© Borgis New Med 2022; 26(4): 80-84 DOI: https://doi.org/10.25121/NewMed.2022.26.4.80

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Nasal glial heterotopia in a 5-month-old girl – a case report

Nosowa heterotopia glejowa u 5-miesięcznej dziewczynki – opis przypadku

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Keywords

nasal glioma, nasal glial heterotopia

Summary

Nasal glial heterotopia is a rare congenital mass that is more commonly noted in infants. We report a case of nasal glial heterotopia in a 5-month-old girl initially presenting with unilateral nasal obstruction. Diagnostic imaging showed a tissue mass in the nasal cavity. The mass was resected via endoscopic surgery. We present the approach to radiological examination and clinical course of the case. In cases of infant nasal glial heterotopia recurrence of the mass is possible if it is not completely excised, so it is essential to be careful during the procedure and perform routine follow-ups. Differential diagnosis includes mainly meningocele and encephalocele, polyps and nasal dermoid cyst. Although nasal glial heterotopia is a rare condition, it should be taken into consideration in diagnosis of infants with nasal obstruction.

Słowa kluczowe

glioma, nosowa heterotopia glejowa

Streszczenie

Nosowa heterotopia glejowa należy do rzadkich wad wrodzonych, spotykanych częściej u małych dzieci. Przedstawiamy przypadek nosowej heterotopii glejowej u 5-miesięcznej dziewczynki, pierwotnie konsultowanej z powodu upośledzonej drożności nosa po jednej stronie. Diagnostyka obrazowa wykazała obecność masy tkankowej w stropie lewej jamy nosa. Zmiana została usunięta w całości z dostępu endoskopowego. Przedstawiamy przebieg diagnostyki i leczenia. W przypadkach nosowej heterotopii glejowej całkowite usunięcie zmiany jest warunkiem braku nawrotów, stąd niezbędna jest zarówno staranna diagnostyka, precyzyjne usunięcie, jak i ścisła kontrola pozabiegowa. Diagnostyka różnicowa obejmuje przede wszystkim takie zmiany, jak: przepuklina oponowa lub oponowo-mózgowa, polipy oraz torbiel dermoidalna. Pomimo że nosowa heterotopia glejowa jest zmianą rzadką, powinna być brana pod uwagę w procesie diagnostycznym u dzieci z zaburzeniami drożności nosa.

Introduction

Nasal glial heterotopia (NGH), also often referred to as nasal glioma, is a congenital benign mass that is made up primarily of astrocytes including the gemistocytic type and a fibrovascular connective tissue stroma intermingled with neuroglial fibers (1, 2). All glial heterotopias have variable histological characteristics but share similar traits (2). This mass can form either intranasally, extranasally or both. NGH is predominantly found in newborns and infants, with rare exceptions in adults (3).

Intranasal nasal glial heterotopia should not be confused with meningoencephalocele, nasal tumors and nasal polyps. The latter is less likely to be found in small children, rather they are generally found in teenagers and adults. While the final diagnosis is made based on histopathological findings, differential diagnosis and the course of treatment are guided primarily by imaging studies.

The objective of this paper is to raise awareness among healthcare providers, especially pediatricians and otolaryngologists, about NGH in order to treat patients with this condition efficiently.

Case report

A 5-month-old female patient was admitted to the Pediatric Otolaryngology Department in a tertiary care center with unilateral nasal obstruction caused by a mass in her left nasal cavity.

The nasal lesion was first observed by her parents during the first 2 weeks of life during nose cleaning. The parents observed no fatigue during feeding and no epistaxis. Two months prior to referral to our department a contrast-enhanced computed tomography (CT) scan was performed, which showed the mass in the left nasal cavity originating most likely from the roof of the nose and a possible defect of the floor of the anterior cranial fossa (fig. 1). No contrast enhancement of the lesion was described. It was not possible to rule out the communication between the mass and the cranial cavity. A magnetic resonance imaging (MRI) study was subsequently performed after the CT scan which showed the mass to be directly adjacent to the dura of the left frontal lobe with slight modeling, without evident dural involvement (fig. 2). The parents reported that the mass slowly grew over time, producing a nearly complete obstruction of the left nasal cavity during the diagnostic process. The patient was referred to our department for further management.

