MALIGNANT TUMORS DEVELOPING IN SACROCOCCYGEAL TERATOMATA

HERMANN LISCO, M.D.*

BALTIMORE, MD.

FROM THE DEPARTMENT OF PATHOLOGY, THE JOHNS HOPKINS MEDICAL SCHOOL AND HOSPITAL, BALTIMORE, MD.

CONGENITAL MALFORMATIONS and tumors designated as dermoids or teratomata have for centuries greatly impressed and stimulated investigations by many students of biologic and medical problems. The literature dealing with various aspects of the sacrococcygeal group of teratomata alone is so voluminous that no attempt can be made to review it in this paper, nor is it feasible to discuss the various classifications or the theories concerning the nature and embryologic genesis of teratomata. Recent reviews of the subject are those of Ewing,[1] and MacCallum.[2] The latter presents a detailed and

TABLE I

TEN CASES OF MALIGNANT DEGENERATION OF TERATOMATA
From the Literature

No.	Author	Year	Sex	Age at Time of Death	Type of Malignant Tumor	Metastases	Autopsy
	Certain Cases						
1	Rudolphy[4]	1897	♀	12 months	Papillary adenocarcinoma	Lungs	Yes
2	Fletcher and Waring[5]	1900	♂	29 months	Adenocarcinoma	Lymph nodes	Yes
3	Hinterstoisser[6]	1908	♀	23 months	"Alveolar sarcoma"	Lungs Liver Lymph nodes	Yes
4	Bergmann[7]	1911	♂	17 months	Papillary adenocarcinoma	Lungs	Yes
5	Stewart, Alter and Craig[8]	1930	♂	3 years, 3 mos.	Papillary adenocarcinoma	Lungs Liver Lymph nodes	Yes
6	Renner and Goodsitt[9]	1935	♀	11 months	Carcinoma	Invasion of rectum	Yes
7	DeVeer and Browder[10]	1937	♂	21 months	Embryonal carcinoma	Lymph nodes	Yes
	Probable Cases						
8	Gramm[13]	1902	♀	21 months	"Chondrosarcoma"	Lungs Lymph nodes	Yes
9	Pandalai, Forsyth and Stewart[11]	1924	♂	12 months	Papillary adenocarcinoma	?	None
10	Susuki[12]	1936	♂	3 years	Adenocarcinoma	Lungs Liver Lymph nodes	Yes

critical discussion of the theories of origin and many references to the original literature are given.

The histologic structure of sacrococcygeal teratomata has been carefully analyzed and described. The unusual nature and behavior of these growths is undoubtedly responsible for the great number of publications, which is entirely out of proportion to the frequency with which these tumors occur.

* Now at the Dept. of Pathology, Harvard Medical School, Boston, Mass.

Minor variations and insignificant peculiarities are added in some of the case reports of recent date, while other papers are devoted to considerations of the treatment of these tumors. Treatment, indeed, currently represents the major part of the problem of sacrococcygeal teratoma.

Malignant tumors developing in sacrococcygeal teratomata have occasionally been observed. The extreme rarity of such changes and the presence in our files of two cases showing the development of malignancy prompt this report.

An effort has been made to establish the actual number of similar cases in the literature (Table I). Several instances of alleged malignancy, and cited as such, have been omitted from this list on account of insufficient description or apparent faulty interpretation. This refers particularly to cases published in the latter part of the last and early part of this century.

CASE REPORTS

Case 1.—History No. H.L.H. 98937: E. E. Y., a white, female baby, was first seen at the age of ten months. She was referred to the hospital by her local doctor with the diagnosis of a tumor of the right buttock. This was present at birth and had slowly increased in size ever since.

Family History.—This was negative except for the occurrence of frequent tumors on the maternal side. Both parents are well.

Patient's History.—Birth was normal. On the day after birth the mother first noticed that the right buttock was slightly larger than the left. It slowly and progressively increased in size, especially during the eighth and ninth months. The buttock was always firm and at times warm. The anal opening was dislocated toward the vagina. Bowel movements had been quite normal.

Two months ago the child refused to sit or to lie on the right side. The appetite became poor, but there was no loss of weight. A diagnosis of a tumor of the right buttock was made and confirmed roentgenologically. An operation was advised, and the child was transferred to the Harriet Lane Home of the Johns Hopkins Hospital.

