SACROCOCCYGEAL TERATOMA*

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THE sacrococcygeal region is a common site for the occurrence of a variety of fistulae, cysts and tumors. Because of the multiplicity of possible factors in their derivation and the variability of their structure, no rigid classification of these lesions has as yet been effected. Many of them are relatively common (pilonidal cysts and sinuses, sacral meningomyeloceles,† *etc.*) and seldom offer diagnostic difficulties. Much less frequent are the teratomatous tumors which in rare instances have been described as an integral part of an anomaly of the neural canal. These in general are the tumors presenting on the dorsal aspect of the sacrum. Another group, histologically similar, have no important connections with the spinal cord or its membranes but are attached to the coccyx or distal portion of the sacrum. They lie chiefly in front of the sacrum and coccyx but, when large, protrude posteriorly and resemble superficially the sacral meningomyeloceles. Three cases of this type herein reported were in fact, referred to one of us (J. B.) with the diagnosis of meningocele.

We are presenting four cases of sacrococcygeal teratoma and, for comparison, one example of sacral meningomyelocele to illustrate the respective diagnostic features. The relative ease with which the teratomata were extirpated, and the excellent results obtained, contrast strikingly with the difficulties and discouraging prognosis usually attendant upon the surgical treatment of the sacral meningomyeloceles.

CASE REPORTS

Case 1.—H. C., a two months old male, was admitted to the Pediatric Service of Dr. Carl Laws at the Long Island College Hospital February 10, 1925. He was the fourth child of normal parents. At birth two small lumps were present over the lower end of the spine but subsequently they gradually became larger and fused into a single tense tumor which "felt as if it had fluid inside it." No weakness of the lower extremities had been observed nor were there any urinary or rectal difficulties.

Physical Examination.—There were no abnormalities other than a large tumor attached to the buttocks (Fig. 1). The mass was covered with true skin, its surface was lobulated and its base sessile, displacing the anal orifice to the left. The lobulations were fairly soft, seemed cystic but could not be reduced. Rectal examination disclosed a smooth elastic tumor filling the pelvic outlet to such an extent that the examining finger impinged upon the under surface of the symphysis pubis. The external anal

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†The term meningomyelocele is employed throughout this paper to indicate not only this specific anomaly but also the related meningoceles, the syringomyeloceles and the syringomeningomyeloceles.

sphincter appeared normal. No cutaneous sensory disturbances could be demonstrated over the buttocks, and the lower extremities showed no abnormalities of function. When the child cried the mass did not increase in size or become more tense. Roentgenologic examination showed the coccyx to be displaced posteriorly.

Operation.—By Dr. Emil Goetsch. Under ether anesthesia, the base of the tumor was outlined by an elliptical cutaneous incision leaving the skin covering the tumor in situ. By sharp and blunt dissection the mass was freed from the surrounding structures without difficulty. The tumor was found to be attached to the coccyx which was therefore amputated at the sacrococcygeal junction. There was no demonstrable attachment elsewhere. The wound was closed without drainage. There was an immediate postoperative rise of temperature to 104.2° F. which gradually subsided during the following three days. A small area of the wound was superficially infected but slowly granulated and at the end of a month was completely healed. The child was discharged from the hospital April 7, 1925, in excellent condition.

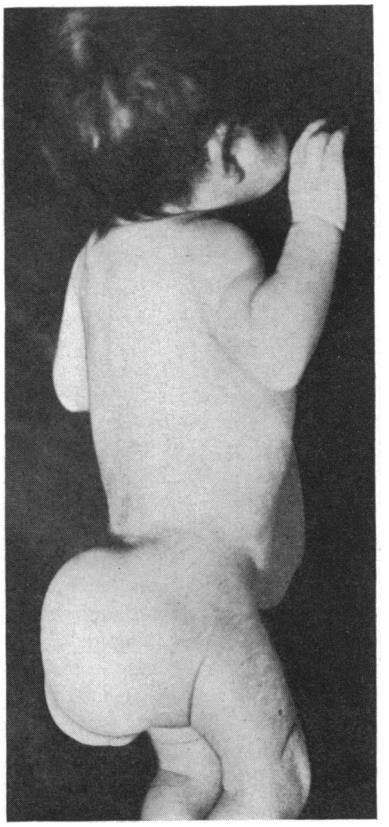

FIG. 1.—(Case 1.) An example of a sacrococcygeal teratoma with a moderately large externally visible portion.

