there are large cysts lined with high columnar epithelium and in their walls there are deep depressions, resembling the glands of Lieberkühn, which extend into the tissue. Close beneath the epithelium there is a layer of non-striated muscular tissue, the fibres of which are arranged in
small bundles. Beneath, there is a connective-tissue layer, which contains large numbers of blood-vessels, the appearance of the whole being very similar to that of the submucous tissue of the intestine. Under this is a thick layer of smooth muscular fibres having a definite course, and beneath this again masses of muscular fibres are seen extending in the opposite direction. This area is strongly suggestive of a section of the alimentary canal.

In another part of the specimen there is a very irregular cyst with long projections lined with columnar epithelium extending into the tissue beneath, and in the vicinity are conglomerate glands lined with columnar epithelium. No definite structures which could be regarded as belonging to the sexual system are found, nor are there any which agree in character with the liver, spleen, or kidneys. Lymphatic tissue is abundantly present, both in the form of small cellular aggregations around the vessels and in larger masses which correspond perfectly in their structure to that of lymph glands.

In this case a teratoma of the testicle was present at the birth of the child. It grew rapidly, was removed, and there was no return, the wound healing by first intention. The tumor on microscopic examination contained tissues which could be referred to all three of the germinal layers. A short while after birth a tumor appeared on the head, which was partly removed. It continued to grow rapidly and finally attained a size greater than that of the normal head. On microscopic examination this tumor also was found to contain tissues referable to all of the germinal layers. There is little doubt that the cyst lined with cylindrical epithelium and with definite layers of non-striated muscular fibres on the outside should be regarded as representing intestine. The tumor of the head was especially remarkable for the presence of a large amount of tissue belonging to the central nervous system. Most of this had no very definite arrangement. In places, however, the structure of the cerebellar cortex was definitely shown.

There can be no question that both tumors were of independent origin, and that the tumor of the head could not be regarded as due to metastasis from the testicle. The independent development of the
second growth is shown by the fact that it contained tissues which were not found in the tumor of the testicle. We must regard the metastasis of tumors as due to the conveyance of cells from the primary tumor to some other part of the body, by the blood or lymphatic vessels, where they develop into similar growths. In the case of the teratomata it would be necessary to assume that the metastasis is due to the conveyance, not of cells, but of portions of tissue representing all the complicated structures of the teratoma. Such masses of cells could not be carried through the capillary circulation or the lungs. And even if a metastasis could be supposed to have taken place in fetal life, such masses of cells and tissue would have lodged either in the placenta or liver.

The general etiology of teratomata is unknown. They represent a variety in which there is a more complex structure than is found in any other class of tumors. The most diverse tissues may be found, and these are frequently arranged in the form of more or less definite organs. Tissues which represent all of the germinal layers of the embryo may be found in them as in the present case.

The theories regarding the formation of tumors may be briefly stated as follows: (1) The influence of a trauma exciting the cells to proliferation. (2) The influence exercised by parasites. (3) Abnormalities of development, in consequence of which certain cells or portions of tissue retain their embryonic character and afterwards develop into tumors. (4) The influence of pathological conditions which may separate certain cells or portions of tissue from their organic connection. The third of these theories, that of Cohnheim, is commonly used to explain the teratomata. It is assumed that in the course of development certain cells not belonging to the part may accidentally become enclosed in the developing organ. These cells retain their specific type and may give rise to new growths which will have the characters of the tissues into which these cells should have developed. It is very probable that the simple teratomata, especially dermoid cysts, may be explained in this way. As regards the internal organs, teratomata are most common in the ovary and the testicle. With our present knowledge of embryology it is impossible to see how
cells belonging to the epiderm or the entoderm could have been enclosed in either the ovary or the testicle, since both these organs develop from cells which are early differentiated from the mesotheliun. It is only possible to assume that the tumor of the testicle must have developed from the cells of this gland.

In the case of the teratoma of the head it is difficult to give any explanation. This tumor was outside of the brain and lay between the dura and the skull, neither of which structures took part in its formation. A survey of the literature failed to reveal a similar case. Teratomata within the cranium are not very uncommon. They are found most frequently at the base of the brain in the situation of the pituitary body. They have also been described in the ventricles of the brain. The only instance which we have found in the literature in any way analogous to our case was one described by Areetaes* and reported before the Royal Medical Society of Athens. The specimen was obtained from Dr. Milliaresis of Cephalonia. This case appeared to be one of twin pregnancy in which one fetus was an acardiac parasite. The second child was a male of a size corresponding to that of a fetus at the 7th month. The head was as large as in a case of hydrocephalus and had in the region of the anterior fontanel an opening with ragged torn edges. Through this it is probable that the first embryo had come, because it could be easily thrust through the opening, and there was room for it inside the skull. There was a third very incompletely formed embryo within the meninges. The brain substance was pressed towards the base of the cranium. There was no defect in the brain. The third fetus seemed to be connected with the medulla oblongata by a pedicle. It represented a more or less uniform mass, but two upper extremities and one lower extremity could be seen. Areetaes thought, from a closer examination of the third fetus, that there might have been parts of another fetus included in this. The whole description of the case is very imperfect and it is difficult to make out the exact relation of the various fetuses and the portions of them.

It is possible to suppose that there may have been in our case a

* Virchow's Archiv, xxiii, p. 428, 1862.
double germinal area, one area developing into a fetus which included within its skull the other undeveloped germ. The cases of double monstrosity in which the two embryos are united head to head are usually explained by the supposed presence of two germinal areas in the ovum arranged in such a way that the two cephalic extremities come in contact in the process of growth. It is possible that one of these may not develop and in the course of growth may become included within the cranium of the fetus. This explanation would apply much more easily to those teratomata which are found within the ventricles of the brain, because in that situation they could have been enclosed in the medullary groove. In our case the tumor lay entirely without the medullary groove, and the embryonic tissue must have been enclosed not in the medullary canal, but within the lateral plates. We could not regard such a structure as due to fission with inclusion of the separated portion, after the differentiation of the embryo had taken place to any degree. It is not impossible that it may have arisen from fission at a very early cell stage, the separated portion becoming included, and afterwards going on to the development of a tumor.

Recent experiments in embryology have shown that on separation of the cells of the blastoderm, the cell or cells so separated may develop either into perfect embryos or, retaining a partial connection, form double monsters. There is no case known in which separated cells of the blastoderm remain enclosed in other masses of cells and afterwards undergo development. We could not explain why the teratomata formed from such cells should develop by preference in certain situations. Although these experiments have thrown much light on the formation of double monsters, they have not given us any explanation of the teratomata. The great number of tissues found in this case could only be explained by the supposition that either an entire germinal area was included or cells from a very early stage of segmentation.

The points of surgical interest in the case are: (1) The very close way in which the original tumor simulated a congenital hydrocele, even to the extent of twice yielding a certain amount of straw-colored fluid. (2) The complete recovery from the first operation. (3) The
fact that complete removal of the tumor of the head was probably possible at the time when it was first noticed, could a diagnosis of its exact situation have been made. But the diagnosis presented insuperable difficulties, for a tumor apparently of malignant nature had been removed from the scrotum and was followed some months later by a tumor presenting clinical aspects of malignancy, which was naturally attributed to metastasis. (4) That such extensive pressure of the motor area of the brain could exist without any trace of motor symptoms.

DESCRIPTION OF PLATE XXXV.

Fig. 1.—Head of child from a photograph taken during life.
Fig. 2.—Section through the tumor of the head; about two-thirds of the natural size.