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Nasal glial heterotopia: Four case reports with a review of literature

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ABSTRACT

Background: Heterotopic neuroglial tissue is defined as a mass composed of mature brain tissue isolated from the cranial cavity or spinal canal. The nose and the naso-pharynx are the most common sites of location. Only 264 cases are reported in the world literature. In this study, we present cases of nasal glial heterotopia we treated in our department. The aim of this paper is to study clinicopathological aspects of these cases of nasal glial heterotopia.
Methods: From 2013 to 2018, we included in our department all the babies who suffered from a neuroglial heterotopia in the head and neck region. Clinical and radiological findings of these cases were analyzed. Histopathological examination was the mainstay of diagnosis.
Results: Four cases of glial heterotopia of the nasal cavity were included and presented in this study.
Conclusions: Glial Heterotopias of head and neck are more common in the nasal cavity. Clinical and radiological findings as well as histopathology and immuno-histochemistry are essential to diagnose these lesions.

Introduction

Glial heterotopia is a rare non-hereditary, benign and congenital malformation, which is composed of normal glial tissue isolated from the central nervous system. Nose and nasopharynx are the most common sites involved. The incidence of nasal glioma is one in 20 000–40,000 live births with female preponderance. A total of 264 cases reported in the world literature since the first description by Reid in 1852 [1,2].

In the nasal region, the lesion could be situated external, on the dorsum or on the side of the nose, or could be situated internal in the nasal cavity.

We clinically and histologically analyzed cases of nasal glial heterotopia treated in our department of maxilo-facial surgery.

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Materiel and methods

From 2013 to 2018, we included all the babies who suffered from a neuroglial heterotopia in the head and neck region in our department of maxilo-facial surgery in the "Hôpital-Femme-Mère-Enfant" in Lyon, France. Clinical and radiological findings of these cases were analyzed. Histopathological examination was the mainstay of diagnosis.

Then, we realized a review of the world literature on PubMed concerning this pathology with key-words: "nasal glial heterotopia" or "nasal glioma".

Results

Four cases of nasal glial heterotopia were treated between 2013 and 2018 in our department of maxilo-facial surgery in the "Hôpital-Femme-Mère-Enfant" in Lyon, France. There are two females and two males with a mean age of 2.18 years (range, birth to 4 years). None of the patients had a history of prior surgery to the nasal area or reported a history of trauma. We analyzed clinical symptoms and radiological findings. Details are reported in Table 1.

All patients had a tumor mass. Three cases had no clinical symptoms. Only one (case number 4) had nasal obstruction, chronic otitis media and nasal drainage. The lesions were either extranasal, situated on the nasal dorsum or on the side of the nose (n = 3) (Figs. 1–2) or simultaneously extranasal and intranasal, inside the nasal cavity or paranasal sinuses (n = 1). The relationship between the lesions and the central nervous system was documented by magnetic resonance imaging (Fig. 3) and confirmed during surgery. No radiographic evidence of a bony abnormality in the nasal cavity, paranasal sinuses or base of the skull was documented.

At the time of the surgery, no cerebrospinal fluid leak or sign of bone erosion was noted (Fig. 4). The lesion is removed with caution (Fig. 5) and a microscopic analysis is performed.

Microscopically, all cases consist of unencapsulated mature glial tissue composed of astrocytes and abondant fibrillary background. Without ependymal, choroïd plexus or leptomeningeal component. The glial tissue could be difficult to identify, especially when there was an associated inflammatory component or significant fibrosis. The glial tissue is identified by immunoreactivity for glial fibrillary acidic protein (GFAP) or S100 protein. No Calcification or ependymal cystic degeneration was observed (Fig. 6).

Discussion

Nasal glial heterotopia is a non hereditary, benign, congenital malformation, embryologically related to encephaloceles [3,4]. It is a rare lesion occurring once in 20 000 to 40 000 live births with a total of 264 cases reported in the world literature since the first description by Reid in 1852 [1,2,5].

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 [6,7].