Pathologic Examination.—Gross: The specimen consisted of a coarsely lobulated mass covered in part by normal skin. It measured approximately 12 x 10 x 6 cm. On section it was found to contain several irregular shaped cavities filled with straw colored fluid. These cavities had smooth lining surfaces and their walls were trabeculated. The solid portions were divided into nodules of varying consistency, separated by connective tissue septa. No parenchymatous tissue could be identified as such. *Microscopically* the solid portions of the tumor consisted mainly of lobulated masses of brain tissue with varying degrees of gliosis. An occasional island of bone formation was noted. Scattered in an irregular manner were clusters of cells suggesting compound racemose glands. The cystic spaces were lined with epithelium, resembling bronchial and gastro-intestinal mucosa. Interspersed throughout was much adipose and connective tissue.

Five years later, in April, 1929, it was reported that the child was living and well with no evidence of recurrence of the tumor.

Case 2.—R. T., a three months old female, was admitted to the Surgical Service of Dr. Emil Goetsch at the Long Island College Hospital July 30, 1925. This was the second child of normal parents. It had been delivered by a midwife who noticed a small tumor over the lower end of the spine. This mass was soft and covered with normal skin. There were no abnormalities of the lower extremities. There had been slight increase in the size of the mass since birth.

Physical Examination.—The child was well developed and weighed 12 pounds. Nothing of importance was noted except in the coccygeal region where a round mass could be seen. The overlying skin was freely movable and showed no discoloration. Bimanual palpation with a finger in the rectum disclosed an oval shaped resilient mass about the size of a small lemon apparently attached to the coccyx. There was no increased tension of this structure during forceful crying nor could its contents be reduced. The external anal

sphincter seemed normal. No cutaneous sensory changes or motor weakness of the lower extremities could be demonstrated.

Operation.—Under ether anesthesia, the mass was exposed through a transverse cutaneous incision. It was ovoid in shape, had a smooth surface, and at its cephalad end was fused with the first coccygeal segment. The remainder of the coccyx could not be identified as such. The tumor with the involved coccyx was amputated at the sacrococcygeal articulation. The wound was closed without drainage. Shortly after operation the temperature rose to 104° F. but returned to normal within 24 hours. The remainder of the patient's stay in the hospital was uneventful and she was discharged August 16, 1925.

Pathologic Examination.—Gross: The specimen consisted of an oval shaped mass measuring 6 x 4 x 3 cm. On section it was found to be composed of a solitary cyst with a smooth glistening lining and an irregular intramural nodule 2 x 3 x 2 cm. The cut surface of this nodule was grayish white and appeared to be composed of cellular areas and connective tissue septa.

Microscopic.—Throughout, numerous small islands of brain tissue were distributed. In close proximity to one of the larger areas of brain structure there was a pouch lined by squamous epithelium. In other areas convoluted glands lined with ciliated columnar epithelium were seen. The lining of a number of small cystic spaces resembled gastrointestinal mucosa.

Recent examination showed a normal girl now ten years old, without any demonstrable abnormalities.

Case 3.—S. B., a ten day old female, was born at full term in the Methodist Episcopal Hospital June 15, 1932. Following the delivery there was seen a large lobulated tumor protruding from the region of the buttocks and displacing the anal orifice to the left. During the next ten days the mass grew appreciably larger. The infant urinated and defecated in a normal manner. No disturbance of the function of the lower extremities was noted.

Physical Examination.—The child was a well developed and well nourished female infant with a prominent tumor mass protruding from the region of the right buttocks, approximately one-third the size of the infant's trunk, and completely covered with normal skin. On transillumination shadows could be seen which suggested septa between several cystic lobules. Rectal examination disclosed that the mass extended into the pelvis and almost completely filled its outlet. The anal sphincters functioned normally. No sensory or motor changes could be demonstrated.

Operation.—June 28, 1932. Under local anesthesia an elliptical incision was made about the base of the mass. By dissection, it was easily freed from the surrounding structures. A solid portion was fused with the coccyx necessitating division of the sacrococcygeal articulation. The wound was closed without drainage. There was a sharp postoperative rise of temperature to 103° F., but otherwise the postoperative course was uneventful. At the close of the operation a transfusion of blood was given.

