A MALIGNANT TERATOMA OF THE PERINEUM.

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The clinical history of the case, for which I am indebted to Drs. W. H. Whitehead and I. H. Manning, of Rocky Mount, N. C., is as follows:

The patient was a male child, born in October, 1898. In July, 1899, the child's father noticed a small tumor, about the size of an acorn, situated in the middle line half way between the anus and scrotum. The tumor was subcutaneous, and neither painful nor tender. In October, 1900, it began to increase in size quite rapidly, as the result, the father thought, of several blows upon it; and the penis became persistently erect. The child had no other abnormality, and seemed in excellent physical condition. The growth was removed by Dr. Tiffany, of Baltimore, in February, 1901. There was a speedy recurrence in loco, so that under date of March 18, 1901, Dr. Manning wrote me that the site of the wound was occupied by a sloughing cauliflower mass as large as an orange, and that a lymph node in the right groin was the seat of a metastasis, the size of a walnut, ulcerating through the skin.

At the time of the operation two portions of the growth were sent in alcohol to me for examination. One of them, almond-shaped, and about 3 cm. in its longest diameter, consisted largely of bone. Pieces decalcified in 5% nitric acid showed the following structure. Both surfaces of the piece are covered by epidermis, which, though quite thin, can in most places be divided into the three layers characteristic of that structure, with here and there an epithelial peg. The epidermis rests upon a corium which contains rudimentary papillae, sebaceous follicles, and hair-roots. A little deeper in the section are numerous plates of bone with rather large marrow spaces. In addition, there are found a number of cyst-like spaces lined by an epi-
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dermis, which rests upon connective tissue containing sebaceous follicles and hair-roots. These spaces contain amorphous debris and a few hairs—in short, in the interior of the tumor are several minute dermoids. There is another space quite different from those just described. It is lined by mucous membrane, the epithelium of which is composed of stratified columnar ciliated cells. The epithelium presents a sinuous outline, being thrown up into minute ridges by a loose cellular submucosa. Outside of this is a quite distinct circular band of smooth muscle tissue. There is still another space lined by stratified squamous epithelium, which is more or less parallel with the preceding. Indeed, the two run together throughout a considerable extent of the piece, and at one point there is a wide opening between them, with quite a sharp line of demarkation between the two sorts of epithelium. In addition to these structures, there were noted a very few striped muscle cells, and a plate of cartilage which stained deeply with hematoxylin. The tumor is fairly well supplied with blood-vessels and lymphatics.

Microscopical examination of the second piece of tissue previously alluded to, shows it to consist of striped muscle tissue extensively infiltrated by masses of new growth. The muscle is, for the most part, in various stages of atrophy and degeneration. Presumably it is a portion of one of the perineal muscles. The tumor masses may be divided roughly, according to size, into large and small. The latter are more or less circular in shape. In some cases it can be made out quite clearly that they are contained within spaces lined by flattened endothelial cells quite distinct from the tumor cells; while in other cases there is no clear line of separation between them, the two passing into each other by a gradual transition. These spaces contain no blood, and are probably lymphatics. In the spaces the cells are cuboidal or spindle-shaped, and have large nuclei, of which some contain well-marked nucleoli, others only chromatic granules. In some of the spaces the arrangement of the cells is distinctly papillary in character: on a blood capillary as a stalk the cells are placed in a thickness of one or more layers; sometimes the
capillary can be traced to a vessel in the wall of the space. The larger masses possess no characteristic shape, and are evidently the result of extensive infiltration of the muscle. It is somewhat remarkable to find the muscle fibers resisting the attack to the extent that frequently muscle fibers, though more or less degenerated, are found within large masses of tumor cells. In these masses the cells are inclined to be spindle-shaped; they are often packed closely together and arranged in columns, separated from one another solely by blood capillaries, which, indeed, constitute the only stroma that the growth possesses. Transitions between the two sets of tumor masses are abundant. A striking feature in both categories is furnished by spaces circular on cross-section and of various sizes, which contain only amorphous granular material and leucocytes. These seem to be due, in some instances at least, to necrosis, in one situation of tumor cells, in another of muscle fibers. In either case the resulting cavity is not filled by tumor cells; on the contrary, the spaces seem permanent, and are lined by tumor cells.

The clinical history of the case and the microscopical examination of the specimens make it certain that we are here dealing with a congenital tumor of a teratoid nature, which has given rise to a malignant neoplasm. Before attempting to proceed to an exact diagnosis let us summarize the leading features. The tumor was found to contain several minute dermoid cysts, plates of bone, and two structures which are susceptible of interpretation as fetal organs in an exceedingly rudimentary condition, viz., a cylindrical cavity lined by mucous membrane, whose epithelial cells were stratified, columnar and ciliated, surrounded by unstriated muscle tissue; and another cavity communicating with the above, but lined by stratified squamous epithelium. Nothing was seen which could be looked upon as representing the central nervous system. The location of the tumor exactly in the center of the perineum should also be borne in mind.

