UNUSUAL CONGENITAL ANOMALIES OF NEUROSURGICAL INTEREST IN INFANTS AND CHILDREN*

EDGAR A. KAHN, M.D., AND LLOYD J. LEMMEN, M.D.

Department of Surgery, Section of Neurological Surgery, University Hospital
Ann Arbor, Michigan

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This report is based upon some of the rarer congenital anomalies encountered in the Neurosurgery Clinic at the University Hospital during the past few years. No attempt has been made to review the literature on this subject as this has been done so thoroughly by Ingraham and his associates. We wish rather to discuss the practical solution to some of the problems one faces in dealing with these unusual congenital malformations.

Case 1. B. W. was referred on July 5, 1949 to the University Hospital by Dr. R. Atchison of Northville, Michigan, 9 hours after delivery, because of a large congenital defect of the scalp. The pregnancy had been normal, but the description of the delivery is most interesting. Dr. Atchison was told by the nurse that the membranes had ruptured. It was a vertex presentation and on seeing only the shiny scalp defect, it was naturally thought that the membranes were still unruptured. He picked up a hemostat and was about to open the membranes, when fortunately at that moment the patient had a severe labor pain and with this further bulging he realized that it was the child’s head presenting. The remainder of the delivery was normal.

The family history records that a sibling with an entirely similar scalp defect, but no other recognizable congenital defects, died of meningitis at 12 days. This sibling had been seen by us 3 days post partum but the condition was unlike anything we had encountered previously and we deemed the lesion inoperable. The parents were not related to each other and there was no history of a similar lesion in either family.

Examination. A scalp defect, 5×7 cm., extended between the anterior and posterior fontanels, in which there was no normal skin. The defect was covered only with a shiny, thin transparent membrane beneath which could be seen large venous spaces. It was well demarcated from the normal scalp. Palpation gave the impression of incomplete closure of the skull over the midline at the center of the defect (Fig. 1A). There was also present a deformity of one finger and both feet in that some of the distal phalanges were absent. Several hours after admission, the membrane became dry and lost its sheen.

X-ray films showed the anterior fontanelle to be within normal limits. The posterior fontanelle was difficult to make out, but there was no obvious defect in the calvarium between the two fontanels as had been suspected on palpation.

Operation. As soon as studies were completed, the scalp defect was excised under local infiltration with metycaicne ½ per cent. No abnormality was found in the underlying bone. After mobilization of the scalp bilaterally down to the ears

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and by means of rotation flaps, the entire defect was closed primarily (Fig. 1B). Convalescence was uneventful.

_Histological Examination_ (Dr. E. Cawley). “As the defect is approached, the epidermis becomes thinned and gradually fades out, exposing markedly atrophic corium approximately one-third the thickness of normal corium. There are no sebaceous or sweat glands. A striking feature seen in the atrophic portion is calcification of the hair follicles. Diagnosis: Congenital ectodermal defect of the scalp.”

_Comment._ Congenital defects of the scalp are extremely rare, only 76 cases having been described up until 1930. The majority of these were in the same location. A search of the literature revealed no case subjected to surgery. Individuals with large defects have died as a rule of meningitis or hemorrhage from the sagittal sinus or the large veins beneath the defect. It should be stressed that these large congenital scalp defects should be operated upon as an emergency procedure. Procrastination will lead to infection making a plastic procedure most difficult.

Case 2. D. C., a 3-month-old white boy, was admitted on Jan. 8, 1947, because of a mass extending from the bridge of his nose (Fig. 2A). This appeared to have increased somewhat in size since birth and fluctuated when the infant cried. The mass, covered by normal skin, was walnut-sized and extended into the palpebral fissure of the right eye and almost into that of the left. A bony defect of the skull was palpable around the periphery of the mass. Neurological findings were entirely normal.

X-rays of the skull showed a spherical soft tissue mass presenting through a defect involving the medial halves of both supraorbital ridges and the inferior frontal area, as well as the expected position of the superior nasal bones.

