Atypical Myelomeningocele with Associated Anomalies
Case Report

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The occurrence of a large skin-covered lumbar mass in a newborn presents a challenge in diagnosis and management. Such possibilities as dermoid, teratoma, myelomeningocele, and others must be considered. We have recently encountered a case of myelomeningocele with associated anomalies which proved to be quite bizarre.

Case Report

The patient was a 1-day-old white girl, born of a full-term pregnancy to a 36-year-old mother, gravida V, para V, via elective caesarean section which was carried out for suspected, but unverified, placenta previa. The prenatal and family histories were normal.

Examination. Physical examination revealed an alert, well-nourished, well-developed infant with a large pedunculated skin-covered lumbar mass (Fig. 1). Firm elements were palpable. A midline dimple, with what appeared to be the outlet of a dural sinus tract, was visible just inferior to the mass. The patient moved her legs minimally in response to pinprick. Her hips were flexed. There was no clonus and reflexes were symmetrically depressed. There was a questionable sensory level at mid-abdomen, anteriorly. X-rays of the lumbar spine showed widening of the lumbar canal and a large soft-tissue mass overlying the lumbosacral region (Fig. 2).

Operation. At the age of 3 days, a midline incision was made from the lower thoracic region to the mid-sacral area, bisecting the well-covered mass. The lower end of the incision stopped just cephalad to the midline dimple. The subcutaneous tissue was found to contain a mass of fat, of granular consistency. This was separated from the overlying skin; the dome of the mass was quite firm.

Incision into the dome revealed a large, flat, curved piece of cartilage (Fig. 3 left and right). Beneath this was found a pad of velvety tissue forming the roof of a cyst. Within the wall of the cyst was a vascular pedicle between the halves of a diplomyelia. Part of the vascular pedicle seemed to be derived from abnormal vasculature seen on the cord surface (Fig. 4). The walls of the cyst were merged with dura laterally, arachnoid from the posterior cord surface, and fibrous tissue from the center of the bifid cord elements. Careful dissection and stimulation revealed neural elements within the sac. Laminectomy was not necessary as the posterior lumbar vertebral elements were widely spread into a spina bifida. The sac was carefully dissected free from its central dural and vascular attachments. The dural edges were closed. Paraspinal muscle fascia was used to cover the dura.

Microscopic Examination. The specimen showed cystic cavities lined by neuropil. No ependymal cells were evident (Fig. 5). No mucosal epithelium or other entodermal derivatives were seen. A cavity with dense fibrous tissue walls was also noted. Numerous vascular channels were apparent within the walls of the cyst. The cartilaginous plate appeared to be histologically normal cartilage.
The velvety plate beneath it was composed of chondroblastic tissue, differentiating into cartilage. Histopathologically the findings were consistent with a diagnosis of myelomeningocele.

**Postoperative Course.** The course was remarkable in that the child did not develop hydrocephalus. Although the poor sphincter tone has persisted, there has been improvement in neurological status with hip, thigh, leg, and foot movements occurring both in response to painful stimulation, and spontaneously.

**Discussion**

The association of myelomeningocele with other abnormalities is well recognized. Abnormal lipomatous tissue in the region of the meningocele, hydrocephalus, diplomyelia, abnormalities in the posterior fossa such as the Arnold-Chiari malformation, and varying degrees of spina bifida are encountered. A number of unusual and interesting cases were found in the literature, but we were seldom able to find a case of myelomeningocele as bizarre as this one. A more descriptive name for the mass we found would be the cumbersome angio-chondro-lipo-mye- lomeningocele in association with spina bifida, diplomyelia, and dermal sinus tract.

The major differential diagnoses included teratoma, dermoid, and perhaps an ectopic twin. No hair, sebaceous glands, nor skin tissue were found within the mass. The similarity of the cartilage to a piece of ectopic pelvis was considered. However, the pelvis normally develops in sections.

The possibility of a teratoma was considered. Several reported cases of teratoma...
were reminiscent as they contained similar tissues, such as cartilage and fat.\textsuperscript{8,9,16,18,25} However, they also contained columnar or squamous epithelial-lined cavities as well as other entodermal tissues (respiratory, gastrointestinal tissues, thyroid).\textsuperscript{17,26} Our case contained only mesodermal and ectodermal structures. Although we do not feel our case represents a teratoma, there may be developmental similarities. The etiology of teratomas has been thought to be a congenital defect arising from an “imperfect separation” of the neural tube from the entodermal tube at an early embryonic stage and has been discussed as a neuroenteric tube, entodermal-ectodermal adhesive syndrome, or a split notocord syndrome.\textsuperscript{3,4,6,11,20,22,23}

Myelomeningocele is often thought to be a consequence of fusion failure of midline structures. Another theory is that this is a consequence of local tissue overgrowth.\textsuperscript{5,24} Rather than this sort of origin we feel that our patient’s defect may have arisen from a hydromyelia, as suggested by Gardner, in association with “imperfect separation” of the neural tube from the skin.\textsuperscript{12} We think this may well represent a progressive cystic dilation of the central canal of the cord.\textsuperscript{2,18} The absence of ependyma is not inconsistent.\textsuperscript{24} This dilating cord might have become partially adherent to the skin and subcutaneous tissues; in effect, a reverse dermoid. Rather than skin elements being enclosed within the spinal canal, expanding cord elements have become partially enclosed and adherent to the skin. The tissues involved are all native to the area. The diagnosis of myelomeningocele remains the most likely.

**Summary**

We have presented a case of a newborn infant with a congenital lumbosacral mass. This appears to have been a bizarre myelomeningocele in association with a vascular malformation of the cord, spina bifida, diplomyelia and a dermal sinus tract. We have briefly discussed the pathology and etiology, and reviewed reports of comparable cases.
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References