

Case Report

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Glial Heterotopia Nasal Septum in an Adult: A Case Report

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Abstract

We report a case of Heterotopia of nasal neuroglia from the right nasal septum in a middle-aged man. This Heterotopia is a very rare Heterotopia in the nasal cavity. It presents as a mass of extracranial brain tissue that is not connected to the brain. It often occurs in neonates and children, and is extremely rare in adults.

Case description

A 35-year-old male patient was admitted to the Department of otorhinolaryngology of our hospital on August 1, 2022 due to "bilateral nasal congestion with headache for more than 6 years". Patients with no obvious incentives in bilateral nasal congestion six years ago, I have a side of the heavier, lay down at night and a change in the weather congestion is aggravating, usually have a sneeze, the symptom such as flow cleaning stuff, no flow pus, hyposmia, no facial pressure and swelling of the nasal cavity, outpatient doctor after checking with the nasal septum deviation, bubble on the right side one" She was admitted to the hospital, in good health, with no history of meningitis and no special family history. Hospital physical examination: nasal appearance is good, see: under nasal endoscope (Figure 1) double nasal mucosa is ruddy, nasal septum front-end clearly to the left deviation, block the left nostril, before the left nasal cavity structure is not seen, on the right side of the nasal visible light pink neoplasm growth, qualitative soft, smooth surface without damage, middle turbinate partial properties, and the mass roots part on the septum.

CT of paranasal sinus in our hospital on June 29, 2022 (Figure 2) showed: 1. Soft tissue shadow in ethmoid sinus cavity, obvious left deviation of nasal septum, inflammation 2. Chronic inflammation of the right maxillary sinus and ethmoid sinus. After the relevant examinations were completed before surgery, the patient underwent the operation of "septum correction, right septum mass extraction + right maxillary sinus and ethmoid sinus opening" under general anesthesia on August 3, 2022. During the operation, the tumor was found to be closely connected with the right lateral mucosa of the nasal septum, without adhesion to the right middle and inferior turbinates, and the base of the tumor was derived from the right lateral mucosa of the nasal septum. The basal area was about 1.5 cm x 1.0 cm in size, and the tissue was slightly tough and not easy to bleed. A small amount of clear fluid was found to flow out of some of the tumor tissues after biting lead, and no obvious bleeding was observed. After the tumor specimen was collected for examination, the nasal septum mass was completely removed with the power cutting system. It was found that the inner surface of the tumor had cystic membrane, and the boundary between the tumor and the nasal septum mucosa was

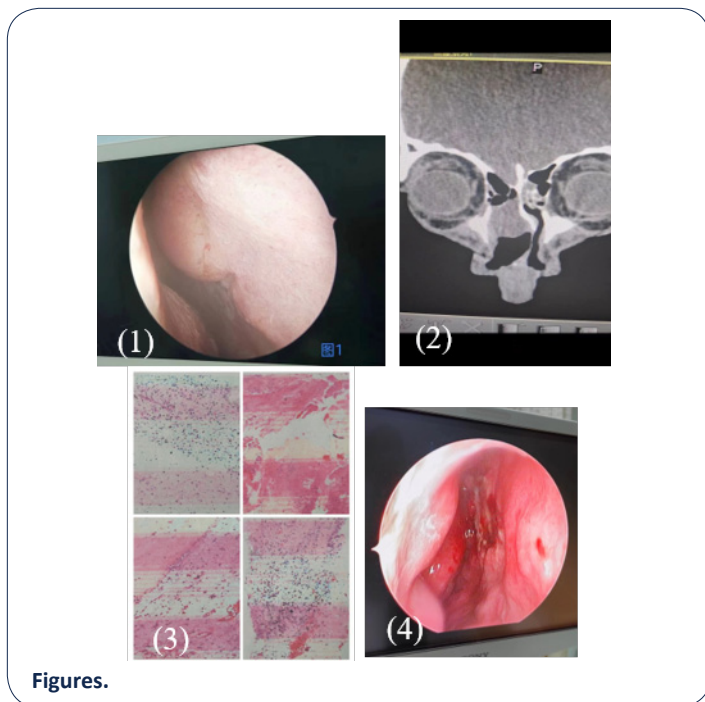
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clear. The nasal cavity was filled with absorbable naxi sponge after operation. Postoperative pathology showed ectopic brain tissue (glial ectopic) and inflammatory polyps (Figure 3). Symptomatic supportive treatment such as anti-inflammation and hemostatic treatment was given after the operation. The patient improved and was discharged from hospital one week after the operation. Reexamination one week after the operation showed satisfactory correction of the nasal septum, a small amount of dry scab attached to the right nasal septum operation area, and nasal cavity was cleaned, and the nasal septum mucosa was intact, and the tumor did not relapse (Figure 4). The patient felt nasal congestion disappeared, still slightly felt forehead pain, a small amount of clear liquid flowed out when the right nasal cavity bowed its head, after the operation, the headache disappeared, nasal cavity still flowed clear nose, and other complaints of special discomfort.