_Operation._ On Jan. 13, 1947 the sac of the encephalocele was circumscribed superiorly by an incision down to the peristeum. The incision was then carried down to the root of the nose where the scalp and skin of the face were dissected from
the sac until the entire defect was exposed. The bony defect was larger than a 25-cent piece. The sac, which contained fluid and heterotopic brain, was opened. This brain substance was excised until fairly healthy cortex was encountered. This meant excising the tips of both frontal poles, following which the brain lay free within the cranial cavity. The crista galli was seen. Dural flaps were fashioned and a good closure was obtained. A flap of periosteum was swung over the defect leaving a pedicle for blood supply. The scalp and skin of the face were sutured in more or less of a triangular fashion with one layer of silk (Fig. 2B).

Histologically there were multiple islands of neural tissue with some glial elements. Some of the tissues showed organization suggestive of cortex.

![Image of patient's face before and after surgery](image_url)

**Fig. 2. Case 2. Pre- and postoperative photographs of anterior encephalocele.**

**Course.** In the past 3 years the patient has done well except for several convulsive episodes. Grossly the defect is filled by solid bone which is confirmed by x-ray. His intelligence is normal.

**Case 3.** S. M., a newborn baby, was admitted on April 1, 1946 because of a huge mass in the interscapular region noted at the time of delivery (Fig. 3). This mass was 5×10 cm., with its long axis in the long axis of the body. Huge veins beneath the skin that covered the cystic mass were a striking feature. On deep palpation firm tissue was present. There was no neurological abnormality.

X-ray films of the chest and spine showed a large mass extending posteriorly from the mid-dorsal spine. There was marked widening of the neural canal in the area of the origin of the broad pedicle of the mass.

**Operation.** On April 19, 1946 under local anesthesia, an incision was made around the lower part of the base of the mass, where the large veins were encountered. There was then entered a cavity containing a large amount of bloody fluid. The mass extended laterally from a defect in the vertebrae which was about 2.5×1 cm. There was no dural sac surrounding this vertebral defect as in the ordinary myelomeningocele. Some fluid appeared in the defect but its origin could not be determined. The entire mass was removed down to what was thought to be epidural space, but the anatomy was poorly defined. The remaining tissue lay free in the
vertebral defect. A fascial closure could not be obtained. Muscle flaps and a layer of subcutaneous tissue were sutured over the defect. The skin was closed in a triangular manner, that on the right being exceedingly thin.

**Course.** The child, now 4 years old, is stated by the family physician to be normal in all respects.

**Pathological Examination.** Section of the mass removed revealed two large cystic spaces which occupied the center except for a rim of tissue about 1 cm. in thickness beneath the surface epithelium. On microscopic study this area was found to be composed predominantly of aberrant brain with embryonic neurons and neuroglial elements. There were numerous vascular channels in and surrounding the aberrant brain tissue. Cartilage, mucous glands, nerve fibers, ganglia, and mucosa, which in one area resembles respiratory type epithelium, and in another area intestinal type mucosa, are present in only a limited area. In the deeper sections there was ependymal lining with areas of choroid plexus. Diagnosis: Trigeminal teratoma.

**Comment.** This is an unusual case in that the sac one ordinarily sees in a myelomeningocele arising from the vertebral defect was absent grossly and histologically. The differential diagnosis between teratoma and myelomeningocele was established in this case only by a detailed histological examination of the entire specimen. This midline mass probably prevented the normal bony closure, thus producing the vertebral defect.

**Case 4.** S. T., a 2-year-old boy, was admitted on Oct. 10, 1949 because he had meningitis secondary to a congenital dermal sinus infection in the lumbar region. One month prior to admission a small papule developed in this area which
discharged pus. On Sept. 26, 1949 muscle twitchings, stiff neck and fever developed. Lumbar puncture revealed a cell count of 13,900/c.mm., 99 per cent of which were polymorphonuclear leukocytes. The culture, however, was negative.

The patient was treated for meningitis by Dr. M. Cooperstock at Northern Michigan Children’s Clinic with sulfadiazine, streptomycin and penicillin to which he readily responded. As soon as the symptoms of meningitis had subsided, he was referred to the University Hospital.

Examination. There was an erythematous, pigmented area of skin over the level of L4, at the center of which was a dimple surrounded by a tuft of hair. One cm. to the right of the dimple was a small opening which exuded thick yellow pus.