Pathologic Examination.—Gross: The specimen measured approximately 22 x 12 x 10 cm. Its surface was irregular and the larger lobules had thin bluish semitranslucent walls. It appeared that about one-third of the mass was made up of solid tissue. Unfortunately the specimen was lost without further examination being made.

The child returned recently for examination and was found to be normal except for considerable atrophy of the buttocks, more marked on the right side (Fig. 2).

Case 4.—J. S., a six months old male, was admitted to the Kings County Hospital August 28, 1935. At birth there was observed a protrusion about the size of a lemon situated just dorsal to and slightly to the right of the anus. It had grown much larger in the interval and displaced the anus to the left side. For three months there had been a moderate degree of constipation relieved by enemata. No urinary disturbance or weakness of the lower extremities had been noticed.

Physical Examination.—The findings were unimportant except for the presence of

a rounded mass about the size of the infant's head attached by a broad base to the region of the buttocks. This could not be reduced nor was there any increase in its size when the child cried. The external anal sphincter was functioning but there was leakage of fecal material due to its being stretched. The urinary apparatus seemed normal. No motor or sensory disturbance was demonstrable in the lower extremities.

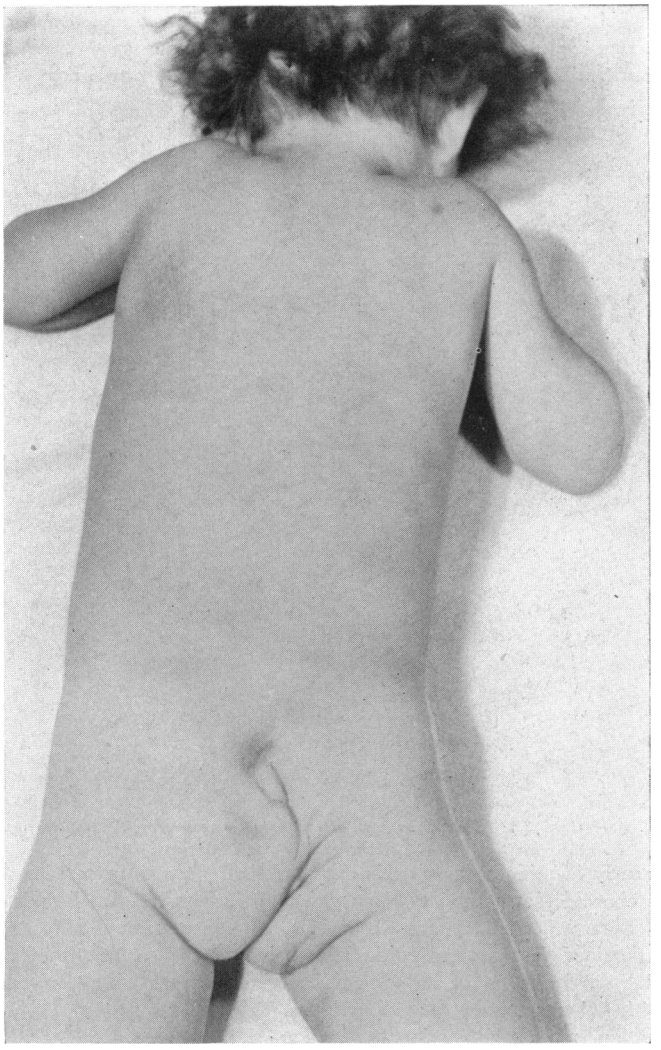

FIG. 2.—(Case 3.) Four years after operation showing atrophy of the buttocks.

Roentgenologic examination of the pelvis showed opaque structures in the mass resembling rudimentary phalanges.

Operation.—August 3, 1935. Under ether anesthesia an elliptical incision was made about the dome of the tumor. The right gluteus maximus muscle was atrophic. The mass was easily dissected from the surrounding structures and was found to be firmly attached to the coccyx. The sacrococcygeal articulation was therefore divided. The wound was closed with drainage. At the end of the operation a transfusion of blood was given. There was a sharp postoperative rise in temperature to 105° F. which returned to normal on the third day; the subsequent course in the hospital was uneventful.