When we come to consider its exact position among such tumors we are met first of all by the difficulties which arise from the lack of a uniform nomenclature. The classification proposed by the late
Birch-Hirschfeld is commendable by reason of its simplicity. He divides these tumors into the following classes (omitting those congenital mixed tumors which develop from the anlagen of the Wolffian body):

A. Tumors developed as the result of invagination and snaring off of portion of the ectoderm or endoderm, or of either of these layers along with mesoderm—dermoid cysts and the so-called enterocysts.

B. Tumors developed in the same way as above, except that all three layers of the blastoderm are involved. These he calls "teratoids." They contain representatives of all of the germ-layers, but no distinct organs. It might be added that a tumor to be placed in this class should be easily referable to the fetal anlagen in its vicinity.

C. Tumors which are really rudimentary embryos. These are the true teratomata, and may be divided into two varieties: the autochthonous, or the teratomata of the genital glands; and heterochthonous, or those which result from the inclusion of one embryo by another—the cases of foetus in foetu.

Following this classification, it seems clear that our tumor belongs either to Class B or to Class C. Birch-Hirschfeld himself, in discussing this classification, admitted that the division between the two classes was more or less artificial, and thought that it might be very difficult to distinguish between them in some cases. It may be said that the present tumor offers such a case. No assistance is to be obtained from a study of similar cases; for, as far as I can learn from an examination of the literature, all the tumors of a teratoid nature which have been found in the perineum belong to Class A of Birch-Hirschfeld's classification, that is, were either dermoids or enterocysts referable to the changes which occur in the cloaca and uro genital sinus during the development of the external genitals. It is true that in the region dorsal to the rectum numerous teratoid tumors have been discovered, which were quite complex in structure, and which, as a rule, were susceptible of explanation by the theory of invagination at the caudal extremity of the embryo—a region
which is remarkable because of the existence of the cloaca and neurenteric canal in addition to a considerable mass of undifferentiated cells. The neurenteric canal in particular is invoked to explain the occurrence of columnar ciliated epithelium in such tumors. It is difficult to understand, however, how such invaginations could produce teratoids anterior to the rectum; and, as has been said, none have been described in that region. It is conceivable, though, that such a tumor as the one under consideration might arise from a partial snaring off of a large portion of the caudal extremity of the embryo at a very early stage, the separated part being included by a regenerated caudal extremity. Such a separated portion would contain anlages to which the various structures contained in the tumor might be referred. The cavity lined by stratified squamous epithelium might be considered as derived from the ectodermal plate which closes the cloaca, and the other cavity might be considered as a representative of the cloaca or, more likely, of one of the genital ducts. The fact that such a great variety of epithelial cells is found in different regions of the urogenital tract, would make it impossible absolutely to deny that the stratified columnar ciliated epithelium found in the tumor originated in that tract. It is a fact, too, that portions of the malignant growth remind one forcibly of the appearances in the fimbriated extremity of the Fallopian tube. It is not impossible that the malignant growth took its origin in the lining of a rudimentary Müller’s duct—in which event we might term it a mesothelioma, as the mesothelium furnishes the lining of this duct. The fact that no evidences of a central nervous system were found is not a valid objection to the above theory, as all of the tumor was not at my disposal.

On the whole it would seem simpler to regard this tumor as a true teratoma in the sense employed in Birch-Hirschfeld’s classification. If we adopt this view the question of its genesis next arises. The parthogenetic hypothesis, which has received new force from the brilliant experiments of Loeb, could not obtain here, however applicable it may be to teratoma of the genital glands. Accordingly we should have to conclude that the tumor is a true foetus in
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foetu in a very rudimentary condition, and that the avenue of inclusion was the urogenital sinus, the closure of the genital folds completely covering it in. Under this interpretation the rudimentary organs referred to would represent the anterior portion of the alimentary canal and the respiratory tract, in accordance with the well-known fact that in teratoma the cephalic extremity receives most development. The absence of brain may be explained by the fact that only a portion of the tumor could be examined. However, brain is not found in all teratomata.

That a mass composed of such tissues should give rise to malignant growth does not seem surprising; rather the wonder is that malignant growths do not arise more frequently in connection with teratomata. Montgomery has recently examined the literature with reference to this point, and, omitting certain tumors of the ovary whose nature was obscure, he could collect but ten cases of malignant teratomata.

In the present case a diligent search was made to determine from what tissue of the teratoma the malignant growth took its origin; but nothing was found to shed any light upon this question—possibly because not all of the tumor was at my disposal. The diagnosis, therefore, has to be made as far as possible from the histological structure alone. It would seem most in accordance with this structure to regard the tumor as an endothelioma which, taking its origin in the endothelium of the lymphatics, developed in and along these spaces finally to break through and infiltrate the tissues of the perineum diffusely.

EXPLANATION OF PLATES.

PLATE XXVIII.

Fig. 1. Section through middle of the teratoma, A small dermoid cyst, B cavity lined by stratified squamous epithelium, C cavity lined by stratified columnar ciliated epithelium, D smooth muscle tissue.

Fig. 2. Tumor masses in lymphatics.

PLATE XXIX.

Fig. 3. The same under higher magnification.

Fig. 4. Through the malignant portion of the tumor.