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Cystic Temporofacial Brain Heterotopia

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Brain heterotopia is a rare developmental anomaly that has been reported to occur in the pharynx, orbit, nasal cavity, and maxillofacial region [1–11]. Cystic temporofacial brain heterotopia has been described in only four previous reports [1, 7, 8, 11]. We present a fifth case, review the pertinent literature, and discuss optimal methods of preoperative diagnosis.

Case Report

A full-term male neonate presented with a large left preauricular and maxillofacial mass. Initial clinical diagnosis was cystic hygroma. Preoperative evaluation included sonography, CT, and multiplanar MR; MR and CT revealed a large, unilocular, cystic mass in the left infratemporal fossa; the cyst was up to 4 cm in diameter (Figs. 1A–1D). There was extensive facial and calvarial deformity, including upward and medial bowing of the left lateral orbital wall, displacement and thinning of the left zygomatic arch, and upward and medial bowing of the middle cranial fossa (Figs. 1C and 1D). CT at the skull base showed mild enlargement of the left foramen ovale and a small left parasellar lipoma, but no communication between the cyst and the intracranial contents could be established with high-resolution multiplanar MR (Figs. 1A and 1B). The final preoperative impression was that of a large parotid duct cyst; surgery was performed on the third day of life.

The surgical specimen consisted of a collapsed cystic structure; its maximum dimension was 4–5 cm, and it had no organized internal architecture. Microscopic examination revealed disorganized neural tissue including glial elements and neurons. Small clefts within the lesion were lined with low cuboidal epithelium consistent with ependyma. Rare papillary epithelial areas suggestive of choroid plexus were also present. Immunoperoxidase stain for glial fibrillary acidic protein (GFAP) was positive in the neural tissue. Additionally, the epithelial-like areas also stained positively for GFAP, confirming ependymal differentiation in these cells (Fig. 1E). Dense fibrous tissue was present in the wall of the cyst, but there were no other tissues present to warrant a diagnosis of teratoma. Final pathologic diagnosis was CNS heterotopia.

Discussion

Cystic temporofacial brain heterotopia is an extremely rare lesion that has been described in only four previous reports [1, 7, 8, 11]. These lesions vary primarily in the degree of intracranial-extracerebral connection. The cases presented by Lasjaunias et al. [8] and Robbins et al. [11] are similar to our case in this respect. In these cases, no connection was shown between the cyst and the intracranial contents. Misra et al. [1] reported a similar brain cyst, but the cyst was associated with a bony dehiscence of the middle cranial fossa on the side of the lesion. In our case, axial CT disclosed mild enlargement of the left foramen ovale (Fig. 1D), and an abnormal left parasellar fat deposit was apparent on MR (Fig. 1B). However, the large extracranial cystic mass did not have intracranial extension. In one case of intracranial, extracerebral glioneural heterotopia (reported by Wakai et al. [7]), the heterotopic brain cyst was primarily located intracranially with a tonguelike extension into the oropharynx through a dehiscent foramen ovale. The leptomeninges overlying the brain were intact, and the mass was partially encased in its own dural covering. These authors note that the palatine shelves fuse during the eighth to ninth weeks of fetal life and suggest that the oropharyngeal component of the mass disturbed the fusing process before the ninth week of fetal life. Although any conclusion at this point is speculative, all these cases (Wakai et al. [7], Misra et al. [1], our own case reported here, Lasjaunias et al. [8], and Robbins et al. [11]) appear to form a continuum with respect to the degree of intracranial-extracerebral connection. This is reminiscent of the apparent continuum between nasal encephalocele and nasal glioma. The most commonly accepted theory of the pathogenesis of nasal glioma is that it represents brain substance isolated from the intracranial contents during sutural closure and is therefore of developmental rather than neoplastic origin [2]. Similar considerations may apply to the general class of cystic brain heterotopias, since in all cases the relative integrity of the bony middle cranial fossa in the region of the foramen ovale appears to be directly related to the degree of separation of these masses from the brain and meninges. Further support for these concepts is provided by recently described transalar sphenoidal encephaloceles [12]. There is clearly a close analogy between nasal encephalocele vis-à-vis nasal glioma and transalar sphenoidal encephalocele vis-à-vis infratemporal fossa brain heterotopia.

