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Skull-Base Meningoencephalocele Presenting as a Unilateral Neck Mass in a Neonate

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Summary: A unique skull-base meningoencephalocele presenting as a neck mass in a neonate is reported, and skull-base encephaloceles are reviewed.

Index terms: Brain, hernia; Skull, abnormalities and anomalies; Skull, base; Infants, newborn

Case Report

A term neonate was noted at birth to have a soft, mobile, 8-cm mass in the right side of the neck inferior to the mandibular angle. The neonate, who was otherwise healthy, had several episodes of apnea related to airway obstruction by the mass.

A contrast-enhanced computed tomography (CT) scan showed a large complex cystic and solid neck mass that involved the right carotid, retropharyngeal, and parotid spaces (Fig 1A) and widened the ipsilateral jugular foramen (Fig 1A). Focal calcification was seen within the mass (Fig 1B). The mass extended into the middle of the side of the neck. A limited magnetic resonance (MR) examination showed the mass to be predominantly isointense with gray matter on T1- (Fig 1C) and T2-weighted images. The brain on MR imaging was normal with no dysmorphic structures.

Subtotal excision of the mass was performed. The lesion was firmly attached to the skull base and intimately associated with the internal carotid artery. The pathologic specimen contained meninges, glial tissue, choroid plexus, and ependyma consistent with a meningoencephalocele (Fig 2A and B).

The child has done well with partial resolution of ipsilateral cranial-nerve deficits involving nerves IX, X, and XI.

Discussion

The jugular foramen is between the petrous portion of the temporal bone and occipital bone, and originates from persistence of the embryologic foramen lacerum posterius, the space between the basiocciput and the auditory capsule (1). Cephaloceles usually protrude from the skull along a suture line or at the junction of several bones (2). The connection with the cranial fossa can be broad based or pedunculated (3). Encephaloceles may contain recognizable organized protruded brain tissue or, as in this case, more disorganized neural tissue with remnants of ependyma, choroid plexus, or glial tissue (3).

Meningoencephaloceles of the skull base are generally classified into five major anatomic types: frontoethmoidal, sphenoorbital, sphenomaxillary, temporal, and nasopharyngeal (2, 4). The basal nasopharyngeal encephaloceles are further subdivided into the transethmoid, sphenoethmoid, sphenonasopharyngeal, and basioccipital-nasopharyngeal subtypes (2). Rare reports of encephaloceles herniating through defects in the tegmen of the petrous bone, greater sphenoid wing, or center of the basilar portion of the occipital bone have been described (2, 5, 6). Our reported case, with involvement of the jugular foramen, does not classically fit into the above classification scheme.

There is no known cause for meningoencephaloceles. Basal meningoencephaloceles are the rarest form, occurring with an approximate frequency of 1 in 35 000 births (7). Basal encephaloceles are commonly occult and therefore might not be diagnosed until adulthood. Encephaloceles occasionally are associated with malformation syndromes, cleft palate, craniostenosis, agenesis of the corpus callo-

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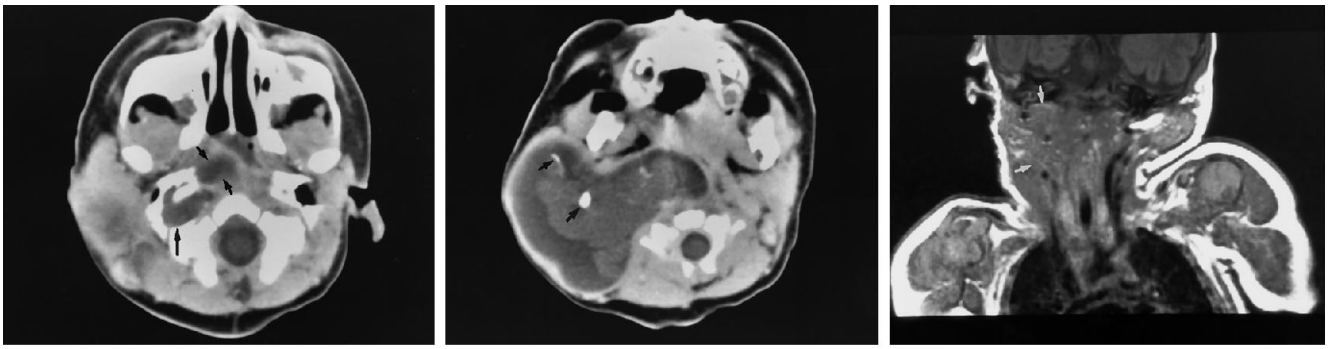

