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Short Report

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Intranasal Encephalocele: A Case Report

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Congenital midline nasal masses are rare anomalies. Nasal cerebral heterotopias (nasal gliomas) and nasal encephaloceles have an actual or potential central nervous system connection (1,2), and represent rare inborn malformations of the central nervous system (3,4).

Nasal encephaloceles are herniations of the intracranial contents through a defect in the anterior skull base (1,3,5). Guthrie and Dott (6) suggested that these lesions are not true cerebral herniations. It was concluded that the meningeal and brain protrusions exist first and that the bony defect is formed later (7). Nasal encephaloceles can be divided into two main groups: frontoethmoidal and basal encephaloceles (5). Basal encephaloceles can be classified as transethmoidal, sphenoethmoidal, transsphenoidal and frontosphenoidal (1). Frontoethmoidal and basal encephaloceles are very rare (1:5,000) (3,5).

The pathogenesis of encephaloceles may be explained by a disturbance in the separation of the surface ectoderm and neurectoderm in the midline just after closure of the neural folds. It should be regarded as a "late" neurulation defect taking place during the 4th gestational week. Apoptosis appears to be related to this separation process. Diagnostic CT or MR imaging delineates the anatomy of the herniated mass (3,4,6).

Case Report

A 24-month–old girl was referred to our hospital for treatment of a soft mass on the bridge of her nose and noisy and somewhat labored respiration. She had been suffering from sleep apnea attacks.

Physical examination disclosed a 1.5 cm in diameter spheric mass in the midline of the nose that extended to the intranasal compartment. It was pulsatile and increased on crying. Sagittal cranial MRI showed a subcutaneous mass on the nose and a cystic mass in the nose filled with a cerebrospinal fluid-like material, without any intracranial extension or evidence of defect in the base of the skull (Fig. 1).

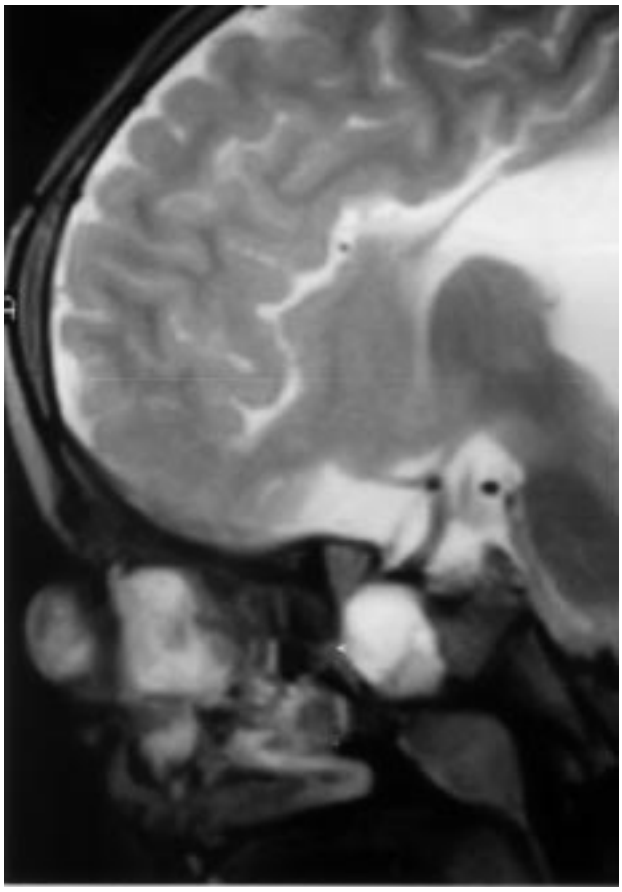

Figure 1. T2- weighted sagittal cranial MRI showed a subcutaneous mass on the nose and a cystic mass in the nose filled with a cerebrospinal fluid-like material, without any intracranial extension or evidence of defect in the base of the skull.

The tumor was approached through a lateral rhinotomy. The lateral rhinotomy exposed an unencapsulated subcutaneous mass that extended intranasally through a gap between the nasal bone and the upper lateral cartilage. As the mass was attached to the inferior turbinate bone submucosally, it was removed en bloc along with the inferior turbinate bone after the cystic material was aspirated. The cribriform plate was exposed by lateral and transverse osteotomy by lifting the nasal bones medially on a hinge. We detected a channel-like duct between the mass and cribriform plate intraoperatively; this had not been detected on cranial MRI, and it was ligated and sutured.