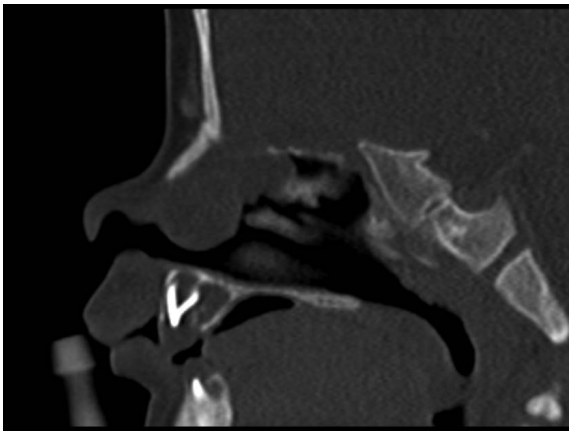

Fig. 1. Parasagittal view of the left nasal cavity and floor of the anterior cranial fossa showing a possible communication between the mass and the anterior cranial fossa

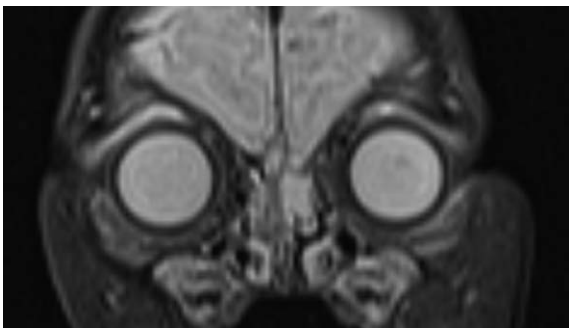

Fig. 2. Frontal view in a T2-weighted MRI scan showing no connection between the mass and the brain tissue

Examination

On admission, an endoscopy was performed which confirmed the location, size and possible origin of the mass (fig. 3). The mass was soft, and covered with pink mucosa. Furstenberg's test was negative.

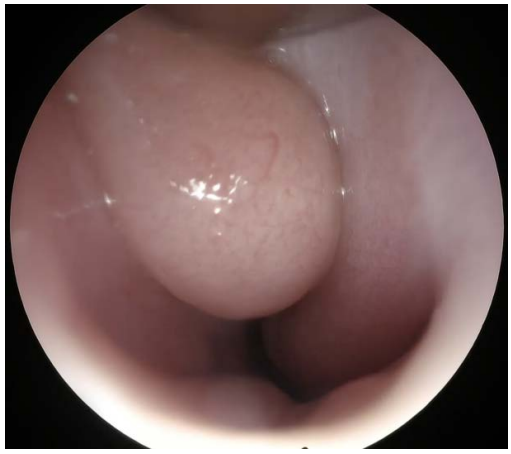

Fig. 3. Endoscopic examination of the left nasal cavity on admission – a mucosa-covered mass visible

Procedure

The patient received intravenous ceftriaxone prior to surgery. A neurosurgical consultation was obtained with a plan for a combined otolaryngological and neurosurgical management to address a possible CSF leak. An intranasal endoscopic procedure under general anesthesia was performed in which the mass was completely excised from the nasal cavity (fig. 4a). A defect of the skull base was observed with no intraoperative cerebrospinal fluid (CSF) leak present (fig. 4b). A nasal pack (PVA sponge) was put in place for hemostasis.

Post-operative course. The excised mass was sent for histopathological examination which confirmed the initial diagnosis of nasal glial heterotopia. The nasal pack was removed one day after the surgery, and no bleeding or CSF

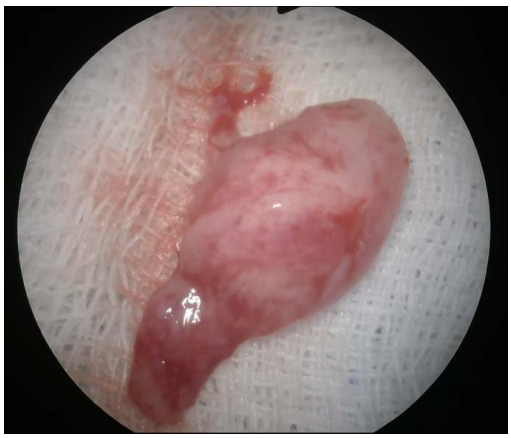

Fig. 4a. Completely excised mass

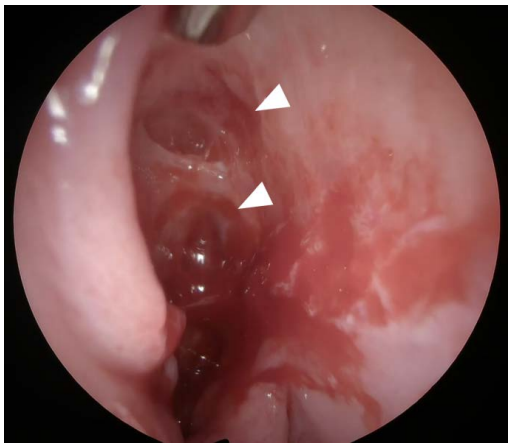

Fig. 4b. A double defect (shown by arrows) of the skull base is visible after the complete removal of the mass

leak was observed. The patient was discharged home two days later without complications.

