SACROCOCCYGEAL TERATOMA

REPORT OF A CASE OF DOUBLE TUMOR IN A NEWBORN INFANT

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Benign and malignant neoplasms behind the sacrum and between the sacrum and rectum are not uncommon. The literature contains reports of many such tumors, and although the majority are found in newborn and young children, a number are recorded as occurring in adults. Many different types of tissue have been found. Tissue of nervous origin, including ganglion cells, ependyma, glia, choroid plexus and embryonic nervous tissue have predominated, especially in tumors and cysts lying behind the bone. Other reports mention giant-cell tumors, sarcoma, carcinoma, myoma, fibroma, chondroma, osteoma, chordoma, and angioma. Occasionally a true tail has been found. Structures which apparently contained epithelium of bronchial and intestinal nature have also been recorded, and in cases in Chiari's series pancreatic, suprarenal, and liver tissues were found. In still another type of case the tumor masses show evidence of fetal inclusions, as organs or limbs. Some of these cases have been explained as parasitic twins. Schultz mentions Schwalbe's classification based on a mono-germinal origin. According to this classification, the tumors can be divided into four groups depending upon the degree of differentiation of the misplaced cells or tissue. In the large series of cases reported by Schultz from the literature, the majority showed some relationship to defective development of the caudal end of the central nervous system. In all of the cases reported, however, only a single tumor was present either in front of or behind the sacrum.

Hundling reviews the subject thoroughly and quotes Galbet's statistics as to the occurrence of these tumors. Hundling himself reports 19 cases from the Mayo Clinic, many of which were ependymal-cell gliomas. All but one of his cases, however, occurred in adults, the exception being a postanal dermoid in a three-year-old girl. In Hundling's series, which included five ependymal-cell gliomas, four dermoids, three carcinomas, and two myomas, there were no cases in which the tumor extended both before and behind the bone. In this respect, none of his cases is comparable to the one presented in this paper.

Hansmann reports two instances. One, in a one-day-old infant dying of bronchopneumonia following operation for harelip and cleft palate, and presenting other anomalies, was an ependymal-cell glioma forming a mass attached to the anterior surface of the sacrum and extending through the third sacral vertebra to the anterior surface of the spinal cord. The second patient, a one-month-old female, showed an
external growth at the lower end of the spine, not continuous with the bone. It contained glial tissue, nerve structures, and bone, and attached to it was a piece of well formed bowel.

Weintraub and Young report a case in a colored boy, aged eleven, with a history of trauma followed in a week by the appearance of a lump in the rectal region. Three months later there was removed from the area between the rectum and the coccyx a tumor mass which had a pedicle extending through a defect in the bone. A canal was present in the pedicle. The tumor was diagnosed microscopically as adenocarcinoma with embryonal tissue. In this case, although the mass was found on both sides of the bone, there was only one tumor present.

Utter and Bates report two cases showing some clinical similarity in that in both constipation and vomiting were present. One occurred in a female five months old, one in a female of seven and a half months.

In the case reported by Stewart, Alter, and Craig, in a boy of two years and eleven months, a sacrococcygeal teratoma was found with metastases to the liver, lungs, and inguinal nodes. This is one of the rare cases of these tumors showing generalized metastasis.

Several instances of tumor both anterior and posterior to the sacrum and coccyx have been reported. In the majority of these cases there has been definite connection between the two masses through a defect in the bone. The case to be presented in this paper is of special interest in that there were apparently two separate tumors, one behind the sacrum and another between the sacrum and rectum. Also, the first tumor was found to be benign while the second tumor showed histologically very active malignancy.

Case Report

J. H., a female, was delivered Nov. 18, 1933, by cesarean section. The mother was a hospital dietitian thirty years old. Three years previously, because of dysmenorrhea and sterility, she had had an x-ray examination with the injection of iodized oil into the
uterus, showing retroflexion with stenosis of the internal os. One year later dilatation and curettage were done, and a stem pessary was inserted, which was allowed to remain three months, after which a smaller pessary was inserted for an additional three months. Pregnancy occurred one year later and the child was carried to term without complications.

The baby, a female, weighed eight pounds at birth and was normal in every way except for a semi-solid, lobulated cystic mass attached by a broad base over the sacrum and extending down over the anus in such a position that it was referred to as a human tail (Fig. 1). The mass was completely covered by skin and in areas where the skin was thin was of a dark blue color, transmitting light very poorly. The base extended down to within about 2 cm. of the anus. There was no change in shape when the child's position was changed or when she cried. Pressure on the neck or flexing the spine did not change the size of the tumor. Rectal examination at this time revealed nothing in front of the sacrum. Roentgenograms of the lower spine and pelvis showed no evidence of developmental defect. The sac projecting below the gluteal folds cast an even shadow, and there was no evidence of calcification, nor were there any osseous structures.

