Review Article

Nasopharyngeal neuroglial heterotopy - Choristoma: Case report and review of the literature

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Abstract
We describe the case of a nine-month-old patient with a nasopharyngeal choristoma. The case presented includes the retrospective review of the historical, radiological, surgical and histological assessment of this pathology as well as a literature review of this entity. This case was presented in an infant with difficulty feeding, nasal obstruction and failure to thrive, evaluated with flexible nasal endoscopy, CT and MRI. The lesion was then surgically removed without complications. Nasopharyngeal choristoma is a rare congenital non-malignant mass, which may present within a range of symptoms and severity according to its size, growth and location.

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Case

A 9-month-old female patient was referred to our department due to nasal obstruction, oral breathing, snoring, intermittent feeding difficulty and failure to thrive since the second day of life. She was delivered vaginally at 37-weeks of gestation, the mother had no prenatal control. She had no other medical problems or past medical history.

During the first month of life a mass at the oropharynx became apparent. During physical examination, a white
mass of approximately 3 cm × 2 cm occupied the oropharynx (Fig. 1), it was also evident during flexible nasal endoscopy an obstruction of 100% of the nasopharynx. The patient was assessed with magnetic resonance imaging (MRI) under general anesthesia. MRI confirmed a well-circumscribed lesion in the oropharynx and nasopharynx that appears iso-intense on T1-weighted imaging, and hyperintense T2-weighted imaging. It has a superior cystic component and an inferior solid nodule in close relation to the oropharynx (Fig. 2). No bony defect was evident in the study. Computed tomography (CT) was obtained for surgical planning.

She was scheduled for surgical resection, during which it was found that the stalk of the tumor was adhered to the anterior and posterior pillars of the right tonsil. Excision was accomplished by monopolar cautery without complications (Fig. 3).

Microscopic histopathological findings showed a white polypoid mass of 3.0 cm × 2.2 cm × 2.5 cm, identifying adipose tissue with degenerative changes, connective tissue and cerebral tissue of mature aspect, as well as fragments of choroid plexus structures. In the periphery, stratified squamous epithelium was found (Fig. 4). These findings confirm the diagnosis of nasopharyngeal tumor with neuroglial heterotopy (choristoma).

The patient had an uneventful postoperative course and was doing well at a 2-month follow-up visit (Fig. 5). A magnetic resonance was performed 3 months after surgery, without evidence of recurrence or residual tumor (Fig. 6).

Discussion

A choristoma is a mass of mature cells that arise at an aberrant location. Most cases are presented at birth or early childhood. It is a very rare entity, but the most common location in the oral and maxillofacial region is the nasal area, followed by the palate, tongue, cheek, scalp and orbit. All type of choristomas have been described, more commonly in females, including heterotopic central nervous system tissue choristomas. However, some cases have been described in males. There are different types of choristomas, including glial, gastric, hairy polyps, among others depending on the type of mature cells that make up the tumor.

Choristomas are masses with growth rates similar to that of the surrounding tissue with no tumor like behavior, therefore they should be considered a malformation and not a neoplasm.