Gliomas are locally aggressive lesions usually present at birth. 90% of cases are diagnosed before the age of 2 years [8].

Usually, nasal glial heterotopia is present at birth as a mass in the nasal region. Three types of clinical presentation were identified: extranasal (60%), intranasal (30%) or both (10%) [9]. Clinically, external nasal gliomas present as masses that do not transiluminate and whose volume remains stable despite crying and efforts. Hypertelorism may be present [10]. Although this lesion has a slow growth rate and is benign without any potential for malignant degeneration, late treatment may result in distortion of the septum and nasal bone or infection.

The differential diagnosis of a congenital nasal midline mass is broad and should include neurogenic tumors, ectodermal tumors and teratomas. Nasal gliomas represent encephaloceles which have lost their intracranial connection; however, a fibrous stalk is found in 15–20% of cases as a relic of this connection. Overall, only 10–15% of gliomas have a connection to the dura. Intranasal gliomas are two to three times as likely to have such a connection compared to their external counterparts [11].

Evaluation of a congenital midline mass include a computed tomography or a magnetic resonance imaging for searching intracranial extension.

Biopsy or fine needle aspiration of nasal masses is contraindicated because of the increased risk of meningitis or perhaps the removal of functional brain material from an encephalocele [3].

The treatment of choice is complete surgical excision. 4–10% cases of post-operative recurrence are to be deplored [12,13]. In our

Table 1
Characteristics of patients.

Case	1	2	3	4
Age	8 months	1 month	4 years	4years
Sexe	Female	Female	Male	Male
Location	External, dorsum	External, dorsum	External, dorsum	External+internal, lateral nose
Size	1cm	1,5cm	2cm	2cm
Nasal obstruction	No	No	No	Yes
Chronic otitis	No	No	No	Yes
Nasal drainage	No	No	No	Yes
Neurospinal fluid	No	No	No	No
Cerebral connection	No	No	No	No

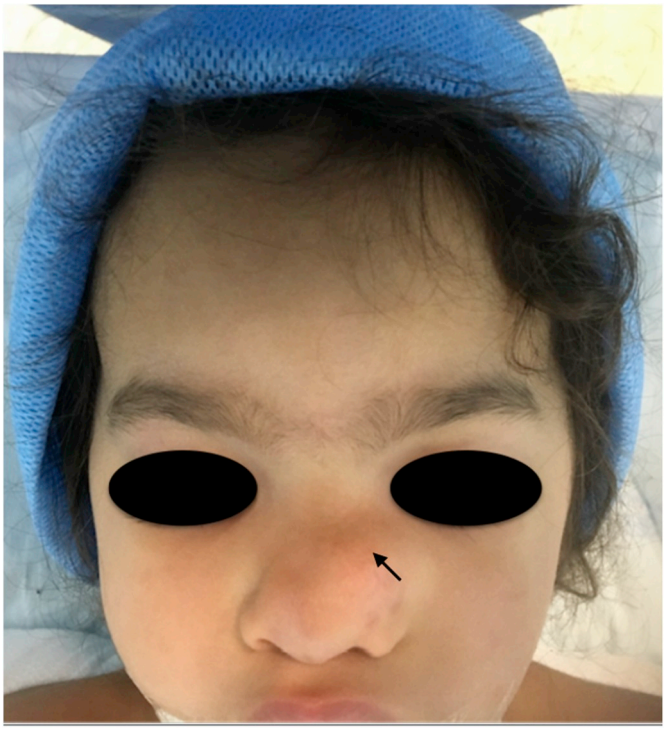

Fig. 1. Full view of the case number 1 at preoperative with an extranasal lesion (→).