Pathologic Examination.—Gross: The excised mass (Fig. 3) was irregularly ovoid and was partially covered by normal skin. It measured 13 x 10 x 9 cm. Immediately beneath the skin was a large dermoid cyst filled with cheesy material which comprised the greater part of the entire tumor. A nodule of solid tissue, protruding into the cyst, was covered by epidermis with a macerated surface. From this nodule a number of long hairs projected into the cyst cavity. The remainder of the tumor was continuous with the dermoid anlage and lay just outside of the superior pole of the large cyst. This portion was composed of solid gray white tissue in which numerous small cystic cavities were distributed. The latter contained, for the most part, translucent gelatinous material. Several nodules of bone were also encountered on sectioning the tissue. *Microscopic* examination of various portions of the tumor (Fig. 4, A, B, C, D) showed

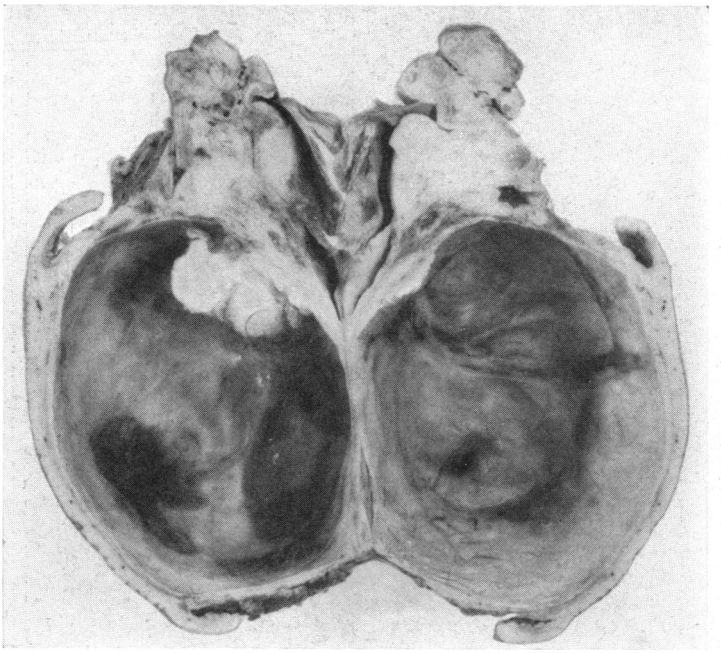

FIG. 3.—(Case 4.) Shows the teratomatous mass removed, containing a large dermoid cyst.

structures clearly identifiable as gastric and intestinal mucosa, pancreatic tissue, gliotic brain tissue, choroid plexus, and salivary glands. There were also islands of bone with active bone marrow, cartilage, smooth muscle, myxomatous and adipose tissue. The large cyst contained desquamated epithelium, hair and the secretions of dermal glands situated in the solid portion of its wall. The smaller cysts were derived from mucus secreting epithelial membranes of various types.

Examination nine months after operation showed considerable atrophy of the buttocks, more marked on the right side. Otherwise the child was well developed and appeared in perfect health.

Since this paper was submitted for publication J. S. (Case 4) was readmitted to the hospital with mild abdominal distention and inability to defecate. Rectal examination disclosed a hard, fixed mass encircling the rectum just within the internal anal sphincter. The extent of the mass could not be determined. There was a rapid growth of this obviously malignant tumor during the next two months and death occurred November 20, 1936. Autopsy disclosed extensive invasion of the pelvis, buttocks and retroperitoneal lymph nodes. Histologically the tumor was found to be of undifferentiated cell type and was interpreted as an embryonal carcinoma.

Case 5.—S. S., a three year old male, was admitted to the Kings County Hospital January 10, 1934. At birth there was noted a small protruding mass at the lower end of the spine, which was soft and had a bluish color. The mass had remained about the same size since birth. There had been urinary and fecal incontinence. The child

