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Basioccipital Meningocele

Carlos R. Martinez,¹ J. Michael Hemphill,²
 Fred J. Hodges III,^{1,3} Bob W. Gayler,¹
 George T. Nager,⁴ Donlin M. Long,⁵ and
 John M. Freeman²

Cephaloceles—developmental transcranial meningoencephaloceles and meningoceles—are not rare. The reported incidence varies between one and three cases per 10,000 births [1–3]. Most cephaloceles occur in the occipital region, involving the supraoccipital or, less commonly, the interparietal parts of the occipital bone [3–5]. Sincipital cephaloceles are less common, and basal cephaloceles are rare [6–10]. We believe the small meningocele through the occipital part of the clivus in an infant with recurrent bacterial meningitis reported here is unique.

Case Report

A white infant boy had two episodes of bacterial meningitis during the first 6 months of his life. After the third episode of meningitis, at age 7 months, a thorough diagnostic evaluation was carried out at another hospital. Physical examination was normal except for a small hairy area in the low lumbar region. Serum biochemical and immunologic studies were normal, as were radiographs of the skull and the entire spine. Polytomograms of the cribiform plate and the temporal bones were reported to be normal. Computed tomography of the head was normal. Metrizamide (Amipaque) myelography and surgical exploration of the lumbar region, at age 9 months, revealed a small arachnoid cyst at the L2–L3 level. No other abnormalities were found.

A few months later two more episodes of meningitis prompted his referral to the Johns Hopkins Hospital. Repeat lumbar myelography disclosed no abnormalities except for post operative changes. Repeat exploration of the low lumbar region disclosed a 2 mm dural defect at the L3 level which was obliterated. The patient did well for 4 months. Three more bouts of meningitis followed and he was readmitted. The temporal bone polytomograms from 14 months earlier were reviewed; they disclosed a 5 mm round defect with slightly sclerotic margins in the clivus, located 1 cm below the sphenoccipital synchondrosis. Repeat polytomograms in lateral (fig. 1A) and frontal (fig. 1B) projections confirmed the presence of the defect. Nasopharyngeal soft tissues were normal. Iophendylate (Pantopaque) myelography demonstrated a small outpouching of

the subarachnoid space extending to the defect in the clivus (fig. 1C). Since several episodes of bacterial meningitis had been associated with upper respiratory infections, the clivus was explored.

During operation, there was a sudden leak of cerebrospinal fluid as the lower part of the adenoids was incised. Further dissection allowed visualization of a circular defect in the clivus which contained an extension of the subarachnoid space in a small fibrous sac; this was resected and the defect closed. Biopsies demonstrated a cystlike structure with chronic inflammation and scarring. A few epithelioid cells were believed to be consistent with histiocytes. No definite arachnoid tissue could be identified. The post-operative course was benign and the patient has done well and been free of meningitis during the 18 months since operation.

Discussion

The term cephalocele is used to indicate herniation of meninges or meninges and brain substance through a defect in the cranium. A meningocele contains only meninges, while a meningoencephalocele consists of herniated meninges and brain tissue [3, 6, 11]. This distinction, however, is not maintained consistently through the literature, in part due to the difficulty in deciding in certain cases whether or not brain tissue is present. In this case, chronic inflammation and scarring made the histologic interpretation difficult. No definite arachnoid or nervous tissue was identified. However, the myelogram and the surgical findings provided compelling evidence that the sac in the bony defect was an extension of the subarachnoid space.

A unique feature of this case is the location of the bony defect. We did not find a single case of a transclival cephalocele in the English radiologic or neurosurgical literature. Cronin and Penoff [12] reported a case of a meningocele involving the anterior margin of the foramen magnum and the anterior arch of C1 in a patient with multiple anomalies of the upper cervical vertebrae and bilateral complete clefts of the primary and secondary palate.

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¹ Russel H. Morgan Department of Radiology and Radiological Science, Johns Hopkins Medical Institutions, 600 N. Wolfe St., Baltimore, MD 21205. Address reprint requests to C. R. Martinez.

