Vernix caseosa meningitis and laryngeal stridor in an infant with myelomeningocele

Case report

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The authors present the case of an infant with laryngeal stridor, apneic spells, and an open lumbar myelomeningocele; granulomatous vernix caseosa meningitis was found at autopsy. They suggest that chemical meningitis which also involved the cranial nerves might be held partly responsible for the laryngeal stridor.

KEY WORDS • chemical meningitis • stridor • myelomeningocele

We have found little in the medical literature on the chemical meningitis occurring in infants with myelomeningocele. Vocal cord paralysis is a well-known complication of this malformation. We are reporting a case of granulomatous vernix caseosa meningitis in an infant with myelomeningocele and laryngeal stridor.

Case Report

This baby boy was delivered at 39 weeks' gestation by assisted breech, following premature rupture of the membranes and a 3-hour labor without evidence of fetal distress. He weighed 2640 gm at birth, and was flaccid and apneic. He had a myelomeningocele, 2.5 cm in diameter, located at the L3-4 level; fluid gushed from the anomaly as soon as it was exposed. The infant improved with tracheal intubation and intermittent oxygen, but on extubation, hoarseness and an inspiratory stridor were present, and lethargy persisted. An apneic spell occurred at 9 hours of life.

Examination. The patient was admitted to the University Hospital at 11 hours of age; he displayed hypotonia, a decreased but symmetrical Moro's reflex, a decreased but noticeable sucking reflex, and symmetrical facial and eyelid movements. A marked high-pitched inspiratory stridor and bilateral inspiratory rhonchi and coarse rales were heard. There was some spontaneous leg movement, and right knee jerk and ankle jerks were present. Cremasteric reflex, anal tone, and spontaneous voiding were noted. The head circumference was 31.5 cm (10th percentile); the anterior fontanel measured 2 × 2 cm, with the sagittal and metopic sutures measuring 1 cm across. Skull transillumination was negative.

Periodic apneic spells necessitated reintubation. On laryngoscopy the cords closed very slightly on expiration but did not abduct on inspiration. Palate and pharyngeal
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movements and the trachoebronchial tree were normal. Chest films showed minimal lower lobe consolidation; skull films were normal.

Laboratory Studies. The hemogram was normal. Blood gas studies initially showed a pO2 of 37 mm Hg, pCO2 of 29 mm Hg, pH of 7.36, and subsequently a recurrent metabolic acidosis. Serum calcium initially was 3.3 mEq/l and subsequently was normal. Unexplained hyponatremia, with a sodium of 120 mEq/l on the third day, occurred but was later corrected to normal. Cultures from the myelomeningocele and cerebrospinal fluid (CSF) leakage, umbilicus, stool, and throat were negative.

Course. Treatment consisted of support with intravenously administered 10% dextrose, sodium bicarbonate and electrolytes, calcium supplements, nasotracheal intubation, and antibiotics. Steroids were given initially for possible laryngeal edema. However, the apneic episodes subsequently recurred and ventilatory assistance and oxygen were given. The myelomeningocele was covered with sterile Vaseline gauze.

On the third day the lesion was repaired, the nerve roots into the spinal canal were freed, and a good dural closure achieved. Postoperatively the apneic spells became more frequent and oropharyngeal secretions increased. Laryngoscopy on the sixth day of life showed pharyngeal paralysis, not present 2 days before. Arching of the back with extension of the neck and occasional spasms of the legs were noted the day after surgery, and bacterial meningitis was suspected. The bladder was distended, requiring manual expression of urine. Apneic episodes became more prolonged and the patient died on the ninth day.

Postmortem Examination. Widespread acute bronchopneumonia was found at autopsy, along with minimal bilateral subdural hemorrhagic fluid. The ventricular system was slightly dilated and the cerebral aqueduct was stenotic but patent.

The brain weighed 340 gm after fixation. The cerebellum was grossly malformed, very hypoplastic and asymmetrical, with the left hemisphere smaller than the right (Fig. 1). The vermis was replaced by a grayish membrane. The pons was small in proportion to the mesencephalon, and its brachia were hypoplastic; from the rostral cross sections, it was slightly rotated counterclockwise on its axis as a result of the cerebellar asymmetry. The surface of the brain stem and the cerebellum was grayish, soft, and studded with multiple rounded whitish elevations of 1 to 2 mm. The spinal cord was hydromyelic and showed similar small grayish lesions throughout, particularly in the lumbar segments. No change of this nature was found in supratentorial or ventricular areas.

Microscopic study of the myelomeningocele sac revealed acute and chronic inflammatory change, with foreign body giant cell reaction, and an abundance of keratinous debris and lanugo hairs. The epidermal lining was desquamated in some areas.