Physical Examination.—The patient was a ten-months'-old well-developed and well-nourished white female baby, that was extremely irritable and cried when made to sit up or to lie on the back. The baby preferred the knee-chest position and was standing on a wide base. There was marked lordosis. Both buttocks were quite prominent. The right buttock was larger than the left. There was a large, firm and smooth downward-projecting mass in the right buttock. The overlying skin was quite warm and there were dilated veins on the surface. Dilated vessels were also seen in the skin over the lower lumbar spine and the sacrum. There was also some slight yellowish pigmentation of the skin over this area and a mild depression could be seen over the sacral spine which was covered by a slight growth of hair. The sacrum seemed to be pushed backward. The rectum, on digital examination, was found to be pushed over to the left by a smooth, firm, rounded mass in the right side of the pelvis. The posterior vaginal wall was pushed forward slightly into the vaginal cavity. Behind it, a firm mass could be felt in the pelvis. The anal opening was wide and there was prolapse of rectal mucosa. About the anus a ring of bluish discolored skin about 3 cm. in diameter was seen.

The abdomen was somewhat protuberant. No masses could be felt anywhere in the abdomen. A few axillary, cervical and inguinal lymph nodes were palpable but not enlarged. A diagnosis of sacrococcygeal teratoma was made. The roentgenograms confirmed the diagnosis of a large tumor, seemingly arising from the region of the pelvic outlet. Collections of calcium deposits were seen in it. It was thought that these might represent teeth. Laboratory data were negative.

On surgical consultation, it was decided to irradiate the tumor before operation in order to reduce, if possible, the size of the tumor. From October 16, 1936, to November 9, 20 roentgen treatments were given. Half of these were directed to either side of the tumor. The distance was 50 cm.; a 2 Mm. Cu. and a 1 Mm. Al. filter were used, with 200 K. V., 20 Ma., a total of 1,740 r. in all was given in the course of this treatment.

The growth of the tumor seemed to come to a standstill, whereas it had been increasing in size before treatment. The patient tolerated these irradiations well. Twice there was a transient diarrhea.

The patient was discharged in good general condition, and was to be followed further in the dispensary. Just before discharge a soft cystic mass was felt at the dividing line between the two buttocks.

About three months later, at 15 months of age, the patient was admitted again. She had had a persistent upper respiratory infection while at home. Otherwise, she had been well until six days before admission, when it was thought that the tumor was growing again. At this time several areas of fluctuation were felt in the tumor, which now involved both buttocks, particularly the right.

During a period of 12 days, nine roentgen treatments were given. The dosages were as follows: 200 KV., 20 Ma., skin-target distance 50 cm., ½ Mm. Cu. and 1 Mm. Al. filters. A total of 792 r. was delivered. Five irradiations were directed toward the tumor from the left and four from the right side. There was no change in the tumor at that time, and the patient was again discharged for further observation in the Out-Patient Department. The child remained at home for three and one-half months. She received one treatment with radium in the second month (one month previous to her third admission).

Soon after the radium treatment the child became irritable and refused to stand or walk. She looked pale and lost weight. Two weeks before admission she developed obstinate constipation. She was admitted for the third time at the age of 19 months. The tumor had grown considerably larger and two soft lumps of large size had appeared on the surface of the mass. There was more distortion of the rectum and of the genitalia. Both legs were partially paralyzed and the muscles atrophied. In the right lower quadrant of the abdomen there was a firm mass projecting from the right pelvis and extending to the midline.

Operation.—June 18, 1937: Under general anesthesia, a large, partially firm and partially soft tumor was shelled out and dissected away from the bladder and the rectum. The tumor was found to extend up into the pelvis and was firmly attached to the coccyx, which was resected. The sacrum was curetted, since the tumor was attached to it also. A nodular mass of tumor was seen to extend up into the abdominal cavity and this could be removed also. The patient stood the operation well, and immediately postoperative had a formed stool. Several transfusions were administered. The operative wound drained and healed slowly.

After an initial improvement during the first three weeks after operation, an abdominal mass could be felt. It extended up to the umbilicus. Small ulcerations appeared about the rectum and the child became more and more cachectic. The abdominal mass increased rapidly in size and the liver became enlarged. The child died two months after the operation, at the age of 21 months.