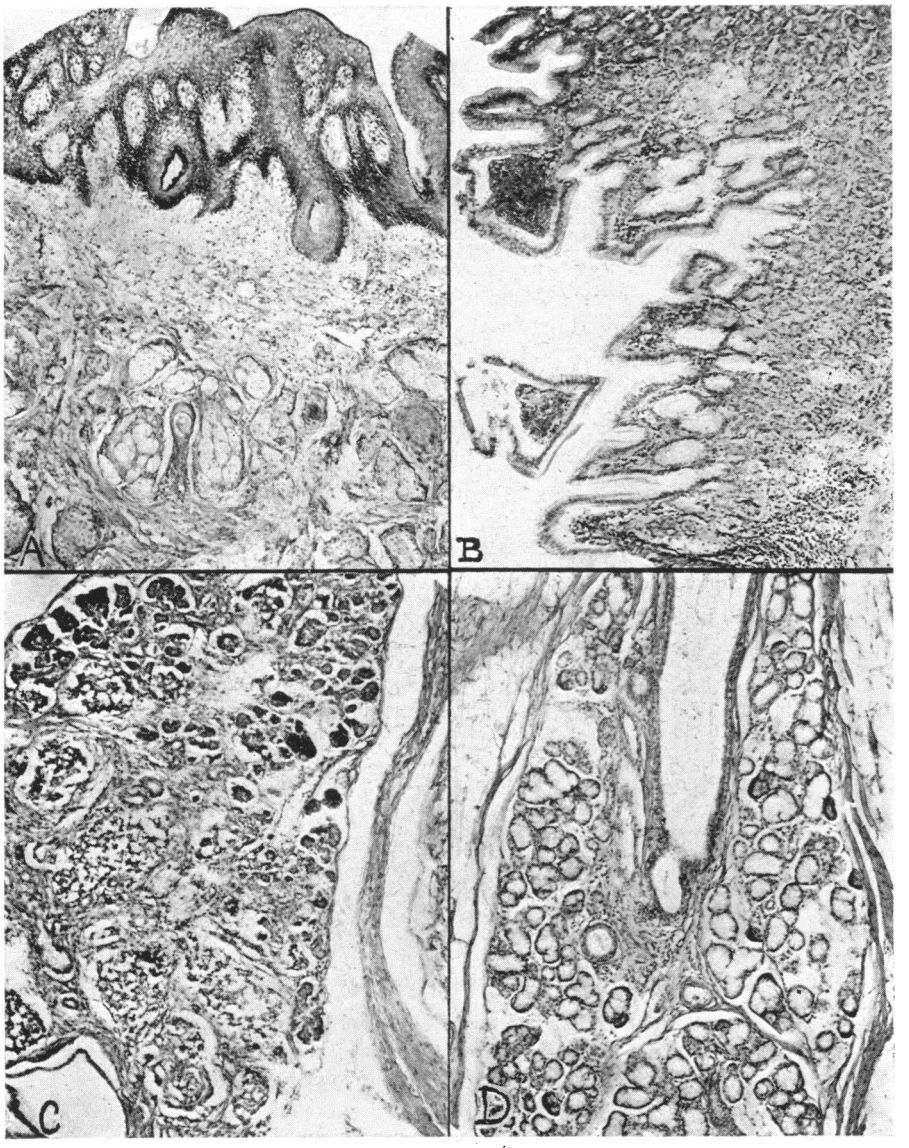

FIG. 4.—Photomicrographs of portions of the specimen shown in Fig. 3. (A) Skin with dermal glands and hair follicles from the dermoid anlage. (B) Gastric mucosa. (C) Pancreatic tissue. (D) Mucus secreting glands probably belonging to the salivary apparatus.

had never been able to walk without assistance. He was brought to the hospital because of the urinary incontinence.

Physical Examination disclosed an undernourished, pot bellied, markedly dehydrated child with a rounded tumor in the region of the sacrum (Fig. 5). The head, upper

extremities and trunk presented no unusual findings. The sacral mass was covered over its distal portion with scaly, embryonal skin. The mass was cystic, could be partially reduced and became quite tense when the child cried. A bony defect in the sacrum was easily palpable. The anal orifice was open and no contraction of the external sphincter was noted. The thighs and legs could be voluntarily flexed and extended but there was no voluntary control of the feet. Complete cutaneous anesthesia and analgesia could be demonstrated from the first sacral dermatome downward.