Macroscopically, the tumor proved to be a nasal encephalocele. The patient did not develop postoperative CSF rhinorrhea and was discharged home on the 5th postoperative day. Histological examination revealed nontumoral but disorganized nervous tissue (Fig. 2). Immunohistochemical staining confirmed the neural nature of these elements showing positivity for glial fibrillary acidic protein (GFAP) (Fig. 3). Her symptoms disappeared within 2 months. A control MRI performed 6 months later was normal (Fig. 4).

Encephaloceles may be congenital in origin and represent a primary anomaly of the neural tube and its skeletal cover. Each of these lesions may be associated with bony cranial defects and intracranial abnormalities (8). One third of spontaneous encephaloceles occur through multiple defects (9). Acquired encephaloceles may be traumatic (5) or iatrogenic e.g., following sinonasal surgery. The condition requires the presence of bone and dural defects associated with increased intracranial pressure (1). Intracranial or subarachnoid communication cannot be demonstrated if the intracranial

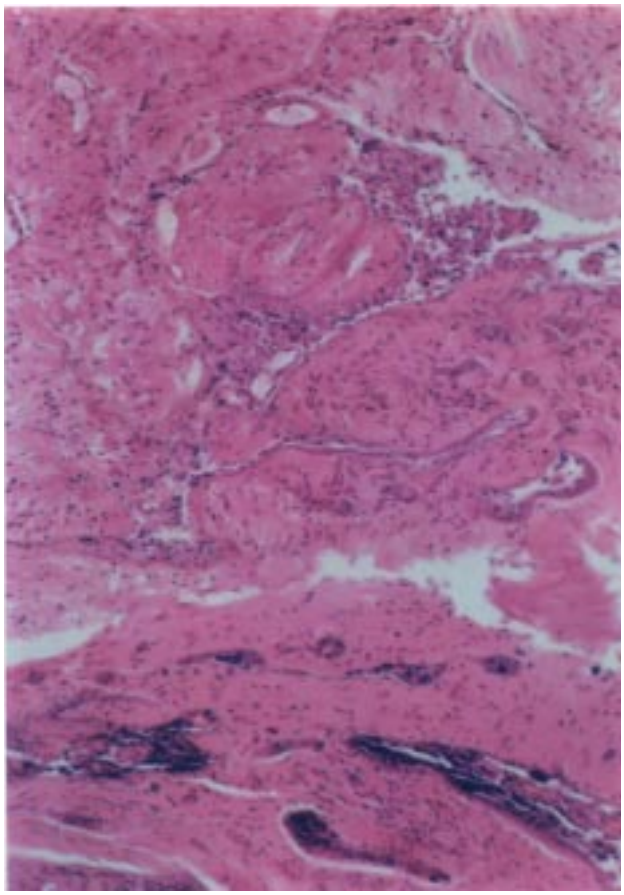

Figure 2. Histological examination reveals disorganized nervous tissue remnants that consist of eosinophylic glial tissue strips intermingled with other connective tissue samples (H&E x 100).

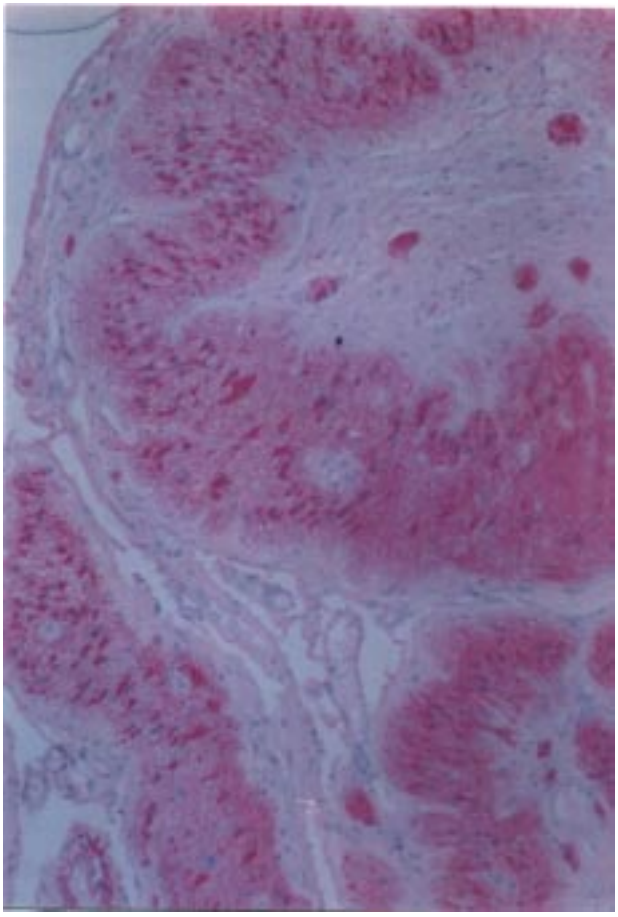

Figure 3. Immunohistochemically, there is strong reactivity for glial fibrillary acidic protein of the glial bundles and rudimentary glial tissue remnants are seen (GFAP x 100).