Autopsy Report (No. 15448, Doctor McAllister). The description will be limited to points of particular interest.

The body is that of an extremely emaciated and dehydrated female baby. Many dilated veins are seen over the abdomen and over the back. There are several small shallow ulcers in the perineal region, and there is a draining sinus leading into the right buttock. There are a few palpable lymph nodes in each groin. Both buttocks are enlarged, the right being larger than the left. No other lesions are seen externally.

Gross Pathology.—A large tumor mass, situated in the retroperitoneal tissue pushes all the abdominal organs upward and anteriorly, including the aorta, the inferior vena cava,

and both ureters, which overlie this mass. The tumor measures approximately 13x13x2.5 cm. The right ureter is obstructed and the vena cava is dilated distally. The main tumor has eroded the vertebrae and intervertebral disks and the right ileum posteriorly. The tumor is soft, somewhat lobulated, and greyish-yellow. Several cysts are found in the tumor, the largest measuring 2.5 cm. in diameter. The tumor surrounds the vena cava and aorta. In the inferior vena cava there is, completely filling the lumen, soft yellowish material.

The entire region of vagina and ureteral orifices is infiltrated by the tumor. There is tumor material in the marrow of the head of the femur.

The large tumor is continuous with a smaller tumor in the right buttock. The draining sinus, mentioned earlier, communicates with this mass which is somewhat firmer and pink. The left buttock is free.

Many small nodules are found in both lungs and in the liver. These are of a greyish-white color and soft. The largest ones measure about 1.5 cm. in diameter. A mucoid material can be squeezed out of them. A few cystic areas are seen in some of them. Bronchial, periportal and retroperitoneal lymph nodes contain similar gelatinous tumor nodules.

The tumor removed at operation measures 12x10.5x10 cm. It is somewhat pear-shaped and surrounded by, for the most part, a smooth fibrous capsule. Portions of fat, numerous adhesions and remains of muscle are seen on one side. The opposite side of the tumor is smaller. It is on this part of the tumor that most of the adhesions and muscle tissue are found, and it shall be referred to as the base. The tumor is rather soft and of a somewhat elastic consistency. When cut in numerous sections, the surfaces present a rather uniform picture—showing numerous cysts varying from a few millimeters to 3.5 cm. in diameter. Most of the cysts are thin-walled and transparent. Many contain a gelatinous tenacious bluish-grey opaque material. Groups of cysts are sometimes surrounded by a membrane of connective tissue, giving it a nodular appearance. Small areas of black pigmentation are occasionally seen in the tumor. A few calcified foci are encountered, but no teeth are found. The capsule of the mass varies in thickness from 3 to 1 cm. At the base and along one side of the tumor there is a slit-like separation of the capsule, dividing it into a parietal and visceral portion. The narrow elongated cavity contains a small amount of greasy material and a few hairs in places. The inner surface of this cavity is roughened here and there. The more solid tissue at the base shows some large areas of necrotic, friable material in close association with some of the cystic cavities.

Microscopic Examination.—The tumor removed at operation shows a dense partially hyalin capsule and a variety of tissues. There are derivatives of all three germ layers. The cysts are lined by ciliated and nonciliated cylindrical epithelium, resembling the mucosa of trachea. Some cysts are lined by squamous epithelium. Islands of squamous cells are often found in the cylindrical epithelium as well. Other cysts are lined by cuboidal or flat epithelium. Many mucous glands, with ducts, are present in the interstitial tissue. The lumina of many cysts are filled with a homogeneous somewhat pink-staining material. In addition, parts of the following structures are found in the tumor: Cartilage and bone, smooth muscle, pigment cells, intestinal mucosa, pancreas with islands of Langerhans, ovarian stroma with structures resembling follicles, choroid plexus, nervous tissue, epidermal epithelium with appendages. There are inflammatory cells in some parts of the tumor, and a sinus is found showing fresh granulation tissue in which a few giant cells are seen.

