Nasal cerebral heterotopia: nasal atretic cephalocele

J.F. Martínez-Lage; J.D. García-Contreras*; B. Ferri-Ñiguez** y J. Sola**


Summary

We report the case of a 4-year-old boy who presented with a congenital mass on the bridge of his nose. A magnetic resonance study failed to rule out a communication of the tumor with the cranial cavity. The lesion was totally removed. Histopathological study of the excised mass showed a peripheral zone of fibrous-connective tissue with a core of glial and neuronal elements. We discuss the origin of these masses and their relation to nasal cephaloceles. We suggest that this type of lesions should be included within the broader spectrum of atretic cephaloceles.


Heterotopia cerebral nasal: cefalocele atrésico nasal

Resumen

Se presenta el caso de un niño de 4 años con una tumoralción congénita en la raíz nasal. La resonancia magnética no pudo descartar comunicación de la lesión con la cavidad craneal. La lesión fue extirpada en su totalidad. El estudio anatomo-patológico de la tumoralción mostró una zona periférica de tejido fibro-nectivo y un centro compuesto de células gliales y neuronas. Se comentá el origen de estos tumores y su relación con el encefalocele nasal. Sugerimos que este tipo de lesión debería incluirse dentro del amplio grupo del cefalocele atrésico.


Introduction

Cephaloceles are congenital herniations of intracranial tissues through a skull defect. Cranial meningoceles contain only cerebrospinal fluid within the meningeal sac. When the malformation includes neural tissue it is termed an encephalocele. Atretic cephalocele refers to a skin-covered subscalp lesion containing meninges and remnants of neuronal or glial tissues. This latter type of lesion has also been named rudimentary, abortive, or occult cephalocele. The significante of abortive cephaloceles depends on its frequent association with "occult" cerebral malformations. So-called nasal glioma is a mass of mature glial tissue arising on the bridge of the nose. There has been some controversy regarding the exactness of the term glioma, as these masses are not considered true tumors but hamartomatous lesions.

We report on a child harboring one of these nasal tumors. In view of the histopathological features of this lesion, which contained neurons and glia, we believe they must share a common origin with rudimentary cephaloceles and suggest they should be termed nasal atretic cephaloceles.

Case report

A 4-year-old boy was referred to our hospital for treatment of a mass on the bridge of his nose, which was noted at birth. Apparently, the tumor had grown recently but caused no symptoms in the child. Past and family history were unremarkable.

On examination, there was a firm tumor, 2 cm in diameter, arising on the bridge of the child's nose and which extended slightly to the left. The mass was covered by normal skin and did not tense up with Valsalva's maneuvers. The child's head size and neurological examination were normal for his age. An ophthalmologic study failed to disclose any ocular abnormality.

Magnetic resonante imaging showed that the nasal tumor was isointense in relation to the brain (Fig. 1). The possibility of intracranial penetration could not be ruled out with this study.

Operation. A transverse skin incision was made and a 2-cm firm nodule, which excavated the nasal bones, was
totally removed. The mass was continuous with a fibrous stalk, which was divided at its insertion in the uppermost portion of the nasal cavity. Postoperative recovery was uneventful; the child being discharged two days after surgery. After a follow-up period of 1 year, the boy was asymptomatic and there was no recurrence.

Histopathological study of the removed tissues showed a firm broad-based tumor, which had a gray-yellowish appearance. Microscopically, there was a fibrous-connec-
tive covering with a central portion of disorganized cerebral tissue made up of streaks of mature glial cells and scarce neurons (Fig. 2). Immunohistochemical staining confirmed the neural nature of these elements showing positive for protein S-100 (Fig. 3). The connective component was enhanced with Masson’s trichromic stain.

**Discussion**

Several types of congenital lesions may arise on the bridge of the nose, namely nasal encephalocele, dermoid cysts and nasal glioma. Dermoid cysts are congenital tumors histologically composed of dermal and epidermal elements. Their origin is attributed to a faulty separation of the cutaneous ectoderm and the neuroectoderm with the subsequent sequestration of skin cells. This process is traced back to week 3-4 of embryonic life. Occasionally, these tumors are connected to the skin surface by a dermal sinus. Rarely, a skin orifice continues with a dermal sinus and ends within the cranial cavity.

Nasal gliomas also originate on the bridge of the nose. They often present as solid tumors firmly adhered to the nasal bones. Histologically they are composed of mature glial cells and only seldom contain oligodendrocytes, ependymal cells or neurons.