When the child was six days old the tail-like mass was removed. It extended down to the sacrum and to the surface of the coccyx but did not penetrate the bone or cartilage. At one stage in the operation, one of the small cysts was ruptured, and a small amount of grayish fluid escaped into the wound. The incision healed by primary union. The child developed normally in every way until the age of ten and one half months. A gradually increasing constipation was then noticed, and inability to void, because of which the family physician was consulted. At this time the bladder could be palpated almost at the umbilicus. On catheterization, some obstruction to the outflow of the urine was encountered, and on rectal examination this was found to be a solid mass about the size of a small grapefruit, filling the entire pelvis. There was barely room for the finger in the rectum between the tumor mass and the symphysis pubis. The mass was posterior to the rectum, apparently filling the pelvis so that it could not be moved in any direction. The rectal mucosa was movable over that part of the mass that could be felt. X-rays again revealed no bony change in the lower spine or pelvis.

It was believed by a radiologist that the tumor was probably too highly differentiated to be affected by x-rays or radium and an attempt was made to remove it surgically. The child was placed in the inverted Trendelenburg position and the coccyx and lower portion of the sacrum were removed. The tumor seemed to be solidly encapsulated, extending up into the pelvis above the sacral promontory (Fig. 2). It was dissected from the anterior sacral wall and was freed from the rectum in the region of the anus, but high up anteriorly the mass had broken through its capsule and was firmly attached to the

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**Fig. 2. Diagram Showing Relationship between Postsacral and Presacral Tumors**

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Figs. 3 and 4. Postsacral Tumor Removed at Six Days

Fig. 3 (above) shows epithelium-lined cysts. In Fig. 4 (below) are seen ganglion cells, nerve tissue, and round cells.

rectum at the lower end of the sigmoid. In this area it was impossible to remove the tumor completely. The wound was closed with interrupted catgut sutures. There was no evidence of recurrence of the postsacral tumor. Although there was very little hemorrhage, there was considerable shock. A transfusion of two hundred c.c. of whole blood was given into the axillary vein, but the child never recovered from the shock of the operation and died a few hours later.

The anesthetic given was phenobarbital supplemented with a very small amount of ether. The respirations were between thirty-six and fifty during the operation. The pulse was around one hundred and forty. Caffein and adrenalin were given before death, but with no definite response.
Fig. 5 and 6. Presacral Tumor Appearing at Ten and a Half Months

The general structure of the malignant portion of the tumor is shown in Fig. 5 (above). Fig. 6 (below) shows epithelium-lined cysts similar to those in the presacral tumor (Fig. 3), adjoining the malignant area.

Pathological Reports: The first specimen, removed Nov. 23, 1933, consists of a mass of tissue from the dorsal sacral region 6 cm. in length, 1.5 to 2.5 cm. in diameter. It is almost completely covered by a thin layer of skin, except for an area 2.5 cm. in length over the proximal anterior surface. The skin surface throughout is smooth. On section the distal half consists of one large cyst cavity containing a clear, pale yellow fluid. The proximal half shows numerous small translucent cysts containing a similar thin, clear fluid. There is also present in this portion some firm yellow tissue between the cyst walls. The tumor appears to have been completely removed. Microscopic examination shows, in section one, the tissue partially covered by a thin layer of skin with numerous skin glands and hair follicles. The subcutaneous tissue in places shows some fat and fibrous tissue with collections of round cells near the blood vessels. In the deeper portions there are large collections of small and large round cells, the exact nature of
which was not determined. In many places, the round cells are collected around lighter areas containing definite neuron cells with a polyhedral form and numerous processes surrounded by a clearer area showing fibrils (Fig. 4). In section two there are numerous small and large cysts, some of which are lined by a thin layer of stratified squamous epithelium and others by a single layer of tall mucous columnar cells. The epithelium throughout is definitely lined by a basement membrane (Fig. 3) The walls of these cysts show a thin layer of smooth muscle surrounding the entire cyst. The remainder consists of fibrous tissue with numerous small young blood vessels. The diagnosis is sacral teratoma.

