Neurenteric cyst with meningomyelocele

Case report

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An 18-month-old girl was born prematurely at 38 weeks. She was the fourth child born to a 36-year-old mother, who had had toxemia of pregnancy.

Examination. Physical examination on the day of birth revealed an alert premature baby weighing 2250 gm. She had a reddish lumbosacral mass resembling invaginated intestine without any apparent orifice, and a soft, thin skin-covered bulge just superior to the mass (Fig. 1). This bulge was fluctuant, and trans-illumination revealed a thick string-like band in the midline just under the skin. The patient moved her legs somewhat weakly in response to pinprick, and her anal reflex was minimal, but she moved her thighs and legs freely with the knees flexed. Lower abdominal compres-
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Fig. 2. Neural arches are widely open and spread apart from the lower lumbar through the sacral levels. At the S-2 segment there is a hemi-vertebral deformity (S), shown more clearly on the sketch (inset).

Fig. 3. Operative photograph showing (A) the neurenteric cyst; (B) the meningocele; (C) the laminal defect; and (D) the paraspinal muscles.

Fig. 4. Photograph (left), and artist’s sketch (right) of sectioned fixed surgical specimen.

sion induced dribbling of urine. There was questionable hypesthesia below the S-3 level. X-ray films of the lumbosacral spine showed widening of the lumbosacral canal, spina bifida of L5-S3, and hemivertebral S-2 (Fig. 2).

Operation. On the day of birth, a circular incision was made around the soft fluctuant mass in the lumbosacral area. The lower ends of the incision stopped just lateral to the lower reddish tumor. The subcutaneous tissue was found to contain normal fatty tissue and a large compressible cyst (Fig. 3). The cyst was dissected, and a tiny neighboring tumor was found in the fatty tissue, which was connected under the skin with the reddish intestine-like tumor. The wall of the cyst merged laterally with the spinal dura, which was opened to reveal lumbosacral fibrous neural elements adherent to the cyst wall containing tortuous small vessels. This cyst was considered to be a meningocele and part of the cyst wall was resected for histological examination. The neural elements were carefully dissected from the wall and were replaced into the spinal canal, and the dura was closed.

The soft reddish mass was cut away from the skin close to its neck, and with the tiny tumor it was followed into the depths of the subcutaneous fatty tissue. There were some crescent-shaped cartilaginous connections to the sacrum, but these masses had no apparent connection with the bone defect of the meningocele at L-5 and S-1. The dermal soft reddish mass with the tiny tumor and subcutaneous fatty tissue containing cartilage were resected as a whole. The paraspinal fascia was reflected to cover the dura at L-5 and S-1, and the wound was closed.

Microscopic Examination. The specimen from the cyst wall showed the typical histology of a meningocele. The main tumor along with the tiny tumor was sectioned to reveal cysts containing slightly cloudy colorless fluid (Fig. 4). The surface of the large cyst wall presenting superficially was composed of colonic intestinal mucosa that was sharply demarcated from epidermal
tissue at the neck (Fig. 5 left). The mucous membrane was supported by connective tissue containing smooth muscle (Fig. 5 right). Deeper, this cyst wall was continuous with the wall of the tiny cyst, the cavity of which did not communicate with the large cyst and was not lined with intestinal mucosa. The underlying tissues were collagenous and fatty, with several small bundles of peripheral nerves and markedly proliferated blood vessels. Cartilaginous tissue was demonstrated in the fibrous region.

Postoperative Course. The postoperative course was uneventful, and the patient did not develop any signs of hydrocephalus. She moved her extremities well despite questionable hypesthesia of her left leg. She had normal bowel movements, and a barium enema failed to reveal any abnormality. Pressure on her lower abdomen no longer produced dribbling of urine, and she was able to urinate well. She was examined 2 years later as an outpatient. She could walk by herself and minimal perianal hypesthesia was noted.