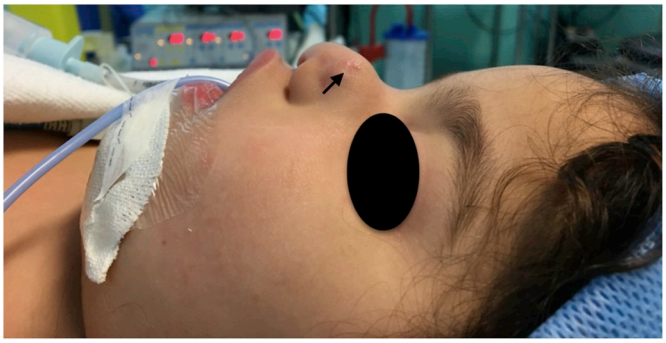

Fig. 2. Profil view of the case number 1 at preoperative with an extranasal lesion (→).

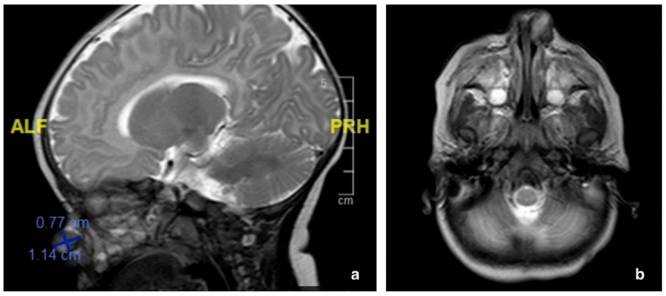

Fig. 3. Magnetic Resonance Imaging of the case number 1: a) sagittal view through the tumor; b) axial view through the tumor.

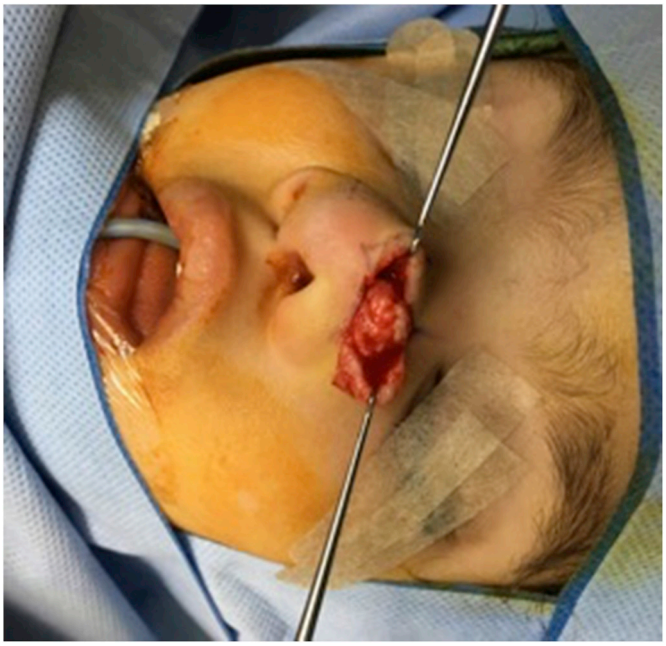

Fig. 4. Intra-operative view of the excision of the lesion of the case number 1.

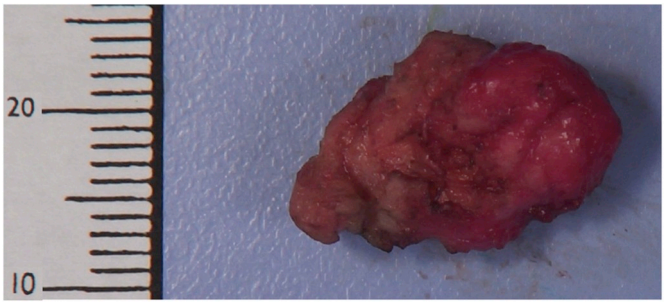

Fig. 5. View of the operative piece.