The differential diagnosis of large temporofacial cystic lesions occurring in a neonate includes cystic hygroma/lym-

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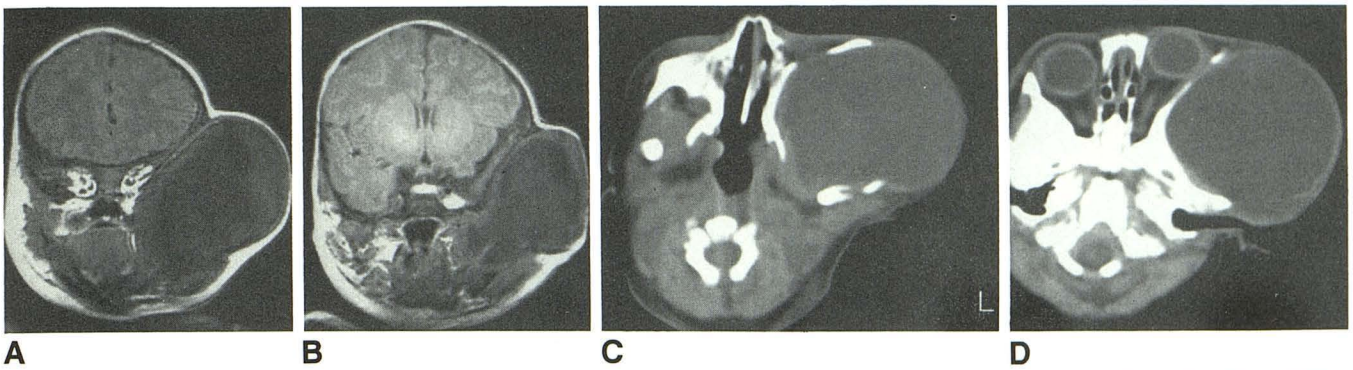

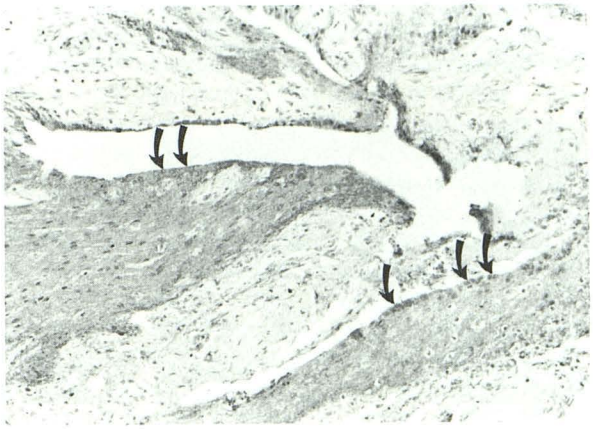

Fig. 1.—*A*, Coronal T1-weighted SE 800/20 MR shows large, extracranial, fluid-filled mass with upward and medial bowing of the left middle cranial fossa and left retrobulbar fat.

***B*, More posterior section shows mass encroaching on parapharyngeal space and additional deformity of left middle cranial fossa. Also note hyperintense fat collection in parasellar region. Fluid-filled mass does not communicate with intracranial contents. Posteriorly, there was deformity of petrous portion of left temporal bone, but encapsulated mass remained separate from the intracranial contents.**

***C* and *D*, Unenhanced axial CT shows a large fluid-filled mass in the left infratemporal fossa with extensive bony deformity of the left maxilla, mandible, and orbit. Note posteromedial displacement of left parapharyngeal fat (*C*) and slight enlargement of left foramen ovale (*D*).**

***E*, Disorganized neural tissue (*stained dark*) intermingled with fibrous tissue (*pale*). Focal clefts are lined by cuboidal or columnar ependymal cells (*arrows*) that arise from looser fibrillary glial background to line spaces within this cystic heterotopia. (glial-fibrillary-acidic-protein [GFAP] immunoperoxidase stain, original magnification ×32)**

phangioma, teratoma, encephalocele, parotid duct atresia, and (rarely) brain heterotopia. In order to establish the optimal surgical approach to such lesions, it is necessary to exclude an intradural component which, if present, would require a craniotomy for curative resection. Therefore, the preoperative neuroradiologic evaluation of such lesions should include plain radiography, CT, and multiplanar MR. Plain radiography and CT show calvarial defects associated with encephalocele and some cases of cystic brain heterotopia. High-contrast, multiplanar MR shows separation of such a maxillofacial mass from the brain and meninges. Percutaneous cyst puncture and drainage may confirm that the fluid composition is equivalent to CSF but does not in itself imply an intradural extension of the mass, since these lesions may contain actively secreting choroid plexus [8]. Selective internal and external carotid angiograms are optional studies that can rigorously exclude pial and/or dural connections and intrinsic lesion hypervascularity [8].

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