Sections from the base of the tumor show an entirely different type of tissue in addition to the above-mentioned structures. It is composed wholly of a single-type of cell which has large polygonal and relatively pale-staining nuclei with little chromatin and but scant cytoplasm. In some regions the cytoplasm of the tumor cells is vaculated. The cells grow in single layers or in massed sheets. They are situated on a delicate vascularized network of connective tissue. Numerous papillary proliferations are seen and the tissue has, on the whole, the characteristic appearance of a papillomatous tumor. In some

areas an acinar structure is noted. Here a pink-staining, homogeneous material is often found in the lumina, formed by the tumor cells.

The number of mitotic figures varies considerably. In focal areas tumor cells are compactly clustered and in other areas the structure is loose. There are foci of necrosis and hemorrhage. There is invasion of adjacent tissues and blood vessels.

It is this tumor which has invaded the bones and retroperitoneal tissues. There is a thrombus of tumor cells in a branch of the left external iliac vein. The metastases in the liver and lung show the same type of tumor cell. Other microscopic studies revealed a chronic cystitis and an acute necrotizing pyelitis of the right kidney.

Anatomic Diagnosis: Congenital coccygeal teratoma, involving right buttock. Papillary adenocarcinoma arising in teratoma and invading right buttock, lower lumbar and sacral vertebrae, right ileum and head of femur. Extension of tumor into pelvic and retroperitoneal tissues. Metastases in liver, lungs, retroperitoneal and bronchial lymph nodes. Urethral and ureteral obstruction by tumor. Chronic cystitis. Chronic and acute necrotizing pyelitis, right. Compression and invasion of pelvic veins and inferior vena cava. Draining sinus communicating with tumor mass in right buttock. History of preoperative roentgenotherapy and radium treatment.

Case 2.—History No. 3822: H. E. B., white, male, age 13 months, was admitted to the Johns Hopkins Hospital, April 12, 1926, because of constipation and anuria.

Family History.—Not remarkable.

Patient's History.—Patient was a full-term child. At the time of birth a small reddish discoloration was noticed just to the left of the coccyx on the thigh. This resembled a birth mark. It measured about 1.5 cm. in diameter. The family physician advised no treatment. At the age of two or three months a small lump of walnut-size was found in addition to the discoloration. Neither the discoloration nor the lump seemed to increase in size during the first four or five months of life. A little later the mother thought that the area of discoloration and the mass beneath it increased in size. The attention of the family physician was called to this but he advised against any interference. The child developed some slight constipation at the age of about nine months. Since the child was given solid food for the first time during this period, it was thought that the constipation was due to the change of food. The constipation persisted, however, and became worse slowly, so that at the age of 12 months, enemas had to be given. A doctor was again consulted and he gave calomel, whereupon the bowels moved freely, though for a short time only.

A week later it was noted that the left leg was somewhat swollen. The area of discoloration and the mass beneath were larger than before, and there were dilated blood vessels in the skin above and around it. The child lately assumed a knee-chest position when awake and seemed to be much more comfortable in this position.

One week before admission, at the age 13 months, the child seemed toxic and was passing very small amounts of urine. Catheterization was performed, and the child was relieved. Repeated catheterization became necessary, which procedure became more and more difficult. As the symptoms of constipation and anuria became worse, and catheterization more difficult, it was decided to hospitalize the child.

When the patient entered the hospital he had not voided for several hours, and the bladder was greatly distended. A retention catheter was introduced and a decompression apparatus used. The retention was relieved, and the child started voiding again in about one week.

Physical Examination.—The patient was a slightly under-developed, weak and irritable child. The abdomen was distended. The bladder dulness extended two fingers' breadth below the umbilicus. On the left buttock, to the left, and close to the sacrum and coccyx, there was a subcutaneous, soft mass covered by bluish discolored skin, measuring about 3 cm. in diameter. It was freely movable, but seemed to be attached to the deeper structures of the sacrum. The superficial veins of the skin were dilated. The left thigh was swollen. On suprapubic palpation an indefinite mass could be felt along-

side the distended bladder. On rectal examination an almost complete obstruction of the rectum was found in the ampulla. A slit-like opening was all that remained of the rectal canal. The rectal mucosa was smooth and intact everywhere. A large tumor mass could be felt; this was firm and filled out the concavity of the sacrum and was attached to both sides of the pelvis. The mass was so firm as to suggest bone formation. Roentgenologically, a distortion of the lower portion of the sacrum was noticed.