Figures.

Discussion

Heterotopia of nasal neuroglia-NGH, also known as nasal glioma, is a rare congenital midline benign tumor. A disease in which nerve tissue grows outside the central nervous system and ectopic glial tissue is deposited extracranial during embryonic development due to abnormal closure of the nasal and frontal bones [1]. This disease was first reported by Reid in 1852, and the concept of glial ectopia was elaborated by Schmidt in 1900. Since it occurs most often in the nose, this disease is called nasal NGH [2]. Nasal glial ectopia is often diagnosed in neonates but rarely found in adults. There is no family history of the disease, the incidence is very low, the male to female ratio is about 3:2, and it is usually not accompanied by other congenital malformations [3].

NGH is not a true tumor, but a congenital malformation. Its etiology and pathogenesis are still unclear. At present, the accepted pathogenesis is "meningoencephalocele" theory [4]. At 8 weeks of embryo, there is an anterior nasal space between the nasal and frontal bones and their deep square nasal cartilage sac. Through this, the dura meets the skin of the outer nose, forming a dural diverticulum. With the development of the frontal osteonasal process, the dural diverticulum gradually separated from the out-

er nasal skin and retracted into the skull, the blind foramen was filled with fibrous tissue, and the dura was completely enclosed in the skull. At the same time, the nasal cartilage sac gradually developed into the nasal septum cartilage, the vertical plate of ethmoid bone and the comb, until the nasal space disappeared. During the retraction of the dural diverticulum into the skull, congenital nasal NGH is formed its part of the neural tissue remains in its retraction path.

Glial heterotopia (NGH) can occur in several sites, including the scalp, orbit, nasal cavity, middle ear and mastoid process, soft palate, tongue base and neck, and other sites. The incidence of ectopic sites ranges from 1 in 20,000 to 1 in 40,000. However, it mainly occurs around the nasal root, leading to ectopic nasal glial cells [2]. NGH can be divided into three types according to the site of occurrence, namely, intranasal type (about 30%), nasal appearance type (about 60%) and mixed type (about 10%) [3-5]. Extrasal nasal glial Ectopia IS MOSTLY FOUND AT BIRTH, AND as the child grows, the tumor gradually increases in size, resulting in a widening of the nasal root and a widening of the pupil spacing. This results in facial deformities and frequent tears, but when crying, the tumor does not grow or harden; Intranasal tumors are usually located near or connected to the nasal septum by connective tissue. The clinical manifestations were dyspnea, nasal congestion, runny nose, mouth opening breathing, snoring, choking during feeding, and feeding difficulties. The diagnosis of intranasal type is generally later than that of extranasal type [2]. This case is an adult patient, who had no related symptoms in childhood after questioning the history. This time, he sought medical treatment because of nasal obstruction and headache, which was caused by nasal obstruction considering ectopic nasal stenosis caused by nasal neuroglia and nasal septum deviation blocking nostrils.

The symptoms and laboratory findings of nasal NGH in adults are nonspecific. The diagnosis mainly depends on preoperative computed tomography (CT) and MRI imaging. CT helps to observe the bone defect of the anterior skull base, while MRI provides supplementary information about the fluid or soft tissue characteristics of the mass. MRI also provides a three-dimensional view and can determine intracranial invasion [9]; Some authors proposed MRI as the first choice for ectopic glioma after summarizing the cases in the past 40 years. If bone defect was considered, CT control was performed [6].

Macroscopic, intranasal HNG is nodular or polypoid, without capsule, smooth surface, medium hardness, gray powder or grayish brown, cut surface grayish white grayish red, cystic cavity may form, cystic containing cerebrospinal fluid like clear fluid; The outer nasal type has skin on the surface, and the inner nasal type is covered with nasal mucosa [2], which does not radiate or expand with the Valsalva maneuver or crying. The ipsilateral internal jugular vein should be compressed, and the Furstenberg test should be negative to observe whether the mass is dilated or pulsatile [6,7].