Fig 1. *A*, Contrast-enhanced CT scan shows the low-density neck mass originating at the level of the widened right jugular foramen (*large arrow*). The lesion involves the retropharyngeal space (*small arrows*) and the right side of the neck.

B, CT scan shows punctate calcification within the mass (*arrows*) and extension into the neck on the right side.

C, Coronal T1-weighted MR image (600/20 [repetition time/echo time]) shows the neck mass (*arrows*) is isointense with gray matter.

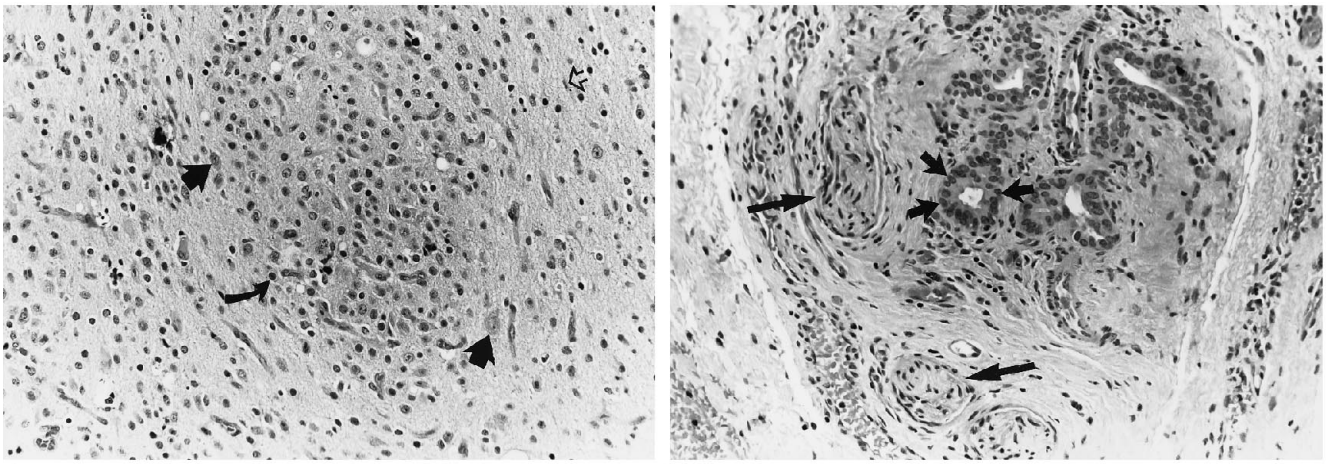

Fig 2. *A*, Histologically, the majority of the specimen consists of cerebral gray matter containing neurons (*solid straight arrows*), oligodendroglia (*curved arrow*), and microglia (*open arrow*).

B, In some areas, peripheral nerve (*long arrows*) and ependymal cells (*short arrows*) are present.

sum, and brain-stem deformities (3). The basal cephaloceles, in particular, are more likely to be associated with midline cerebral anomalies (8). In general, the cephaloceles with better prognoses are less than 5 cm in diameter, do not contain dysplastic tissue, and are not associated with hydrocephalus or anomalies (9). Known complications of basal cephaloceles include local mass effect, cerebrospinal fluid leak, and increased risk for central nervous system infection (2).

Common neck masses in infants include cystic hygroma, hemangioma/lymphangioma, and branchial cleft cyst. None originate from or commonly involve the jugular foramen. These lesions should be easily differentiated from skull-base encephaloceles on the basis of typical CT or MR appearance and location.

In conclusion, skull-base encephaloceles are rare, but should be considered in the differential diagnosis of a neck mass presenting in a neonate, especially if the mass clearly herniates through a basal foramen or suture.

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