A diagnosis of tumor of the pelvis was made, with the probability of a periosteal sarcoma. Hence, roentgen and radium treatment seemed most advisable. This combined treatment was carried out during the following weeks. The roentgenotherapy was continued for 18 days, beginning at a dosage of 15 minutes and gradually increasing up to 20 minutes. He was given a total of five half-hour radium treatments per rectum, making a total of 500 mc. hours.

A few days after entering the hospital the left thigh was noticed to be swollen and there was definite swelling of the left inguinal nodes. The right inguinal nodes were normal in size.

The treatment was well-tolerated, and the tumor mass became much smaller. The intestinal and urinary obstruction disappeared almost completely. The patient was able to void and normal stools were passed. The swelling of the left thigh and the discoloration of the skin had disappeared, and the subjacent tumor mass had decreased to about one-half the size noted on admission. The patient was discharged 19 days after admission.

For two weeks, roentgenotherapy was continued three times a week. The child improved steadily and continued to do well for about one month. Reexamination, in about four weeks after discharge from the hospital, showed a fairly well-nourished child. An indefinite mass could still be felt on palpation to the left of the midline in the pelvis. The mass had decreased in size considerably. The enlarged lymph node in the left groin had entirely disappeared. The left thigh was still a little larger than the right. The mass in the upper portion of the left buttock was still present but it was much smaller and softer than before.

The general improvement of the child lasted for about one month. After this time the tumor began to grow rapidly. Urinary obstruction occurred again.

On June 1, 1926, two months after the final admission, the tumor had spread to the inguinal nodes on the left side and also the right side. Small nodular masses could be felt in the abdomen and in the liver. The tumor at the coccyx had not increased in size. Roentgenograms showed metastases also in the lungs. The child became weaker, and finally died, July 26, 1926, almost four months after the final admission.

Autopsy Report (No. 9375, Doctor Forbus). The report will be limited to the points of interest. (Abdominal incision only.)

The body is that of an emaciated male child, 16 months of age. The abdomen is greatly swollen. The lower extremities are edematous, the left leg more so than the right. Both buttocks are quite prominent. Just to the left of the tip of the coccyx, and extending for about 5 cm. up over the posterior surface of the sacrum, there is a soft cyst-like mass. The overlying skin is of a bluish color.

The abdominal organs are greatly distorted in shape and location, due to the presence of extensive nodular masses. In the pelvis, occupying the retroperitoneal tissues, there is a large tumor mass which completely fills the whole cavity. This has pushed aside all the pelvic organs. The bladder is pushed far upward, in such a way that it rests on the anterior surface of the tumor mass, right above the brim of the pelvis. In attempting to remove the tumor, after a dissection of the pelvic organs which are not involved by it, it is found that there is a direct continuation of tumor tissue into the structures of the left thigh beneath the inguinal ligament. The tumor is very densely attached to the floor of the pelvis and to the anterior surface of the sacrum and coccyx. There is no invasion of any bony structures. There is a distinct continuity between this tumor and the cystic tumor which was described as being located in the sacral region externally. It is easy to remove the sacral cystic tumor from its bed.

The branches of the vena cava inferior are markedly compressed by tumor masses and a thrombus is found in the main stem of the inferior vena cava. There is an ulceration of the mucosa of the rectum. The retroperitoneal tissues and lymph nodes are diffusely invaded and replaced by a rather soft yellowish-grey, brittle tissue which is of a tenacious and mucinous character. There are widespread hemorrhages. There are numerous metastases in the liver, the lungs, and in bronchial lymph nodes. The better preserved, smaller metastases show a homogeneous grey, granular surface with some mucoid material. There is tumor tissue in branches of the portal veins also.

The pelvis of each kidney is somewhat dilated but there is no appreciable dilatation of the ureters. The other organs show no lesions.

Frozen-section of the sacral tumor show a variety of tissues including intestinal mucosa, skin, and nervous tissue. Sections of the invading tumor show a different type of tissue, which will be described later.