Operation. On Oct. 13, 1949, under avertin and ether anesthesia, a horizontal elliptical incision was made excising this area of pigmentation. The dissection was carried down to the deep fascia and a stalk, 2 mm. in diameter, was isolated. The muscles were then stripped from the spine and lamina of a normal vertebra above. This lamina was removed and normal epidural fat showing no signs of infection was uncovered. Beneath this was normal dura. Bone was removed until the point was exposed where the stalk entered the posterior dura. It could then be seen that this stalk was running from the skin directly to the dura and entering it at an acute angle. The dura was then opened longitudinally without entering the arachnoid, excising the stalk with a narrow dural cuff. Attached to the stalk intradurally and adherent to a vascular arachnoid over a nerve root was a small nubbin of tissue, 3×2×1 mm., which was firm and yellowish in color (yellow nubbin). This was excised along with the stalk as a single specimen. The other nerve roots of the cauda equina appeared normal and there was a free flow of colorless CSP. The dura was readily closed.

Course. This was entirely uneventful and a 4-month postoperative check-up by Dr. Cooperstock showed no neurological defect.

Microscopic Examination. From the skin a cornifying stratified squamous epithelial tract can be traced in serial sections (Fig. 4), through the corium down to a blind end (yellow nubbin) lined by stratified ciliated columnar epithelium with a definite basement membrane (Fig. 5). Some of the cells have the appearance of goblet cells seen in respiratory epithelium. There are small mucous glands, inflammatory cells, foreign body giant cells, and a solitary nerve rootlet in the tissue surrounding the blind end. Throughout the sinus tract may be seen keratohyaline and amorphous debris and in the blind end (yellow nubbin) there are necrotic pus cells. Diagnosis: Teratoid.

Comment. The histological structure of the nubbin is very similar to two larger lesions described by Kubie and Fulton and others. Had not infection intervened, we have the potential development, depending upon which process would predominate, of either a cholesteatoma from the kerato-
hyaline from the stratified squamous epithelium or a mucous cyst from the mucous glands. Adams and Wegner\(^1\) collected 6 cases of the latter which caused intermittent compression of the spinal cord. Sachs and Horrax\(^9\) found 14 reported cases of intraspinal epidermoids and dermoids associated with dermal sinuses. They found no teratoid lesions in the cases they collected of this type.

**Case 5.** D. G., a 9-day-old white baby girl, was admitted on April 1, 1949 because the repair of a myelomeningocele noted at birth had not held. She was born following

![Image](A) Junction of stalk and yellow nubbin with transition of stratified squamous epithelium to ciliated columnar epithelium (hematoxylin eosin, X120). (B) Yellow nubbin attached to nerve root. Pus cells are shown in the cavity lined by ciliated columnar epithelium (hematoxylin eosin, X120).

a normal pregnancy and delivery. Because the membrane was so very thin and it appeared that rupture was imminent the family physician closed skin and fascia over it without opening the sac. A wound dehiscence occurred on the 8th day and the dural sac herniated through the open wound without rupturing.

**Examination.** The entire operative incision was open and what appeared to be a purulent dural sac was protruding through the opening. There was a mild temperature elevation which was felt to be secondary to the infected wound. There was no weakness of the lower extremities and the anal sphincter was intact.

**Course.** The baby was placed in Trendelenburg position and furacin dressings
were applied to the wound. She was given full therapeutic doses of penicillin and sulfadiazine. After 8 days the fever subsided and the gross evidence of infection around the postoperative wound had cleared.

Operation. On April 11, 1949, under ether anesthesia, the granulating meningocele sac was circumscribed into fairly normal skin. The dissection was carried down to the base of the sac, the neck of which measured 1 x 0.5 cm. as it entered the bony defect. The sac was opened and the abnormal spinal cord tissue which was adherent to it was dissected free so that the spinal cord lay free in the dural canal.

A good dural closure was obtained using fine chromic catgut. Fascial flaps were sutured over the vertebral defect, giving an excellent closure.

Course. The wound healed by primary intention. Examination 1 year postoperatively showed the child to be normal in all respects.

Histological Examination. There was a marked infiltration of polymorphonuclear cells, plasma cells, and foreign body giant cells in the connective tissue, adipose tissue and aberrant nervous tissue. There were colonies of microorganisms in the more superficial areas of the skin.

Comment. This case shows that with the present day antibiotics an extensive dissection of this type can be carried out in the presence of controlled infection.