On the first follow-up visit one week after discharge, no complications were found. During the second follow-up visit 6 weeks after discharge home no bleeding and no CSF leak were noted, the nasal patency was good and a completely healed surgical site was observed in endoscopy (fig. 5).

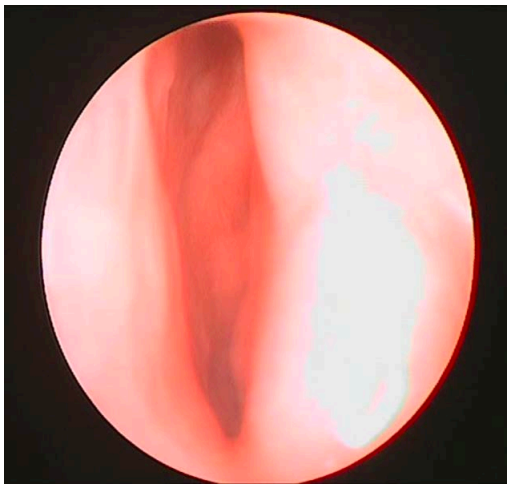

Fig. 5. Endoscopic view during follow-up showing a completely healed roof of the left nasal cavity with healthy mucosa

Discussion

Nasal glial heterotopia, NGH (also known as Nasal Glioma, NG), was first discovered by Reid in 1852 (4), described by Berger as "encéphalocèle" in 1890, and the first complete elucidation was done by Schmidt in 1900 (5). Throughout history, NGHs have been characterized under different names: fibrogliomas, encephalochoristomas, encephalomas, ganglioma, astrocytoma, ganglioblastoma, ganglioneuroschwannospongioblastoma, encephalo-choristoma-naso-frontalis, encephaloma, and choristoma (6). The most commonly found division in literature shows that 60% of NGH cases are extranasal, 30% intranasal and the rest 10% present as mixed with both an internal and external part (7-9). However, another later study conducted between the years 1950 and 1963 including a bigger study group showed 39% of cases found extranasally, 33% intranasally and 27% mixed (6, 10). Around 5% of congenital nasal lesions are due to NGH. While in most cases lesions present within the first year of life, studies have shown that the average peak of occurrences is between ages 5-10 years which could also probably be due to undiagnosed lesions in the early years of life (11). NGH is primarily a pediatric condition, with cases rarely described in adults (4), and a male predominance of 2:1 (4, 12).

Extranasal glial heterotopias are usually firm on palpation and unaffiliated with the enveloping skin, which could appear telangiectatic. Intranasal glial heterotopias appear pinkish or reddish and polypoid on examination, therefore can be mistaken for nasal polyps, however, the latter is very infrequent in patients under 5 years of age (10). Another intranasal pathology that requires differentiation from NGH is encephalocele. Recent molecular histopathological data suggest that these two lesion types may represent manifestations of the same histopathological entity, however clinical distinction is used in practice (13). Furstenberg test could be used to distinguish nasal glial heterotopia from encephalocele: compression of the ipsilateral jugular vein produces an increase in volume or pulsation of a true encephalocele, while it does not have any effect in most cases of nasal glial heterotopias (14).

In our case, the NGH was completely intranasal and showed on examination as a mucosa-covered soft but firm mass. It was observed in early infancy which is typical for congenital lesions, with a negative Furstenberg test strengthening the initial suspicion of NGH over an encephalocele.

Both computed tomography (CT) scans and magnetic resonance imaging (MRI) plays a role in the diagnostic process of nasal masses. MRI offers a superior over CT tissue differentiation possibility, therefore can aid strongly in the diagnostic process. CT scans show the bony structures of the skull base, especially the cribriform plate, which helps to exclude any possible connections between the nasal cavity and anterior cranial fossa. In infants, while a shorter-lasting CT scan without contrast enhancement is sometimes achievable during natural sleep, an MRI scan and a contrast-enhanced CT need to be taken under sedation. The radiation dose produced by CT has to be taken into consideration, especially in small children. The choice of which study to perform is not an easy one and depends both on the clinical evaluation and the availability of diagnostic options. Ultrasound may play a role in extranasal NGH cases diagnosed prenatally (15) but is of little help in small intranasal lesions.

