Epidermoid cyst occurring within a lumbosacral myelomeningocele

Case report

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The authors report the case of an infant who presented with an epidermoid tumor contained in a lumbosacral myelomeningocele. The association of spina bifida aperta and congenital intraspinal tumors is rare. Only two cases of teratoma and two cases of epidermoid tumors arising within a meningocele have been documented previously.

KEY WORDS □ congenital intraspinal tumor □ epidermoid tumor □ meningocele □ myelomeningocele □ spina bifida □ spinal dysraphism

INTRASPINAL neoplasm of cutaneous origin associated with a midline closure defect is a well documented occurrence. Some of these anomalies include spina bifida occulta, cutaneous dimples, angiomas, nevi, or hypertricosis. Nevertheless the concurrence of the common myelomeningocele with congenital intraspinal neoplasms is a much rarer event. To our knowledge, only two teratomas1,4 and two epidermoid tumors3,5 related to cystic spina bifida have been reported previously.

Case Report

This infant was admitted to the hospital on the day of his birth, June 26, 1982, for treatment of a lumbosacral myelomeningocele. Prenatal and family histories were irrelevant to the case.

Examination. The child weighed 2750 gm, and his head circumference measured 35 cm. He had a skin-covered myelomeningocele in the lumbosacral region, 5 × 4 × 3 cm in size. The child was able to move his feet, although they were slightly hypotonic. Both ankle reflexes were absent and his anal sphincter showed an almost normal tone. The rest of the examination was normal.

Routine analyses were within normal limits, X-ray films of the lumbosacral spine disclosed a widened spinal canal. Head circumference started to increase very rapidly up to 39 cm. A computerized tomography (CT) scan showed enlarged lateral and third ventricles. On July 21, a ventriculoperitoneal shunt (VP) was placed. The child was discharged 1 week later, leaving the meningocele repair for a second admission.

The patient was readmitted on August 1, because of anorexia and low-grade fever. Coagulase-negative Staphylococcus was cultured from the cerebrospinal fluid (CSF) obtained through the shunt reservoir. He was treated with systemic and intraventricular antibiotics.

Operation. On November 11, when the CSF had become sterile, the initial shunt was removed and a new VP valve was inserted. At the same time, the myelomeningocele was repaired. The meningeal sac was opened and the neural plaque and related roots were dissected and preserved. A small pearly tumor was found adherent to the surface of the neural plaque. The neoplasm measured 5 × 4 × 3 mm and was removed in toto without difficulty. The meningocele repair was undertaken in the usual manner. Subsequently, the boy required a new revision of the shunt device because of persistent valve infection. He was discharged on January 17, 1983.

Postoperative Course. The infant has been followed closely, and repeated CSF samples have been sterile. He is doing very well from the neurological point of view.
Histological Examination. The cutaneous coverings of the myelomeningocele showed a well structured lining consisting of squamous epithelium. The underlying dermis was in contact with leptomeningeal layers and islands of neural tissue (Fig. 1 left). The neoplasm consisted of a cyst lined by keratinized squamous epithelium, which surrounded non-nucleated keratin lamellae (Fig. 1 right).

Discussion

Congenital tumors include any tumor that exists at birth. The following criteria have been proposed to be applicable to congenital tumors of the spinal cord: 1) tumors of confirmed congenital origin: these tumors are symptomatic at birth or during the neonatal period; 2) tumors with probable congenital origin: these tumors are manifest during the 1st year of life; and 3) tumors with possible congenital origin: these tumors are detected after the 1st year of life, but the initial symptoms can be traced to the neonatal period.

There is another group of neoplasms that, because of their histological findings, are considered to be present at birth even if they are detected later in infancy, childhood, or adulthood. They include epidermoid tumors, dermoid tumors, teratomas, and lipomas, and are considered as congenital tumors of the spinal cord in the pathological sense. In this group several cutaneous anomalies have been found associated with intraspinal neoplasms; namely, dermal sinus, hypertricosis, skin nevi, and angiomas.

Spina bifida cystica is a frequent form of spinal dysraphism. Nevertheless, the concurrence of a congenital neoplasm within the spinal canal associated with myelomeningocele seems to be a very rare event. A search in the current literature disclosed only four cases of congenital intraspinal tumors occurring in association with cystic spina bifida. The patients reported by Arnold and Mitgang had teratomas, and those reported by Kirsch and Hodges and Muscatello were the only patients with both epidermoid tumors and myelomeningoceles.

The origin of dermoid and epidermoid cysts is essentially the same. If invagination of skin elements into the neural canal should occur early in embryonic life, a dermoid tumor will result; if this occurs at a later stage, an epidermoid tumor will develop. In the present case, we can disregard a traumatic origin of the epidermal inclusion cyst, since no spinal punctures were performed at any time (all CSF samples were collected via the shunt reservoir). Regarding the temporal relationship of both the tumor and myelomeningocele, they can both be traced to the posterior neuropore closure, at the 25-somite stage, at about the 4th week of embryonic life.

The etiology of spina bifida remains uncertain, and we cannot relate the two fortuitously associated anomalies to any identifiable factor. Probably the concurrence of an epidermoid cyst within the spinal canal and myelomeningocele is just a mere coincidence in time and place. Nevertheless, it is tempting to assign a common embryological origin for both events.
Epidermoid cyst within a myelomeningocele

References


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