The second specimen, removed Oct. 9, 1934 (Fig. 2), consists of a tumor mass, oval in outline, 8 cm. in long diameter. The tumor is covered on one surface by a fibrous tissue capsule. On the posterior surface, a piece of cartilage is present, evidently part of the coccyx. The opposite surface is irregular and shows the capsule incomplete, with tumor tissue here forming the external surface of the mass. On section, the tissue is yellow-gray, has a fine architecture with a translucent surface, and areas of cavity formation with semi-liquid necrotic material. Microscopic examination shows, in section one taken from the site of the previous operation, no recurrence of tumor tissue. Section two shows the cartilage of the coccyx surrounded by fat and fibrous tissue. There is no suggestion of tumor tissue posterior to the cartilage. A layer of striated muscle forms the posterior wall of the tumor. In front of the muscle and connective tissue surrounding the coccyx there are masses of glandular tissue with small round ducts lined by tall columnar epithelium and small nests of clear-staining polyhedral cells suggesting islands of Langerhans. These areas most probably are masses of pancreatic tissue (Fig. 7). There are also several large ducts lined by tall mucus-secreting columnar epithelium (Fig. 6). The duct cavities are filled with an amorphous pink-staining material. There are numerous smaller mucous glands and ducts, and small nests of lymphoid tissue present throughout. In other sections the cell masses with a myxomatous stroma (Fig. 5) predominate. The cells here are small, rounded, and polyhedral, and in places show some attempt at gland formation; elsewhere there are definite papillary formations with several layers of cells overlying loose fibrous and myxomatous stroma. In other places the tumor cells form solid masses with the small polyhedral and round tumor cells closely packed with a fine fibrillar stroma. The diagnosis is malignant sacrococcygeal teratoma.

Autopsy showed a healed surgical incision over the posterior sacrococcygeal region with no evidences of tumor tissue here or in any of the subcutaneous tissue extending to the bone. The lower segments of the coccyx have been removed and there is a small amount of regional hemorrhage. There is a large defect present anterior to the sacrum behind the rectum. A small amount of gray, friable material is seen on the posterior wall of the rectum. Otherwise, there is no evidence of tumor. There are no abnormalities of the internal genitalia. Microscopically the only organ of interest is the rectum, which shows a small amount of tumor tissue overlying the serosa, consisting of connective tissue with nests of spindle and polyhedral cells showing invasion of the muscle layers. In this portion, there is no evidence of gland formation (Fig. 8).

DISCUSSION

In view of the fact that these tumors can best be explained by Cohnheim's theory of displacement of embryonic rests, it is of interest to attempt an explanation of this case. The question arises as to the relationship of the two masses and whether one or two defects in development occurred. Most probably both tumor masses were present at birth, the first in the form of a pseudo-tail, as seen in Fig. 1, and the second in the form of a small tumor mass, cystic or solid, in front of the sacrum, which, however, was not large enough to produce symptoms and therefore went unrecognized. During the ten and one half months of
the child's life the anterior tumor grew rapidly, until it became of such size as to cause noticeable effects. Although there are certain histologic structures in both tumors which are similar, and although we do not know the nature of the anterior tumor before it took on proliferative and malignant characteristics, we believe that the two tumors probably originated from separate anlagen. Certain tubular structures lined by tall columnar epithelium and some mucous glands are seen in both. However, the posterior tumor contains nerve tissue and round cells, which are absent in the second tumor, while the anterior tumor shows structures suggesting a relationship to the neurenteric canal.

From the point of view of embryology it is not difficult to see why
such diversified structures are found in this region. Among the various structures present here at some time or other in embryonic life are spinal cord, meninges, and regional nerve tissue, bony vertebral canal, caudal end of notochord, neururenteric canal, post-anal gut, and the vestiges medullaires coccydiens of Herrman and Tourneux.

The absence of metastasis in this case is typical of most malignant tumors in this region. However, the evidence of invasion of the wall of the rectum (Fig. 8) and the general histologic picture indicate the malignant character of the tumor. The fact that the earlier tumor did not recur, the microscopic evidence indicating complete removal, and the benign character of the histologic picture speak against the second tumor having originated from some remaining part of the first. Examination of the region showed no evidences of anatomical connection between the two. Considering these facts, we believe that the case is best explained on the basis of two individual congenital tumor anlagen, one anterior to and one behind the sacrum. The posterior one was easily recognized, while the anterior one was recognized only after it took on proliferative and malignant characteristics and grew to such a size as to produce symptoms of obstruction. We cannot say that the anterior tumor was benign or malignant at the time of birth and cannot state what would have happened histologically to the first tumor in the ten and one half months if it had not been removed.

Conclusion

A case of congenital sacrococcygeal tumor is presented in which there were two distinct tumor masses: (1) a cystic mass behind the bone and beneath the skin, which was histologically benign; (2) a highly malignant teratoma anterior to the sacrum and the coccyx found ten and one half months after birth, causing symptoms of obstruction.

There was no anatomical connection between the two masses and apparently no evidence that the second mass arose from the first or from the same anlage as the first.

The posterior tumor is believed to have arisen as the result of some defect in development of the spinal cord and vertebral canal, while the anterior tumor most probably arose from the neururenteric canal or post-anal gut.

Bibliography


Chiari: Quoted by Schultz.