Case 6. M. L., a 4-month-old girl, was admitted on Nov. 7, 1949, with a mass in the lumbar region present since birth. The mass, well covered with skin, was
9×6×2 cm. and pulsated on crying. There was slight weakness of the left lower extremity and the foot was definitely smaller than the right.

X-ray films of the spine showed an extensive separation of the two halves of the vertebral bodies beginning at D8 and extending throughout the lumbar area, with the hemi-vertebrae widely separated in the midportion. At the level of L2 there was a spicule of bone extending posteriorly into the soft tissue (Fig. 6A). (This spicule of bone was absent on the postoperative films.)

Operation. On Nov. 14, 1949, under ether anesthesia, an elliptical transverse incision was made around the mass. A normal spinous process and lamina was exposed above this mass and the dissection was then carried down to where a lipomatous mass was seen coming out of a bony defect. Further dissection revealed a bony mass projecting posteriorly from the anterior part of the bony canal and this was splitting the dural sac. The area of doubling of the spinal cord around the bony mass was about an inch in length; above and below this point the dural sac appeared normal. The dura on the right was firm and to all appearances normal and therefore was not opened. The lipomatous mass was coming only from the dural sac on the left. On dissecting this further, nerve roots could be seen coming from a large mass of rudimentary cord substance infiltrated by fat. Just above the midline bony mass there was a midline dehiscence covered by a soft fluctuant mass which undoubtedly was retroperitoneal tissue. The bony mass was then excised so that it would no longer keep the main dural sac impaled upon it. CSF came only from the area of the lipoma of the cord, which was partially excised. It was impossible to get good fascial flaps, only muscle flaps and subcutaneous tissue being available for closure.

Postoperatively her course was uneventful except that it was necessary to aspirate CSF which collected beneath the wound for about 2 weeks. Four months after operation the involved foot appeared to have normal strength. The child, now 8 months old, is beginning to stand.

Comment. Diplomyelia or diastematomyelia is a term applied to apparent doubling of the spinal cord. The x-ray picture of this condition was recently pointed out to us by Dr. E. B. Neuhauser. Herren and Edwards in a review of the literature found 42 cases in which there was a doubling of the cord, which they believed was due to partial twinning. They found that it occurred in all areas of the cord and was found at all ages of life. Diplomyelia may in rare instances be present unassociated with a bony defect. In a quarter of the cases there was an osseous or cartilaginous mass in the midline separating the two cords. They suggested that this is also a manifestation of partial twinning. Diplomyelia becomes of surgical importance only when mesodermal derivatives, such as bone, ligaments, or cartilage, become secondarily involved in the lesion and anchor the cord, producing a mechanical hindrance to its ascent. This case also shows a marked similarity to the case reported by Saunders in which a dorsal intestinal herniation associated with diplomyelia was verified by autopsy (Fig. 6B).

This is the third case of this type that we have confirmed by operation in the past 2 years. Pickles in a review of the literature found the total number of reported cases including his own to be 47; 6 had been found at operation and 5 of these patients survived.
Case 7. A. K., a 6-day-old girl, was admitted on Dec. 6, 1947 because of a large mass over the sacral region extending from the sacral region through the posterior part of the perineum (Fig. 7A). It was noted that the coccyx could be felt fairly distinctly giving the impression that it was being pushed backward. The mass was 30 cm. in circumference and did not transilluminate. The anus was situated at the anterior margin of the mass, being drawn up to some extent into its wall. The anus was patulous and appeared to have lost its tone.

X-ray films of the spine and pelvis demonstrated a large soft tissue mass. There has been noted, however, in retrospect a small gas shadow within the mass, which must represent intestine displaced downward and laterally (Fig. 7B).

Operation. On Dec. 8, 1947, under ether anesthesia, a curved incision was made over the sacral swelling. A large cyst containing bloody fluid was entered and it was believed that this must be the sac of a myelomeningocele. The base of the sac was dissected free and was exceedingly wide. Another sac was now entered but no nervous tissue was present. The coccyx and sacrum were pushed upward by the swelling. A mass about 5 cm. in diameter, covered with tissue that looked like peritoneum, was then seen. The mass bulged backward markedly with every respiration and it was still thought to be a myelomeningocele. This sac was opened and intestine could now be seen. It was then realized that this was a posterior herniation which contained lower colon and small intestine. There were three heavy layers of fibrous tissue which had been dissected free which were used to repair the hernia.

