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Basal Transsphenoidal Encephalocele: MR Findings

Basal encephaloceles are the rarest form of encephalocele, and transsphenoidal encephaloceles (TSE) represent the rarest form of basal encephalocele [1–10]. The incidence of TSE is estimated to be 1/700,000 live births [1].

Case Report

A 15-year-old girl presented for evaluation of short stature and delayed sexual maturation. Her weight and height were 91 lb. (41.3 kg) and 58 in. (147.3 cm), respectively. She had not yet begun menstruating and had had no development of secondary sexual characteristics. Moderate ocular hypertelorism was present. A brain CT scan was initially interpreted as showing an intrasellar and suprasellar lucent nonenhancing mass (Figs. 1A and 1B). Subsequent MR studies (Figs. 1C and 1D) revealed a transsphenoidal encephalocele.

Discussion

Transsphenoidal encephaloceles develop as a result of defective ossification of the body of the sphenoid bone with subsequent persistence of the craniopharyngeal canal, which normally closes by day 50 of gestation [2, 3]. This permits prolapse of the anterior inferior third ventricle, including hypothalamus and optic chiasm, inferiorly through the sphenoidal bone and into the epipharynx just behind the nasal septum. In rare cases, the encephalocele is wholly contained within the sphenoid bone. Associated findings include agenesis of the corpus callosum, hypertelorism, endocrine dysfunction, airway obstruction, CSF rhinorrhea, chiasmatic syndromes, and various optic and mid-face abnormalities [1–10].

In neonates and children presenting with TSE, meningitis caused by CSF rhinorrhea is a potential problem; facial and cerebral malformations are more likely to be evident [1]. Patients who reach adulthood with occult TSE are more likely to present with endocrine dysfunction or chiasmatic syndromes [1].

Surgery has been recommended for patients with TSE who develop rhinorrhea or who have progressive visual deficits. Surgical results, however, have not always been beneficial to the patients. In part this may be due to the incomplete assessment of the contents of the encephalocele afforded by CT, angiography, and pneumoencephalography (PEG).

In the evaluation of basal encephalocele, MR provides considerably more information than traditional neurodiagnostic techniques. Associated brain anomalies, aberrant vascular structures, and the intrinsic anatomic relationships within the encephalocele are displayed more clearly. Natural tissue contrast between flowing blood, CSF, and brain tissue on MR images obviates injection of contrast material. The position of the intracranial optic nerves, chiasm, and tracts can be assessed even more accurately than they can be with detailed PEG. In several of the 25 cases reported in the literature, surgeons have performed transcranial surgery without knowing the position of the chiasm [2, 4, 7, 8]. In the case of persistent CSF rhinorrhea or progressive visual loss, surgery may be necessary; MR seems to provide the most accurate road map for this delicate intervention.

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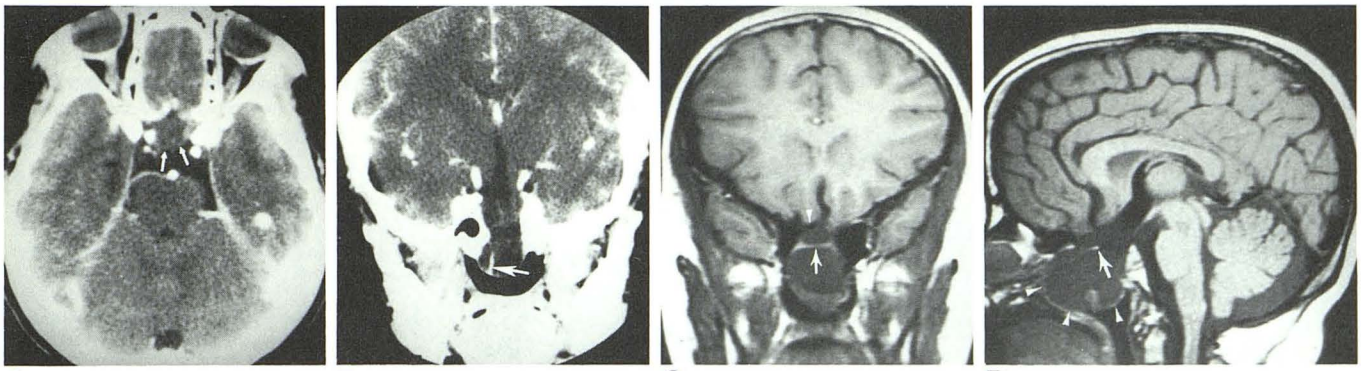

Fig. 1.—*A*, Axial enhanced CT shows a lesion of soft-tissue density within sella, slightly denser than CSF (*arrows*).
***B*, Coronal enhanced CT image shows herniation of third ventricle inferiorly through sphenoid bone into epipharynx. Enhancement of infundibular stalk can be seen inferiorly in encephalocele (*white arrow*).**
***C*, Coronal MR (SE 800/20) shows optic chiasm (*arrow*) displaced inferiorly within encephalocele. Lateral to chiasm and encephalocele wall, internal carotid arteries and aerated sphenoid bone can be seen as regions of signal void. A1 segment of right anterior cerebral artery lies just above encephalocele (*arrowhead*).**
***D*, Sagittal MR (SE 800/20) shows inferior extension of third ventricle into encephalocele. Ptotic optic chiasm (*arrow*) is somewhat thin. Encephalocele bulges into epipharynx (*arrowheads*).**