Discussion

The congenital anomalies associated with spina bifida and/or meningomyelocle include heart disease, intestinal malformation, cleft palate, craniosynostosis, umbilical and inguinal hernia, congenital dislocation of the hips, club feet, skeletal deformities such as the Klippel-Feil syndrome and Sprengel's deformity, pilonidal sinus and genitourinary anomalies. Cutaneous angioma and lipoma are also frequently associated with meningomyelocle.

Cases of meningomyelocle with a neurenteric cyst covered by intestinal mucosa on the skin are not often seen. A few such cases were reported by Bentley and Smith. The differentiation of this cyst from dermoid is easy, because its intestinal mucosa is endodermal. Differentiation from teratoma is difficult because the mass was composed not only of intestinal mucosa but also of fatty tissue, collagenous fibers, peripheral nerves, and cartilaginous bone. Willis defined a
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Teratoma as "a true tumor or neoplasm composed of multiple tissues of a kind foreign to the part in which it arises." Rhaney and Barclay discussed enterogenous cysts associated with spinal abnormalities and concluded that these intraspinal cysts are not true teratomas, since they have neither local neoplastic features nor any tendency to metastasize. We agree with their view, because these anomalies are not single encapsulated tumors, and their origin can be explained in terms of embryonic development. Bentley and Smith proposed the concept of split notochord syndrome for the anomaly, known to occur in developing embryos. They claimed that the yolk sac or gut anlage endoderm could herniate through the separated notochord and adhere to the dorsal ectoderm or skin anlage. Beardmore and Wiglesworth suggested a local ento-ectodermal adhesion as the primary event. Prop. et al. extended their view to the second phase of notochordal growth and suggested that the adhesion might be located in the primitive streak. Although there are some differences in the primary event, their explanations seem to have in common the concepts of ontogenetic alteration, notochordal cleft, separation of the neural tube, and ectodermal-endodermal connections in some stages.

Postvertebral intestinal remnants are subdivided into fistulas, sinuses, diverticula, and cysts by Bentley and Smith. In our case, unlike theirs, no obvious fibrous band was found connecting the intestinal remnants to the cleft of the sacral lamina, and the exposed enteric mucous surface had no orifice. The postoperative barium enema failed to reveal any abnormality. Therefore, postvertebral intestinal cyst might be an appropriate name for our case. One special feature of this case is the duplication of the cyst. This has been described only rarely and can be explained by any of the following possibilities: 1) occlusion of the embryogenic gut and faulty process of canalization or vacuolization; 2) two adjacent ento-ectodermal adhesions; and 3) development of a side-tube from a single diverticulum.

On the basis of our findings and the developmental possibilities, our case can be explained by the following process. The ventrally situated yolk sac or gut anlage endoderm herniated through the notochordal
cleft which had developed in some ontogenetic alteration, and eventually adhered to the dorsal ectoderm or skin anlage and ruptured to produce a fistula. Subsequent differential growth of the embryo closed and obliterated part of the fistula to duplicate the lumen. The external enteric canal then invaginated through the fistulous orifice to expose the mucosal surface, and the mucosa of the closed internal enteric cavity degenerated (Fig. 6).

Spinal and central nervous malformations are commonly associated with enteric cysts. The meningomyelocele is one of them, and its etiology is not well understood. According to Matson, the basic causes include failure of vertebral fusion or neural groove, local herniation, and abnormal proliferative overgrowth of neural elements in the embryo. We think that the association of meningomyelocele and neurenteric cyst must be explained by a single event. Patten stated that overgrowth of the neural plate in the early stages of its development might be a factor in the genesis of meningomyelocele. This overgrowth was induced by a neural tube defect that was experimentally produced in the chick embryo prior to establishment of the spina bifida. If one considers the ectodermal-endodermal connections of a neurenteric cyst which traverses the spinal canal, the intimate association of the cyst with a meningomyelocele can be easily understood.

References


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