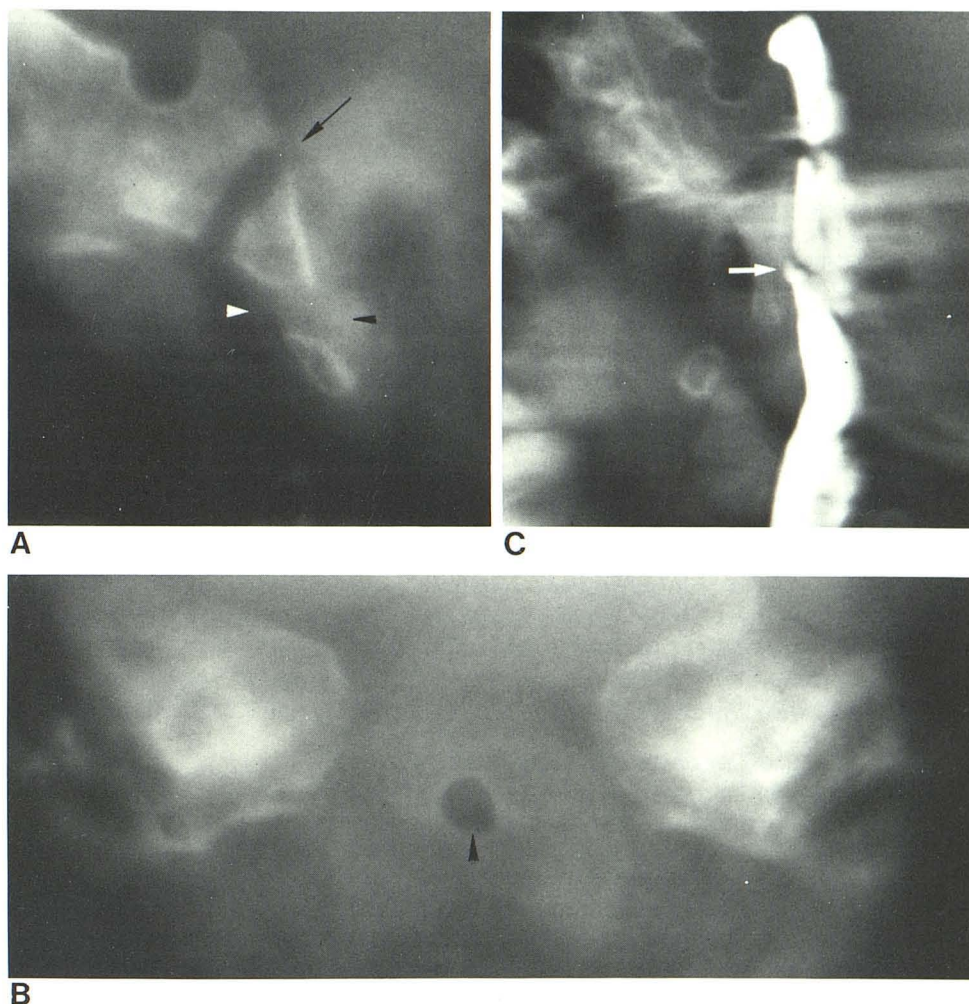
²Department of Neurology, Johns Hopkins Medical Institutions, Baltimore, MD 21205.

³Present address: Edward Mallinckrodt Institute of Radiology, Washington University School of Medicine, St. Louis, MO 63110.

⁴Department of Otolaryngology, Johns Hopkins Medical Institutions, Baltimore, MD 21205.

⁵Department of Neurosurgery, Johns Hopkins Medical Institutions, Baltimore, MD 21205.

Fig. 1A.—**A**, Lateral hypocycloidal tomogram of clivus. Defect (arrowheads) about 1 cm below sphenoccipital synchondrosis (arrow). **B**, Anteroposterior view. Midline position and sharply defined sclerotic margins of defect (arrowhead). **C**, Midline linear tomogram of clivus during myelogram. Filling of small sac (arrow) that extends to defect in clivus.



Several theories have been proposed to explain the formation of cephaloceles [1, 3], and a review of them is beyond the scope of this report. However, considering the embryologic development of the basicranium [13–16], it is possible to speculate that a focal failure of chondrification of the basal plate or failure of subsequent ossification results in a bony defect through which the meninges herniate. Defects in ossification of the basiocciput may indeed result in abnormal configuration of the foramen magnum, such as the keyhole defect illustrated by Di Chiro and Anderson [17]. Similarly, transverse or longitudinal fissures may be found in the occiput [17, 18]. However, to our knowledge, these anomalies have not been associated with herniation of the meninges.

It is also interesting to note that in the newborn cranium, the vestige of the notochord can be found in the midline of the clivus [16]. It is, however, smaller than the defect in our case, and in cross section is not circular. Therefore, persistence of the notochordal channel seems an unlikely explanation for the defect seen in our case.

The importance of the radiographic evaluation in children with recurrent meningitis is apparent in this case. In addition to routine views of the skull, including basal projections, polytomography may be indispensable for complete evalu-

ation of the cribriform plate, the temporal bones, and the base of the skull.

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