Histologically, nasal NGH consists of an unencapsulated collection of astrocytes, eosinophilic glial cells in the connective tissue matrix, and the internal neural tissue is nonfunctional, as no surgical excision resulting in any neurological deficits has been reported. The possible intracranial connections are usually fibrous and do not disrupt the dura [8,9]. According to the study of 10 cases of nasal NGH in foreign countries, it was found that all cases were

composed of neurons and glia in different proportions. There was no obvious ependyma, choroid plexus or light membrane, and the complex epithelium (skin, respiratory or metaplastic squamous epithelium) was intact, although sometimes weakened and atrophic. The glial component is often difficult to identify, especially when there are associated inflammatory complexes, or fibrosis is evident. All cases had chronic inflammatory cells, including lymphocytes and macrophages, and reactive gliosis. In adult patients, fibrosis or sclerosis is significantly more severe and almost completely masks the background glial tissue [8], making it difficult for pathologists to diagnosis nasal NGH.

A strict and comprehensive differential diagnosis of nasal mass must be performed before treatment. The external nasal lesions should be differentiated from common nasal root masses such as hemangioma, dermoid cyst, meningoencephalocele, etc. [8]: hemangioma is soft and multiple. some skin temperature increases, and the pressure can be reduced. Dermoid cysts showed cystic structures by CT and MRI examination. In addition to the imaging features of meningoencephalocele, the mass is usually translucent and can beat with pulse or breathing. The mass can increase with crying force, and the tension can increase. Furstember sign is positive. Intranasal NGH should be distinguished from nasal polyps and meningocele. Intranasal NGH is easy to be misdiagnosed as nasal polyps, the latter is soft, often accompanied by asthma, nasal itching, nasal congestion, runny nose and other symptoms of sinusitis, patients often have a history of allergic rhinitis, the former contrary; Meningocele is the herniation of brain and/or meningeal tissue through a bone defect in the skull. The herniated tissue remains connected to the intracranial nervous system and subarachnoid space, and the meninges or brain tissue usually enters the upper nasal passage through the lamina cribrae. Or they may enter the sinuses and nasopharynx through bone defects in the sphenoid or ethmoid sinuses. In addition, pathological examination of nasal meningoencephalocele usually indicates that the lesion is composed of brain and meningeal tissue, with neuronal cells and ependyma, elements rarely observed in nasal glial ectopia; Although meningocele and nasal glial ectopia have similar clinical manifestations and pathologies, they require different treatment approaches. Therefore, meningocele must be ruled out by clinical and radiographic examination to prevent CSF leakage and meningitis before any form of surgical intervention is performed in patients with suspected congenital nasal glia heterotopia. In such cases, CT and MRI of the nasal cavity are usually performed to determine whether the nasal lesion shows intracranial communication. Although the distinction between nasal NGH and encephalocele depends on the presence of a connection between the mass and the intracranial tissue. However, studies have shown that even with CT and MRI, this connection may be very small or insignificant. In addition, given the developmental abnormalities, bone defects may also be associated with nasal NGH but still show no connection to the brain parenchyma, distinguished by the presence or absence of meningitis and/or CSF rhinorrhea before and after surgery.

For the patients, patients with adult due to congestion in hospital, preoperative examination from the septum pure sense, vesicular sense, nasal polyps, etc to consider is one-sided, most doctors in adult patients, the consultation will not NGH and meningeal encephalocele as the primary differential diagnosis of nasal septum neoplasm, then if blind lines of preoperative biopsy or

surgical removal of the endoscopic surgery. This may lead to serious consequences such as meningitis, brain abscess, or cerebrospinal fluid rhinorrhea [9], so it also reminds us physicians that when nasal mass is encountered, comprehensive preoperative evaluation must be performed, and preoperative biopsy or surgery should not be performed blindly.

Conclusion

Nasal glial heterotopia is a rare benign congenital disease. In our case, ectopic presentation of glial tissue over the nasal septum was extremely rare. Evaluation should include preoperative imaging with thin-cut coronal and axial CT scans and multiplanar MRI scans to rule out any intracranial connections before invasive surgery. In the case of nasal NGH, endoscopic resection is required. Postoperative recurrence and malignant potential are rare. These patients may develop cerebrospinal fluid rhinorrhea and infection after surgery, so close follow-up is important.

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