In 1964, McLaurin reported seven children diagnosed with parietal meningocele among a series of 13 patients with parietal cephaloceles. Since then, some authors have reported small, skin-covered midline subscalp nodules or cysts that have been described as abortive cephalocele, rudimentary meningocele, meningeal heterotopias, atypical meningocele, or atretic cephaloceles. These uncommon lesions present in the vicinity of the lambda and contain meninges, neural rests and, frequently, embryonic vessels. The prognosis for these patients does not depend on this inoffensive congenital defect, but rather on the association of "occult" brain anomalies.
Cephaloceles have been reported in only two locations, the parietal and occipital regions. The origin of rudimentary cephaloceles remains obscure. Several pathogenetic mechanisms have been described including regression of a true encephalocele in early fetal life, persistence of the "nuchal bleb", and persistence of neural crest remnants. Marin-Padilla attributes the development of cephaloceles to a defective interposition of mesoderm between the cutaneous ectoderm and the neuroectoderm. Having el. relate the formation of nasal cephaloceles to a failure in the mechanisms of apoptosis that lead to the separation of skin and neuroectoderm at the foramen cecum, and consider these lesions as true neural tube defects. In the case of nasal gliomas the connection between the infra-and extracranial tissues would, with time, become fibrosed or may even completely disappear.

Yeoh el. prefer using the term "nasal cerebral heterotopia" to describe these lesions instead of that of "nasal glioma". They have also documented the histopathological similarities and differences between cephaloceles and heterotopias of the region of the nasion. They arrived at the conclusion that both anomalies are clinically and histologically identical, the only difference being the presence or absence of an intracranial communication. In the view of these authors, so-called nasal glioma and sequestered cephalocele are merely variants of a spectrum of anomalies. They find no pathological criteria that would distinguish nasal cerebral heterotopia from atretic cephalocele were it not for the neurosurgical evidence of a cerebral connection.

From a clinical point of view, removal of the lesion is indicated in most cases. In atretic cephalocele, the fibrous stalk may be divided safely and needs no intracranial approach. Nasal gliomas (nasal cerebral heterotopias) should be completely removed due to their known propensity to recur.

Given the variety of tissues (of glial, meningeal, or neuronal lineage) that may occur within these anomalies, we suggest that they form a continuum, and that they share a common pathogenetic mechanism; namely defective interposition of mesoderm between the fetal skin and the cranial

<table>
<thead>
<tr>
<th>Name of lesion</th>
<th>Tissue(s)</th>
<th>Osseous defect</th>
<th>Referent(s)</th>
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<tbody>
<tr>
<td>Heterotopic cerebral tissue</td>
<td>Neural</td>
<td>(-)</td>
<td>Commens et al.²,</td>
</tr>
<tr>
<td>Ectopic glial tissue</td>
<td>Glia, meninges, vessels</td>
<td>(-)</td>
<td>Orkin &amp; Fischer¹²</td>
</tr>
<tr>
<td>Heterotopic meningeal nodule, epicranial arachnoid cyst</td>
<td>Meningeal (arachnoid)</td>
<td>(-)</td>
<td>Jackson⁷</td>
</tr>
<tr>
<td>Nasal cerebral heterotopia, nasal glioma</td>
<td>Glial, neuronal, meningeal</td>
<td>(+/-)</td>
<td>Bale at al.¹,</td>
</tr>
<tr>
<td>Abortive/ rudimentary/ atretic / sequestrated cephalocele</td>
<td>Neuroglial, meningeal, vessels</td>
<td>(+)</td>
<td>Currarino³,</td>
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<td></td>
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<td>Theaker et al.¹³,</td>
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<td>Hirsch at al.⁵,</td>
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<td>Yeoh at al.¹⁴,</td>
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<td>Drapkin⁴,</td>
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<td>Martinez-Lage et al.⁹,</td>
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<td>Yokota et al.¹⁵</td>
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Fig. 3. Intense staining with protein S-100 of the glial bundles (S-100, 100X).
We summarize in the Table the names and histopathological pattern of several reported masses of heterotopic tissues of the nasal region and scalp. Accordingly, we propose that nasal glioma and other lesions of embryonic ectopic tissues should be included within the category of atretic cephalocele, as it is probable that all of them have a common pathogenetic origin.

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References