Operation.—Under ether anesthesia, the mass was explored and there was found an embryonal spinal cord attached to the dome of the meningocele cavity. Atrophic

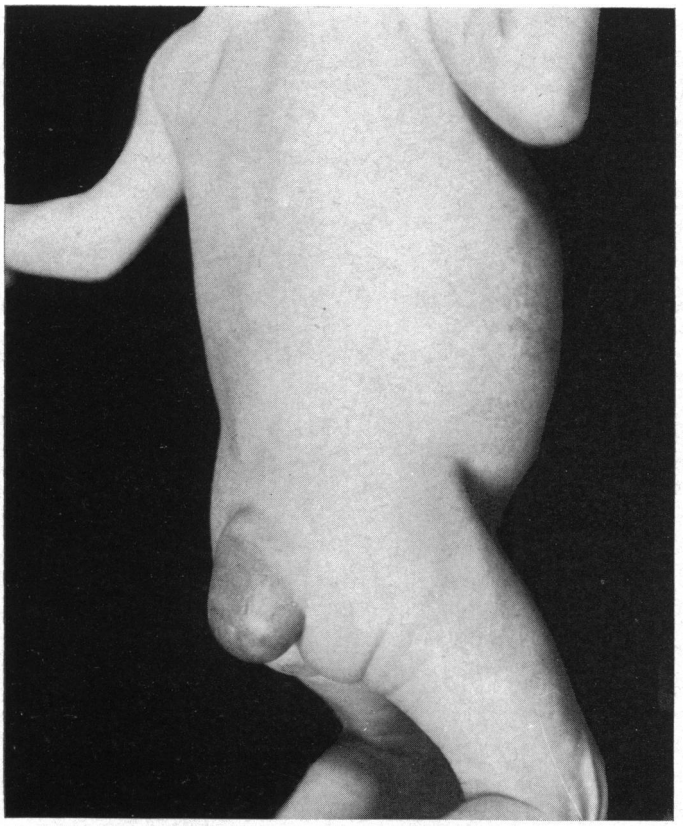

FIG. 5.—(Case 5.) A sacral meningomyelocele, included for comparison.

spinal nerves passed from this cord through the sacral foramina. The spinal cord was detached and the sacral defect repaired. On the third postoperative day there developed evidences of meningitis from which the child died two days later.

Pathologic Examination.—Gross: The specimen consisted of a portion of the wall of a meningomyelocele, covered on its outer surface by wrinkled embryonal skin and on its inner surface by a glistening membrane. Beneath the lining was seen an area of tissue resembling a prolongation of the spinal cord. *Microscopically* there was seen a mass of nerve tissue containing scattered groups of ganglion cells, irregular fiber tracts and patches of glial overgrowth, the latter projecting into the surrounding connective tissue. One surface was covered by atrophic skin.

DISCUSSION.—The subject of malformations and new growths in the sacrococcygeal regions has occupied a prominent position in medical literature of the past 50 years. To attempt a review of the large volume of accumulated data and discussions pertaining to the subject seems unwarranted. The reader is therefore referred to articles by von Bergmann,[1] von Recklinghausen,[2] Kummel,[3] Mallory,[4] Borst,[5] and Schwalbe.[6] A variety of local embryonal structures have been drawn upon to explain the origin of the various fissures, fistulae, cysts and tumors so common in this region. Among these are the fovea coccygea and the coccygeal vestiges of the neural canal, the neurenteric canal, the postanal gut and the proctodeal membrane. According to Ewing,[7] although some of the tumors may, with considerable certainty, be referred to single embryonal structures, the majority of them are more complex and probably involve more than one of these embryonal remnants or some additional anomalies of development. The concept that teratomata may arise from parthenogenetically developing sex cells, a modification of the old "bigerminal theory," has been given much support by the researches of Bosaeus[8] on the origin of ovarian embryomata. He removed from frogs' ovaries unfertilized ova, pricked them with a needle, as Jacques Loeb had done, to stimulate parthenogenetic development, and then reimplanted them into the lymph sac, pleural cavity or ovary of the particular frog from which they had been taken. From these ova complicated teratomata developed that had "essentially the same stucture as the spontaneous adult teratomata or cystic embryomata." MacCallum[9] favors this explanation for the development of teratomata of the gonads where they are often accompanied by chorionic membranes, but holds that the teratomatous tumors in the sacrococcygeal region and brain which approach the complexity of twin inclusions, and even the simplest cysts composed of only one or two types of tissue, are best explained as originating from isolated somatic blastomeres with varying potentialities. A teratoma resulting from parthenogenetic development of a sex cell would be of the nature of an offspring while one derived from an isolated blastomere would be of the same generation as the host, a twin.