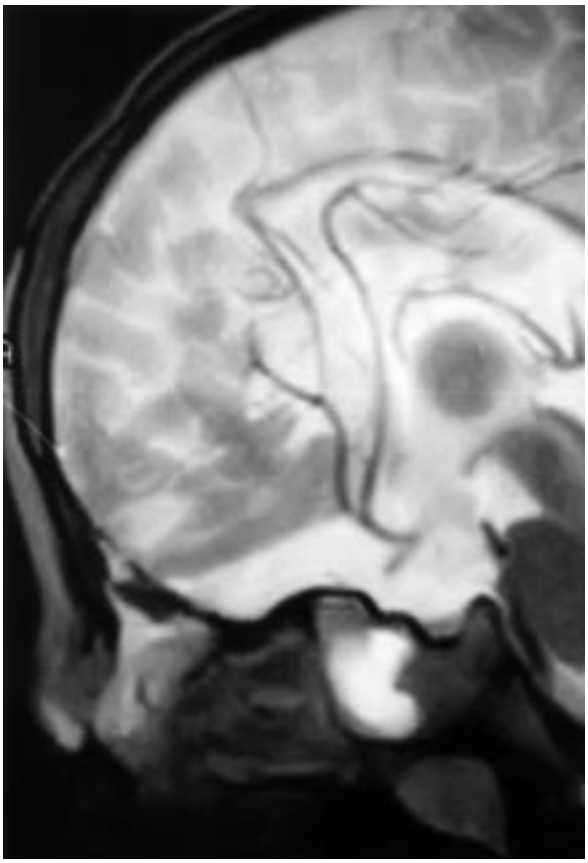

Figure 4. A check-up MRI performed 6 months later was normal.

connection is lost or very small. A pedicle directly connecting the neuroglial tissue with the subarachnoid or ventricular spaces may become detached and eventually absorbed or vestigial (10). The incidence of bony defects noted is as high as 21% (11). A connecting duct was found between the anterior cranial fossa and nasal mass in our patient.

Encephaloceles occur in one out of every 4000 live births. They are classified as sincipital when extranasal near the glabella, forehead, orbit or suboccipital region. Primary encephaloceles are usually present in infancy or early childhood, and are possibly associated with a history of meningitis. They appear as a soft, pulsatile, bluish mass near the glabella and may be transilluminated. An intranasal encephalocele presents with a mass in the nose; it may be mistaken for a nasal polyp and removed with disastrous results. The Furstenberg test will be positive in cases of encephalocele, and X ray films of the base of the skull and CT scans usually show a bone defect (11). We did not observe abone defect on radiologic examination, but one was observed intraoperatively in our case.

Encephaloceles are classified according to their contents and location. Those containing only cerebrospinal fluid and meninges are termed craniomeningoceles; if the lesion also contains neural tissue then they are termed meningoencephaloceles. Nasal encephaloceles can cause complex deformities of the naso-orbital skeleton. As the encephalocele pushes through a defect in the facial skeleton, it causes lateral displacement of the medial orbital walls. Correction of this skeletal deformity is necessary to achieve a normal facial appearance (12).

Frontoethmoidal encephaloceles can be recognized as a facial mass covered with normal skin, while basal encephaloceles may cause nasal obstruction or symptoms related to herniation of basal structures. Therapy for frontoethmoidal encephaloceles consists of excision of the tumor, watertight closure of the dural defect and reconstruction of the skull defect (5).

Traditionally, encephaloceles have been treated neurosurgically via a transcranial approach associated with the possible morbidity of a loss of sense of smell, post-operative intracerebral hemorrhage, cerebral edema, epilepsy, frontal lobe dysfunction with memory and concentration deficits (11,12). The advent of endoscopic sinus surgery has allowed a new intranasal approach for the treatment of basal encephaloceles, minimizing patient morbidity (1).

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