The baby’s postoperative course was uneventful and when seen a year later the wound was solid with no pulsation on straining. She was beginning to walk and had normal bowel movements. The anal sphincter was intact (Fig. 7C). Recent checkup examination by Dr. Cooperstock 2 years after operation reveals the patient to be normal in every way.

Histological Examination. The specimen was a trigeminal teratoma with spaces lined by a respiratory type of epithelium, glial tissue, glands and squamous epithelium.

Comment. This was a most unusual condition and we had never encountered it before. The only vertebral defect was the pushing upward and backward of the sacrum and coccyx. The large posterior hernia was either
primarily associated with the teratoma or secondarily acquired by herniation through the interspace described by Ebner (cited by Watson) between the coccyx and ischiococcygeus muscle.

Case 8. E. L., a 2-day-old baby girl, was admitted on April 17, 1942 because of an enormous cystic mass larger than the baby’s head, which came from the sacrococcygeal area. This was attached by a very wide pedicle and over its surface ran large veins. The mass did not pulsate and no calcification could be seen by x-ray studies. The anal sphincter was intact. Because the mass was becoming necrotic, it was deemed advisable to remove it as a life-saving measure (Fig. 8A).

Operation. On April 18, 1942, under ether anesthesia, the enormous mass was circumscribed into healthy skin. The tumor was partially cystic with no evidence of deep infection. It was dissected from the sigmoid colon and rectum and was totally removed. It was seen to have arisen in the sacral region. In the sacrum there was a defect more on the right than on the left, the coccyx itself being absent. The incision below was carried to within 3" of the anal sphincter. Some heavy subcutaneous tissue and some gluteal muscle were brought together over the rectum and sigmoid colon, giving firm support to these structures (Fig. 8B).

Postoperatively the child did remarkably well and is normal in all respects at the age of 7.

Pathological Examination. The removed specimen weighed 1 1/2 lbs. (baby’s weight 8 lbs. before operation). Microscopically it was a trigeminal teratoma which contained cystic spaces lined by columnar epithelium, glial tissue, esophagus, bronchus with cartilage, thyroid, skeletal muscle and squamous epithelium.

Comment. The huge size that these lesions may assume is well shown in Fig. 7. Most observers in the past have believed that these sacral lesions were in all instances vestiges of a reduced parasitic twin. More recently, however, Patten has pointed out another possible origin. The extraordinarily high incidence of trigeminal teratoma in precisely the region where the primitive streak of the embryo was formerly located may well mean that some of these masses originated by neoplastic growth of the tissue of this territory. In the embryo, the region of the primitive streak is a proliferation center from which cells entering all three of the germ layers are derived. It is quite logical, therefore, if it became involved in neoplastic growth that it would give rise to a teratomatous mass in which derivatives
of all three of the germ layers were represented. It should be emphasized that Patten's interpretation does not question the implication of twinning in a certain proportion of sacral teratomata. The possibility of an additional method of origin in this region, however, may well help explain the high incidence of teratomata in the sacral region as compared with other regions in which unequal conjoined twinning might be involved.

These lesions invariably are attached to the posterior surface of the rectum. The surgeon must consequently be warned against opening this structure. One should therefore keep oriented by having the assistant place a finger in the rectum of the infant during the operative procedure.

We have had 19 cases of similar sacral teratomas, only 3 of which were malignant.

SUMMARY

1. Large congenital defects of the scalp should be operated upon as an emergency procedure.
2. Mild surface infection in a myelomeningocele without sphincter paralysis or weakness of the extremities does not contraindicate surgery.
3. A congenital dermal sinus tract excised as a single specimen at operation shows continuity from the surface to the arachnoid and demonstrates the mechanism of the production of meningitis in such cases.
4. Diplomyelia or diastematomyelia is being recognized more frequently clinically. It becomes of surgical importance only when the cord is anchored by a midline bony mass preventing ascent of the cord from its fetal position.
5. Congenital defects of the neuroaxis in infants and children without gross neurological defects which appear hopeless at first are often amenable to operative procedures.

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REFERENCES