NASAL GLIAL HETEROTOPIA


*ENT Department, **Pediatric Surgery Department, ***Histopathology Department „Sf. Maria” Children Emergency Hospital, Iasi

ABSTRACT

Nervous congenital anomalies including encephaloceles and heterotopic nervous tissue masses (glioma), located in nasal region present some diagnostic and therapeutic particularities because of their potential of intracranial connection. The authors emphasize the importance of preoperative assessment in order to prevent the intracranial complications. The authors report the case of 25 days-old girl with a mass on the dorsum and left wall of the nose, presented since birth. Paraclinical exams, including CT scan, revealed the mass features without an intracranial connection. Surgical excision was performed, the lesion appearing well defined and left nasal bone was found to be eroded, without intranasal extension. Postoperative evolution was favorable. The heterotopic central nervous tissue was confirmed by the histopathology and immunochemistry exams. Although rare, the nervous congenital malformations should be considered in the differential diagnosis of a nasal mass, because it’s possible intracranial connection, requiring a complex pretherapeutic assessment.

Key words: nasal glioma, extranasal heterotopic central nervous system tissue, glial heterotopia.

Introduction

The nervous congenital midline nasal masses including nasal gliomas and encephaloceles are rare anomalies and clinically important because of their potential to connect to the central nervous system, requiring a complex assessment.

Case report

A 25 days-old girl was admitted with a mass on the left side of the nose, presented since birth, without nasal obstruction. There was no other relevant history. Physical exam revealed a 10/15mm, firm, nonpulsatile mass, located on the root and the left side of the nose, without changes on the overlying skin. The mass did not increase in size when the child cried and there was no intranasal mass.

The echography of soft tissues showed a homogenous, well defined subcutaneous mass, without Doppler signal. CT scan revealed a soft tissue mass without an intracranial component or bony defect in the floor of the

Correspondence

Daniela Rusu
ENT Department, “Sfanta Maria” Children Hospital,
62 Vasile Lupu Street, 700309, Iasi, Romania.
anterior cranial fossa (Fig. 1, 2, 3). The patient underwent a surgical excision of the mass which appeared to be subcutaneous and unencapsulated, with a diameter of 15/20 mm, fairly firm. The nasal bone of the left side was found to be eroded without a defect of nasal mucosa and thus, without a tumoral extension in left nasal cavity.

The histopathological exam of the surgical specimen showed nervous tissue with rare neurons and islands of glial cells, surrounded by a fine band of fibrous tissue (Fig. 4, 5, 6).

The patient was well 14 months postoperatively.

Discussion

Encephaloceles and gliomas have a similar embryologic origin but as the encephalocele is a herniation of cranial contents through a defect in the skull, a glioma is thought to be an encephalocele that has lost the intracranial connection. Fifteen percent of gliomas remain attached to dura with a fibrous stalk[2, 3]. Any of these lesions can be associated with a cerebrospinal fluid (CSF) leak so that the biopsy must be avoided because of meningitis risk.

Gliomas are locally aggressive lesions usually presented at birth, and 90% of cases are diagnosed by age of 2 years. Sixty percent of gliomas are extranasal, 30% intranasal and 10% of cases are in both sites, communication of the intranasal components being through a defect in the nasal bone. Extranasal glioma presents as a firm, noncompressible subcutaneous mass, most often located at the nasal dorsum. The mass does not increase in size with crying and do not transilluminate. The overlying skin may have telangiectasias. The intranasal glioma may present with nasal obstruction, epistaxis or nasal deformity and usually arise from the lateral nasal wall (on examination an intranasal encephalocele arises medially). A clinical sign is the absence of expansion or pulsation of the mass following the compression of ipsilateral internal jugular vein (negative Furstenberg test). The distinction between glioma and encephalocele is very important because of the risk of CSF leak and meningitis. Unilateral nasal polyps in children is rare.

Biopsy of a unilateral mass should be avoided until a complex assessment is performed. In our patient the lesion was located extranasally on the root and left side of the nose.

Heterotopic central nervous system tissue may occur at other sites, such as the paranasal sinuses, nasopharynx, tongue, palate, tonsils and orbit and may be referred to as facial glioma[1, 8].

The diagnosis requires radiographic imaging exams (CT and RMN). CT scan reveals mostly bony abnormalities in the floor of the anterior cranial fossa (enlarged foramen cecum, bifidity of crista galli). RMN shows soft tissue details suggesting an intracranial connection.

The differential diagnosis includes developmental anomalies, inflammatory lesions, traumatic deformity, benign and malign tumors.

After a complex assessment, the management of nasal glial heterotopia is complete surgical excision which offers a cure in most cases. Incomplete excision can be accompanied by recurrence (15-30%)[1, 6]. The nervous midline masses which have an intracranial connection require an interdisciplinary approach with a neurosurgeon. A frontal craniotomy is per-
formed, the intracranial lesion is excised and then the dura defect is repaired, and the extracranial mass is removed[7].

The intranasal lesions are approached by lateral rhinotomy or by endoscopic techniques [3, 5]. The extranasal glioma without an intracranial connection may be excised using either a vertical elliptic midline incision or a horizontal incision over the nasal dorsum[2, 7].

The diagnosis of these lesions is confirmed by histopathological exams. The lesion is nonencapsulated, composed of large and small islands of glial cells, in a connective fibrous tissue. The glial tissue can be confirmed by immunoreactivity for glial fibrillary acidic protein (GFAP) or S100 protein[1, 2]. Neurons are rare or absent. Long-standing or recurrent nasal encephalocele tends to contain an excessive fibrous tissue relative to the amount of glial cells and absence of neurons may make it difficult to distinguish from glioma[1, 9].

Conclusions

Any unilateral nasal mass in a child should be evaluated for a congenital midline masses.

The nasal glioma is a congenital CNS malformation in which there is anterior displacement of cerebral tissue that has lost their intracranial connection. This must be distinguished from the encephalocele in order to prevent the risk of cerebrospinal fluid (CSF) leak and meningitis. The diagnosis of nervous congenital midline nasal masses requires a complex assessment which must include CT, RMN. The biopsy of such lesion should not be performed before radiographic imaging scans are obtained. The treatment of these lesions is surgical excision by multiple approaches, including a combined one with a neurosurgeon when the intracranial connection was proved.

REFERENCES