The sections which remain for study (other sections and blocks have unfortunately been lost) show the following: The tumor mass, located subcutaneously in the left buttock, shows a single cyst lined by squamous epithelium. Sections from the cystic part of the pelvic tumor show cysts lined by high columnar mucus-producing epithelium. The epithelium sometimes changes in character from place to place. In some places it is squamous, in others ciliated, in again others it is cuboidal. A few hair follicles are seen in the cysts lined by squamous epithelium. These cysts are surrounded by a rather dense connective tissue. An island of undifferentiated epithelial tissue is found in the interstitial tissue. Smooth muscle as well as nervous tissue is seen.

The malignant tumor is composed of different cells. These cells are somewhat cuboidal; they have little cytoplasm and consist for the most part of large polygonal nuclei with varying amounts of chromatin. The nuclei are relatively pale. These tumor cells are arranged in various ways: They tend to grow in single layers along fine strands of connective tissue, forming small papillary branching structures. In other places they grow in solid columns and in others small acini are formed by tumor cells. Solid nodules of tumor cells are found here and there, surrounded by connective tissue membranes. A cystic appearance is frequently seen and in these parts a pale bluish, homogeneous substance lies between loosely arranged tumor cells. Many mitotic figures are seen. There are necroses and hemorrhages. The metastases throughout the body show the same type of cell and the same variations in arrangement, which is predominantly that of a papillary adenocarcinoma.

The remaining organs show no lesions except for a chronic ulceration of the mucosa of the rectum, which shows loss of mucosa and granulation tissue with an occasional giant cell.

Anatomic Diagnosis: Papillary adenocarcinoma arising in cystic sacrococcygeal teratoma. Extension of tumor into pelvic and retroperitoneal tissues and left thigh. Metastases in liver and lungs, retroperitoneal and bronchial lymph nodes. History of repeated urinary and intestinal obstruction by pelvic tumor and of deep roentgen and radium treatment, with relief of symptoms. Pigmentation of skin. Ulceration of rectum. Slight obstruction of both ureters and slight hydronephrosis, bilateral. Thrombosis of inferior vena cava.

DISCUSSION.—Both cases represent typical congenital sacrococcygeal teratomata. They were both found at, or soon after birth. Derivatives of all three germ layers were found in both tumors.

The outstanding feature in these two cases is the development of a malignant tumor in the teratoma. Both tumors had infiltrated the adjacent tissues and metastasized to various organs. They showed very striking similarities in structure (Figs. 1 and 2).

It has been pointed out by several authors that teratomata—wherever

they occur—are a potential source for the origin of a malignant tumor. MacCallum[2] states: "While it is true that the teratoma itself is benign, it is not at all uncommon to find the development of a distinct carcinoma at some point in its epithelium, exactly as we find it in the body in general." As a rule, one type of cell only assumes the malignant character and spreads throughout the body. The possibility that a variety of cells in a teratoma are stirred to such