In the presented case the initial contrast-enhanced CT scan showed the size and localization of the mass and a possible defect of the skull base, with no conclusions as

to a possible link with brain tissue, so could not be used to rule out an encephalocele. The radiation dose produced by this examination was roughly 2.5 mSv, which is high for a 3-month-old. Although CT is the examination of choice for the assessment of bony structures of the viscerocranium, the anterior skull base is hard to visualize in a three-month-old due to incomplete calcification of the bone. Also, the defect of the roof of the nasal cavity found intraoperatively did not relate directly to the images from the radiological studies.

A careful choice of radiological imaging has to be made especially in infants regarding radiation exposure and the need for sedation for imaging. MRI is considered the first choice of imaging in cases of NGH (4). It is the authors' opinion that in pediatric patients, a CT scan, if needed at all for surgical planning, should be performed in the shortest time possible prior to surgery.

It is contraindicated to perform a biopsy in a patient with a suspected nasal glial heterotopia, as it poses a high risk of meningitis and CSF rhinorrhea, especially if there is a connection of the lesion within the cranial cavity (16).

Intranasal glial heterotopias usually cause nasal obstruction as their primary symptom and can potentially lead to respiratory distress, especially in neonates who rely on nasal breathing. The first line of treatment is surgical resection of the mass with an endoscopic approach preferred for intranasal lesions (17), as performed in described case.

Conclusions

Nasal glial heterotopia is a rare congenital benign growth, which is most commonly seen in small children. Differential diagnosis including mainly other congenital lesions of the nose relies strongly on radiological investigation. An appropriate choice of imaging studies is crucial for preparation for surgery, which is the only treatment. Children with NGH will usually be examined initially by pediatricians before being referred to otolaryngologists, therefore awareness about NGH in both specialists is beneficial. Additionally, challenges in radiological imaging in infants should be taken into account so that radiologists, pediatricians and otolaryngologists may work together to establish a proper diagnosis and treatment.

Consent information

Informed consent was obtained from the patient's parents for the use of medical data for publication purposes.

Conflict of interest
Konflikt interesów

None
Brak konfliktu interesów

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submitted/nadesłano:
2.11.2022
accepted/zaakceptowano do druku:
23.11.2022

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© Borgis New Med 2022; 26(4): 84-88 DOI: https://doi.org/10.25121/NewMed.2022.26.4.84

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Increase in middle ear infections of *Streptococcus pyogenes* etiology among patients of the Department of Pediatric Otolaryngology of the Medical University of Warsaw in correlation with world reports

Wzrost liczby zakażeń ucha środkowego o etiologii *Streptococcus pyogenes* wśród pacjentów Kliniki Otolaryngologii Dziecięcej Warszawskiego Uniwersytetu Medycznego w korelacji ze światowymi doniesieniami

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Keywords

Streptococcus pyogenes (GAS), invasive *Streptococcus pyogenes* (iGAS), acute otitis media (AOM), acute mastoiditis

Summary

Introduction. Beta-hemolytic streptococcus is one of the most common causes of out-of-hospital infections. It can cause local or general infections of varying severity, most commonly in the throat, paranasal sinuses, ear and skin. Some infections are invasive in nature. In the last months of 2022, an alarming increase in infections caused by *Streptococcus pyogenes*, including invasive infections, has been observed in many countries in Europe and around the world compared to previous years.

Aim. The purpose of this study was to compare the number of hospitalized patients with acute otitis media and acute mastoiditis of *S. pyogenes* etiology in the first six months of 2023 with hospitalizations for the same reason in the previous 5 years, and to assess the correlation with the observed worldwide trend of an increase in the number of observed beta-hemolytic streptococcus type A infections.

Material and methods. A retrospective analysis of the data of patients diagnosed with acute otitis media and acute mastoiditis of *S. pyogenes* etiology hospitalized in the Department of Pediatric Otolaryngology in the first six months of 2023 was carried out, and their number was compared with patients hospitalized for the same reason in the previous 5 years.

Results. Between January 2023 and the end of June 2023, 15 patients with acute otitis media were hospitalized in the Department of Pediatric Otolaryngology, in which *S. pyogenes* was isolated by bacteriological examination. Purulent leakage from the ear was observed in 10 patients. Isolated otitis media was diagnosed in 4 patients. In 10 children, AOM was complicated by acute mastoiditis, which was confirmed by CT scan. The most severe complication was sepsis of GAS etiology. Intravenous antibiotic therapy and surgical treatment were implemented in all patients, achieving a cure. From 2018 to 2022, the number of patients hospitalized for the same reason was significantly lower each year.

Conclusions. The number of patients hospitalized in the Department of Otolaryngology in the last two months of 2022 and in the first six months of 2023 due to AOM, as well as AOM complicated by mastoiditis, in which the etiologic agent turned out to be *S. pyo-*