It is generally recognized that teratomata such as we have described are benign growths but that one of their component tissues may undergo malignant degeneration. In reporting a case of "Sacrococcygeal Carcinomatous Teratoma," Stewart, Alter and Craig[10] comment that malignancy in such lesions in childhood is either not as common as it is generally thought to be or the cases have not been reported, for they were able to find only four other instances in the literature. These authors quote a statement by Gant[11] to the effect that teratomata in this region show a tendency to undergo cancerous degeneration unless excised early. Renner and Goodsitt[12] have recently reported the case of an infant from whom a "tail-like mass," a teratoma, was removed shortly after birth. The tumor did not appear to extend into the pelvis and no pelvic mass was palpable at that time. Ten months later the child developed constipation and inability to void, and was found to have a

large malignant teratoma located between the rectum and the sacrum and infiltrating the rectal wall. The intrapelvic tumor, which these authors considered as a second or independent tumor, could probably have been successfully extirpated had it been recognizable at the time of removal of the externally visible mass.

Early complete excision of these tumors is indicated as a prophylaxis against malignant change if for no other reason. Also the external portions are not only unsightly but easily vulnerable, while the intrapelvic portions, which are frequently the larger, may cause various pressure effects.

The meningoceles, meningomyeloceles and meningosyringomyeloceles are all the result of defective development of the neural canal. The protruding mass may show a superficial resemblance to the sacrococcygeal teratomata but the two classes of lesions can usually be differentiated by the following features.

DIFFERENTIAL DIAGNOSIS.—(1) The skin covering of the teratomata is true skin whereas the meningomyeloceles are usually covered with thin "embryonal skin" which is easily eroded and ulcerated.

(2) The teratomata vary widely in size. They may protrude but slightly from the pelvis, indeed may be entirely intrapelvic, or present as huge external masses the size of the infant's head or larger, with a palpable intrapelvic portion as well. The meningomyeloceles, on the other hand, are seldom so large and, except in the case of the rare "anterior meningoceles," there is no extension into the pelvis.

(3) A rapid increase in the size of the teratomata is usually noted in the early months of life. This, according to Hansmann and Berne,[13] parallels the growth of the infant and should not be taken as evidence of malignancy. The meningomyeloceles show no such rapid increase in size.

(4) Most of the teratomata can be recognized as containing both solid and cystic portions. They are not reducible and show no enlargement or increased tension when the child cries, or when the jugular veins are compressed. The meningomyeloceles are distinctly cystic and show the various evidences of communication with the spinal subarachnoid space.

(5) The teratomata are associated with no motor or sensory disturbances except, in rare instances, where the tumor presses on the lumbosacral plexuses. The cases of the meningomyelocele group are commonly characterized by loss of sphincteric control and sensory and motor disturbances of the buttocks and the lower extremities.

(6) Hydrocephalus is not encountered in cases of sacrococcygeal teratoma whereas it is often present in the examples of meningomyelocele, particularly after surgical removal of the meningocele sac.

OPERABILITY.—Regarding the operability of sacrococcygeal tumors, we are aware of the fact that not all of the cases offer the favorable outlook of those we are reporting. In each of our four cases the tumor was composed of well differentiated tissues in which no evidence of malignant degeneration was found. In each instance there was no associated spinal cord or spinal

canal anomaly, the tumor was not attached to other important structures and excision was accomplished without great technical difficulty. The fact that all four cases were in infants with tumors externally visible at birth, made early diagnosis and early operation possible. When this is not the case such tumors may go unnoticed until adult life when symptoms of compression of pelvic structures may develop as a result of sudden growth activity. Some of the cases in the group reported by Hundling[14] were undoubtedly of this type. Teratomatous masses have been encountered in association with spina bifida occulta (von Recklinghausen[2]). Keen and Coplin[15] found a fistulous tract passing through a defect in the sacrum and communicating with the rectum in a case of sacrococcygeal teratoma. Such cases present very different surgical problems from those encountered in our series.

CONCLUSIONS

We are of the opinion that a large majority of the sacrococcygeal teratomata in infants are benign, that they can be clinically differentiated from the cases of the meningomyelocele group, and that they are not attached to important structures and should therefore be operated upon as early as possible with the expectation of good functional results. The only residual abnormality to be expected is atrophy of the gluteal muscles in instances where the tumor is very large.

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