The leptomeninges covering the cerebellum, brain stem, and spinal cord, as well as the cranial and spinal nerve roots, showed a severe inflammatory reaction, with foreign body giant cells, lymphocytes, and macrophages laden with a greenish or yellowish pigment (Fig. 2). Abundant keratinous debris and some lanugo hairs were present in the subarachnoid space. Marked tissue edema and some neuronal shrinkage were seen in the pons, medulla oblongata, and spinal cord. A moderate hydromyelic cavity was found at all levels of the spinal cord (Fig. 3). The pia-arachnoid over the cerebral hemispheres and the lining of the ventricles were not involved.

FIG. 1. Basal and right lateral view of the hypoplastic cerebellum and brain stem. Note flecks of whitish keratotic material in the leptomeninges (arrows).
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**FIG. 2.** Histological section of left cerebello-pontine angle showing laminated keratin debris and granulomatous reaction of pia-arachnoid with foreign body giant cells. H & E, X 180.

**FIG. 3.** Transverse section through hydromyelic spinal cord at lumbar level. Keratinized cells (X), some arranged in laminated layers (arrows) are seen in the subdural space. H & E, X 40.

**Discussion**

Granulomatous chemical meningitis is a known complication of the spillage of keratinous debris and fatty contents of dermoid cysts into the subarachnoid space. It takes the form of acute and relapsing episodes, or of a chronic process imitating a degenerative disease of the brain and spinal cord. Extensive radiculitis and involvement of cranial nerves has been reported; infiltration of the vagus nerves caused vocal cord paralysis in one case.

The term "vernicomyelia" was coined to indicate the invasion of the subarachnoid space and the spinal cord by vernix caseosa. The inflammatory lesions described were of foreign body type, often with superimposed bacterial meningitis. Six of the 24 cases reported were in children within the first 3 months of life. The authors did not mention whether the myelomeningocele sacs were open at birth or if the lesions extended into the intracranial compartment, but penetration into the ventricles and the cerebral substance by keratinous debris has been mentioned elsewhere.

The time of closure of myelomeningocele sacs or the CSF dynamics in the fetus with spina bifida aperta is not known. Intrauterine pressures of 4 to 80 mm Hg occur during pregnancy and labor, and probably the fetal movements or the compression of the engaged head create pressure differentials between the cranial and spinal CSF compartments, and therefore cause tidal fluid movements. The stresses in the birth canal, or even the local scrubbing of a myelomeningocele sac could propel epidermal debris into the CSF spaces.

Since keratinization and desquamation increase rapidly toward the end of pregnancy, this type of meningitis would be more likely to develop in the perinatal period. The difficulties of obtaining adequate samples of...
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CSF and excluding the commonly present bacterial infection may preclude an early diagnosis of chemical meningitis as well as the possible application of recommended steroid treatment. The contribution of this type of meningitis to the scarring of the leptomeninges and thereby to the problem of hydrocephalus in spina bifida deserves further consideration.

Deficit of the third and sixth through twelfth cranial nerves has been reported in association with myelomeningocele and the Arnold-Chiari malformation. Descent and funneling of the brain stem and cerebellum supposedly cause traction, compression, and vascular and CSF obstruction. The associated anomalies also include dysplasia of those structures, cellular depopulation of the lower cranial nerve nuclei, and shrinkage of the nerve cells, lepto- and pachymeningeal scarring, and adhesions to the nervous tissue. The pressure caused by hydrocephalus and foraminal constriction has been proven to be material but not essential to the cranial nerve impairments, which may be variably relieved by CSF shunts and decompression of the posterior fossa. Vocal cord paralysis usually occurs when the hydrocephalus is clinically established, but occasionally stridor appears from birth; in that case it could be wrongly attributed to trauma, particularly following endotracheal intubation. Other local causes might be considered in the differential diagnosis, but abductor paralysis is the usual endoscopic finding in the cases of an underlying nerve deficit. In the latter case tracheostomy is often indicated, as the prognosis is unpredictable.

In their detailed compilation of reported cases, Schwalbe and Gredig emphasized that the Arnold malformation, namely, cerebellar dysplasia and herniation into the spinal canal, and that of Chiari, with additional dorsal pontomedullary herniation and kinking, each constitutes a wide spectrum of congenital anomalies, not coexisting to the same degree in every case. In our patient the marked cerebellar and a less severe pontile hypoplasia, the aqueductal stenosis and the slight hydrocephalus, constitute some common features with the Arnold-Chiari malformation. The lack of herniation at the level of the foramen magnum would cause our case to fall into the category of Type V as defined by Schwalbe and Gredig; however in the original publication of Chiari in 1895, cerebellar hypoplasia without caudal displacement and herniation was defined as Type IV of the malformation. Although the hypoplasia and distortion of brain stem were by themselves sufficient for explanation of the laryngeal stridor, additional irritation of cranial nerves caused by aseptic meningitis might have played a contributory role in the paralysis of vocal cords of our patient. We suggest, therefore, that vernix caseosa meningitis should be considered in the differential diagnosis as an uncommon cause of laryngeal stridor in newborns with open myelomeningocele.

References

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