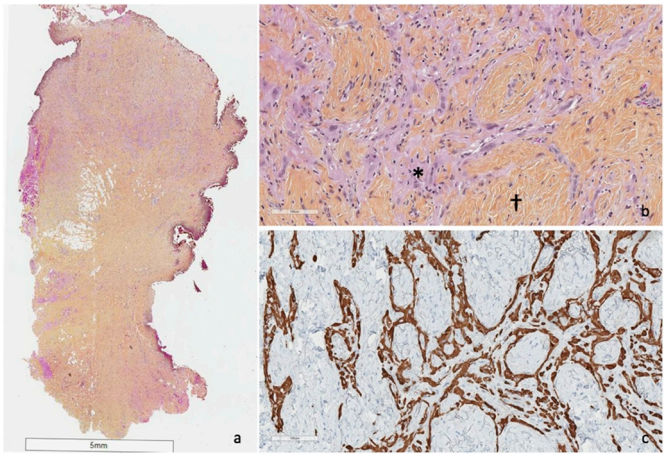

Fig. 6. Histological features of case number 1: a) polypoid lesion topographic view; b) intermixed glial (*) and fibrous component(†) HPS X200; c) GFAP immunostaining positivity of glial tissue (X200).

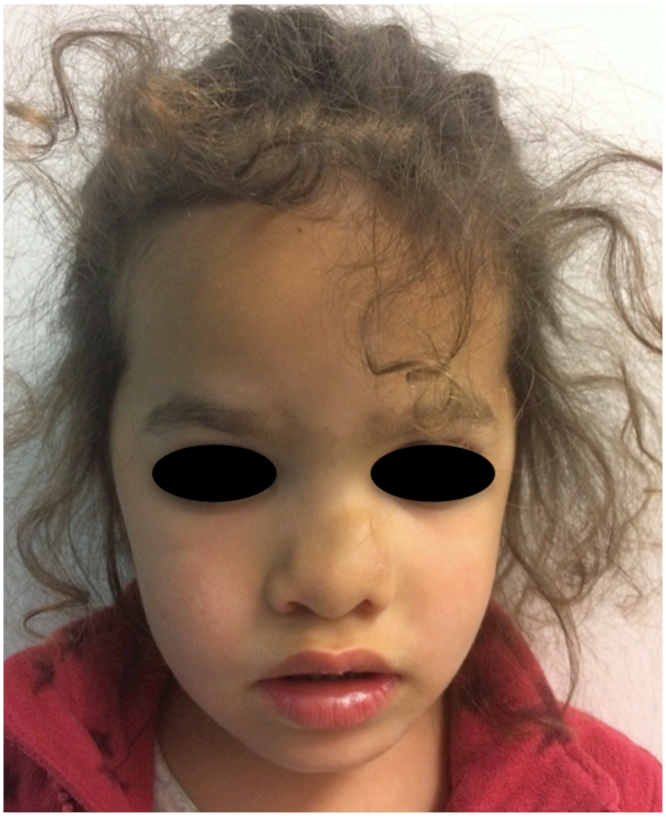

Fig. 7. Full view of the case number 1 one year after surgery.

serie, three patients had no recurrence (Fig. 7) except one patient (case number 4) who developed a recurrence after three months, with a cure achieved after additional surgery.

If there is no evidence of intracranial communication, the external/transfacial approaches are adequate. Neurosurgical consultation may still be necessary if a previously unrecognizable tract is identified intraoperatively to be coursing to the skull base [4]. Intranasal gliomas, if small and lacking intracranial communication, may be excised endoscopically [5].

Histologic examination reveals neuroglial tissue (astrocytes with fibrillary glial processes and connective tissue) without a true capsule. Neurons are usually absent. Rarely, choroid plexus, ependymal-lined clefts and pigmented retinal epithelium are seen especially those of the palate and nasopharynx. The glial tissue can be confirmed by immunoreactivity for glial fibrillary acidic protein (GFAP) or S100 protein [7,10,13].

Conclusion

The rarity of nasal glial heterotopy makes clinical diagnosis very difficult. Intranasal glial heterotopy can be quite difficult because the clinical presentation may not immediately put this diagnosis at the forefront. Diagnosis is easier in a young child, where clinical suspicion of extranasal glial heterotopia is high and histological examination with special stains or immunohistochemistry would ensure correct diagnosis.

Declarations of interest

None.

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