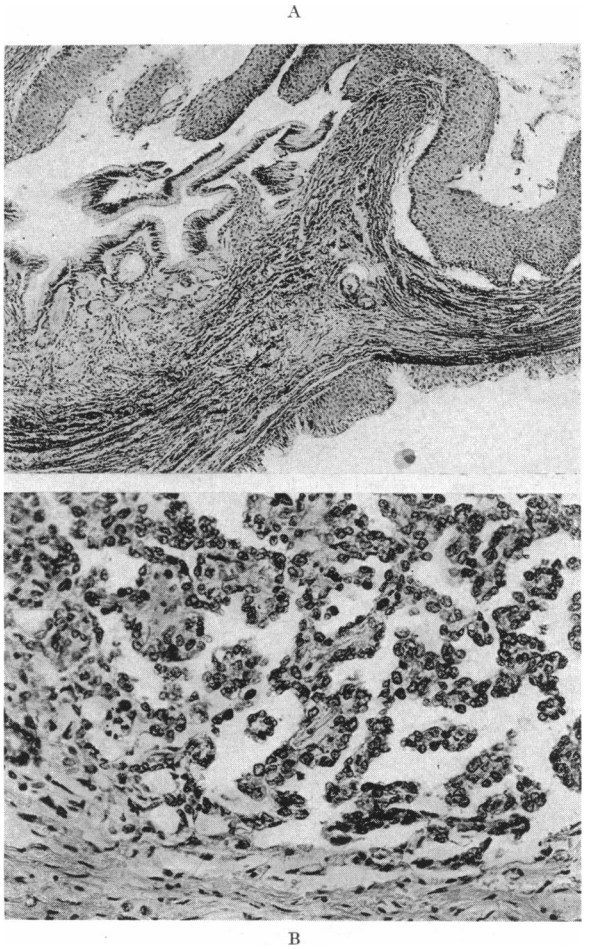
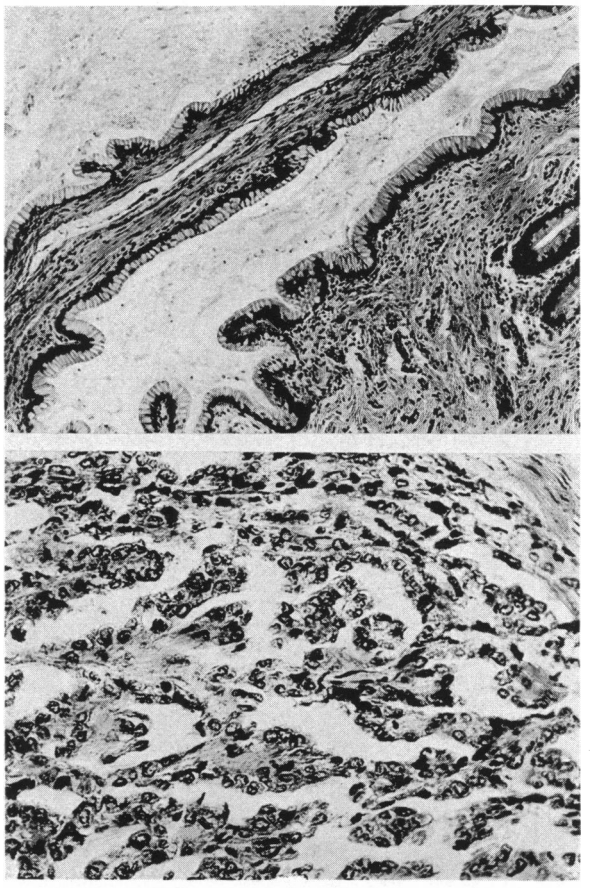


FIG. 1.—Case 1: Path. No. 15448. (A) Portion of teratoma. (B) Portion of papillary adenocarcinoma.

a change and that combinations of tissue metastasize is remote, and has not been shown to occur. However, Schairer[3] reports a remarkable case of a 14-year-old girl who had a teratoma of the left ovary. This was composed of mature and immature tissue elements of all three germ layers. In addition, a very immature and unclassifiable type of tissue was seen in it. There were numerous metastases and in these all tissues of the original tumor were found. The immature cell-forms predominated in the metastases. As it is unlikely

that tissue complexes became malignant and metastasized, as pointed out above, the author explains his case by assuming that a part of the very undifferentiated multipotential portion of the teratoma became malignant. These cells were then swept away by either the blood or lymph stream or implanted themselves on the peritoneum. Here, a number of them proceeded to differentiate and thus images of the original tumors were formed, containing

A



B

FIG. 2.—Case 2: Path. No. 9375. (A) Portion of teratoma. (B) Portion of papillary adenocarcinoma. (Photographs by Milton Nongl).

cysts, cartilage, and neuro-ectodermal tissues. It seems likely that in this case also only one type of cell was stimulated to malignant growth.

The cases reported herewith, and all those collected from the literature showed the development of malignancy, also, of only one type of cell. It is interesting, in this connection, to note the frequency of papillomatous adenocarcinomata. Moreover, all but two cases were malignant tumors of epithelial origin. The case of Hinterstoisser[6] can probably be interpreted, on account of

the minute description, as a carcinoma, while the exact nature of the tumor described by Gramm[13] remains obscure.

