Clival encephalocele

Case illustration

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The clinical presentation and anatomy of this congenital defect approximate findings associated with transsphenoidal encephalocele, a subset of midline fusion anomalies that present with respiratory distress, recurrent meningitis, and cerebrospinal fluid (CSF) rhinorrhea. Preoperative imaging, however, documents a defect in the clivus with herniation of neural tissue into the nasopharynx. This is the first report of a midline fusion defect of the clivus with an associated encephalocele.

Two days after birth, this infant girl suffered respiratory distress. Transnasal endoscopic surgery established a diagnosis of basal encephalocele. Seven years later she suffered recurrent bouts of meningitis. Examination revealed no evidence of CSF rhinorrhea. Magnetic resonance (MR) imaging revealed tissue extending from the rostral portion of the clivus to the nasopharynx (Fig. 1). Computerized tomography (CT) scanning of the head revealed that the defect was posterior to the sella and separated from the sphenoid sinus by a thin rim of cortical bone (Fig. 2). At surgery, the distal knob of the encephalocele was exposed in the nasopharynx, using a sublabial–transseptal approach. The posterior wall of the sphenoid sinus (anterior wall of the clivus) was thin, covering the underlying encephalocele. The bone was removed, the stalk was mobilized and secured with an aneurysm clip (Fig. 3), and the distal encephalocele was resected. The resulting defect and the sphenoid sinus were packed with abdominal fat.

References