Glial choristomas may be solid or partially cystic. These cystic lesions encapsulate fluid similar to cerebrospinal fluid that might result from the production of functioning choroid plexus. There are three theories of the pathogenesis of glial choristomas. One is the protrusion of the neural glial tissue from the developing cerebral tissue,
which becomes isolated from the brain. This theory is supported by the observation of some choristomas that may have a stalk connecting with de intracranial contents. Other theory is the displacement of neuroectodermal cells during an early stage of embryogenesis, which may subsequently differentiate into a variety of neuroglial tissue. The third theory is that the lesion is a neoplasm, but this one is not supported by most authors. They must be differentiated from teratomas, dermoids and hamartomas. Teratomas differ in that they comprise all 3 germ cell layers, and dermoids contain ectodermal and mesodermal elements only. A hamartoma is a mass of non-aberrant mature cells, normally found at the affected site, typically with a single tissue type and disorganization. Other entity from which the neuroglial choristoma must be differentiated is the nasal glioma. A nasal glioma is an extracranial/extracervical congenital tumor made of glial and fibrous tissue and do not contain choroid plexus structures. In our case the mass had mature elements of brain tissue and choroid plexus structures ectopic to their location with some elements typical for their site. No cartilage, bone, dermal or other structures where present suggestive of teratomas or other type of tumors. Presenting symptoms may vary significantly and may include mild to severe airway obstruction since birth, feeding difficulty, cyanosis, choking spells, and failure to thrive among others. Symptoms are influenced by the location of the mass and its size relative to the size of the pharyngeal cavity. For instance, in patients with cleft palate there has been reported a lower occurrence rate of respiratory distress. Several imaging modalities are employed, but the most commonly used are CT and MRI. CT provides a rapid assessment in a critical patient, but MRI provides more details for defining the origin of the lesion and soft tissue involvement. On CT, the lesion is presented as a soft tissue density isodense to brain and with a possible bony defect. If this defect is present, an MRI is helpful to determine an intracranial connection. Despite the usual relation to a bony skull base bony defect, choristomas by definition do not communicate with the subarachnoid space. On MRI the lesion shows isointense signal in T1WI and slight hyperintense signal on T2WI, as the normal brain. Our patient showed the characteristic images of the choristoma on MRI, without bony defect on the skull base, and a cystic component on the most superior aspect. The purpose of the surgery is achieving complete resection and restore normal function (airway, feeding). This can almost always be obtained by different methods.

Fig. 4 Adipose tissue with degenerative changes, connective tissue and cerebral tissue of mature aspect, as well as fragments of choroid plexus structures. In the periphery stratified squamous epithelium was found.

Fig. 5 Oral view of the patient after a two-month follow-up with no recurrence.

Fig. 6 Postoperative MRI control, Sagittal (A) and axial (B) view show no evidence of residual or recurrent tumor.
without major complications. In our case we used monopolar cautery to achieve total extirpation of the lesion. Recurrence has not been documented in any report, even at 5-year follow-up.²,⁶,¹² Uemura had an interesting association between nasopharyngeal glial choristomas and cleft palate. They presented one case and reviewed 17 patients with choristomas, 6 of these cases were accompanied by cleft palate.¹³ Our patient did not have any abnormality in the palate, and remarkably she did not present respiratory distress at birth despite the size of the tumor.

A total of 22 cases of neuroglial choristomas in the oropharynx have been described in the literature so far (Table 1), plus our own case.⁵,⁶,¹¹,¹³,¹⁴

### Conclusion

Choristoma is an aberrant tissue that is not frequently presented in the oropharynx. It is a differential diagnosis of nasopharyngeal obstruction in neonates and infants. If direct examination, upper airway endoscopy and diagnostic imaging suggest this pathology, the treatment must be directed towards total resection to restore airway and normal feeding pathways, which grants the patient an excellent prognosis.

### Declaration of Competing Interest

There is no conflict of interest or financial disclosure to be made.

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Table 1  Neuroglial choristomas in the oropharynx described in the literature.

| Author          | Year | Number of Patients | Gender | Histopathology                                      | Images   | Symptoms                      | Follow-up |
|-----------------|------|--------------------|--------|----------------------------------------------------|----------|-------------------------------|-----------|
| Zhan et al.     | 2014 | 1                  | Female | Astrocytes, oligodendrocytes/ microglialcytes, choroid plexus | CT       | Cleft uvula                   | 2 years  |
| Husein et al.   | 2008 | 1                  | Male   | Choroid plexus, glial tissue, GFAP                  | MRI/CT   | Respiratory Distress          | —         |
| Uemura et al.   | 1999 | 18                 | Female | Astrocytes, GFAP                                    | MRI      | Cyanotic episodes, Cleft palate | 1 year 18 months |
| Al-Amar et al.  | 2006 | 1                  | Female | Choroid plexus, meningeal membrane-like connective tissue. | CT/MRI   | Respiratory distress          | —         |
| Giannas et al.  | 2005 | 1                  | —      | Brain tissue                                         | MRI      | Cleft palate                  | —         |

GFAP: Glial fibrillary acid protein.

²¹ case plus 17 review cases from 1946 to 1975.

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