The first trustworthy report of a malignant tumor arising in a sacrococcygeal teratoma is that of Rudolphy.[4] The description of the original tumor, undoubtedly, characterizes it as a teratoma and the detailed report on the histology of the malignant tumor makes the diagnosis of a papillary adenocarcinoma certain. The case of Fletcher and Waring[5] also seems to be beyond doubt, judging from the clinical and pathologic data. In Hinterstoisser's[6] case a teratoma was described containing cystic cavities, bone and cartilage, and also some structures which were referred to as round cell sarcoma. The child was well for 21 months after operation, and then a pelvic tumor was found. At autopsy there was a large retroperitoneal mass with metastases in lymph nodes, lungs and liver. This was diagnosed as large cell alveolar sarcoma. It is likely, and the description suggests, that the author was dealing with an adenocarcinoma, although its identity cannot be ascertained today. The case of Bergmann[7] (No. 4 of his series) undoubtedly belongs in this series. No malignant changes were found in the teratoma removed at operation but four and one-half months later a large pelvic and retroperitoneal tumor and widespread metastases were found at autopsy. The tumor showed a papillary structure and gland formation. The case of Stewart, Alter and Craig[8] is a typical congenital sacrococcygeal teratoma in which a papillary adenocarcinoma developed. There were metastases in liver, lungs and lymph nodes. In Renner and Goodsitt's[9] case a typical postsacral teratoma was found and removed at operation. Ten and one-half months later a second tumor had developed which was anterior to the sacrum. Both proved to be teratomata, but in the second tumor a papillary tumor with attempts to gland formation was described in addition to the teratomatous structure. There was invasion of the wall of the rectum. DeVeer and Browder[10] describe a sacrococcygeal teratoma in a six-months-old child. This was removed and no malignant changes were found. Fifteen months later the child died of a tumor, with local extension and metastases to lymph nodes. The tumor is described as embryonal carcinoma.

To these seven cases three more can be added, although they do not show as much evidence either of the true nature of the teratoma or of the malignant tumor.

The case of Pandalai, Forsyth and Stewart[11] is another example of a papillary adenocarcinoma, developing in a teratoma. The papillary adenoma and carcinoma are supposed to have shown striking resemblances to tumors of the choroid plexus. There was no autopsy. Susuki[12] in a paper on "Teratomata" describes an adenocarcinoma with widespread metastases originating in the left buttock. He fails to describe the teratoma. The clinical history is suggestive of the presence originally of a congenital teratoma. The case of Gramm[13] has been noted frequently as an example of this type of tumor. As the original paper was not obtainable it is impossible to include this case with any degree of certainty in this series. However, the data given

in other papers[6, 14] do indicate that there was a teratoma, and do suggest that there was a malignant tumor with metastases to the lungs and lymph nodes. It was diagnosed as chondrosarcoma.

Additional cases described as teratomata with malignancy and some of them cited as such as in the literature, are those of Frank,[15] Nakayama,[16] Leopold and Phillips,[17] Heigl,[18] Sawday,[19] Weintraub and Young,[20] and Gruber.[21] These cases have been referred to in an earlier paragraph and it may suffice to say that there seems no justification to include these cases in this list. There was either apparent misinterpretation of the nature of the malignant tumor or absence of any evidence of a teratomatous tumor. Metastases occurred in none of them, and some of the reports are too fragmentary to draw any conclusions (Table I).

COMMENTS.—It is well known that malignant tumors may arise in teratomata in other parts of the body. They occur, for instance, in such tumors of the testis, the ovary, or the mediastinum. The material presented in this paper clearly demonstrates the possibility of malignant degeneration in the group of sacrococcygeal teratomata. This potential danger should be borne in mind in all consideration of treatment. Law,[22] in a discussion of pelvic tumors with sacral attachments, advocates early treatment: "We believe that, owing to the demonstrated tendency to malignant degeneration, these tumors should be removed when recognized." Similar opinions have been voiced by Stewart, Alter and Craig,[8] and others. Irradiation as a palliative measure is undoubtedly indicated in all cases in which malignancy with metastases has ensued. The demonstrated recurrence in some cases and the development of malignancy even after early operation perhaps indicates the necessity of postoperative prophylactic irradiation even in the absence of malignant changes in the tumors removed at operation. Since some of the malignant tumors were found in the tissues at the base of these tumors (near the point of attachment to bone, connective tissue, or cartilage) it appears important to pay particular attention to this region in microscopic studies of operative specimens.

SUMMARY

(1) Two cases of typical congenital sacrococcygeal teratoma are reported. In each of them a papillary adenocarcinoma with widespread metastases developed.

(2) The literature is reviewed and ten similar cases have been found.

(3) The necessity of early surgical treatment is pointed out.

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