

Nasal Glial Heterotopia in a 1 Year Old Child - A Case Report: Histopathology is the Ultimate Gold Standard for Diagnosis

Ashwini Gundawar, Sneha Joshi

Department of Pathology, MIMER Medical College, Talegaon Dabhade, Pune, Maharashtra, India

ABSTRACT

Nasal glial heterotopia (NGH) is a benign congenital malformation wherein abnormally located mature brain (glial) tissue presents as a mass on the forehead or nasal root area. Rarity of this condition makes clinical level diagnosis a challenge. Differential diagnoses for NGH are dermoid cyst, encephalocele, hemangioma, allergic nasal polyp, or chronic otitis media. NGH has no direct communication with intracranial cavity, unlike an encephalocele. However, potential intracranial connection is possible, through cribriform plate or bony deformities. Therefore, pre-operative aspiration and biopsies are contraindicated in childhood swellings in forehead/nasal bridge area. Instead, pre-operative imaging modality investigations are mandatory. It is also important to note the risk for the removal of functional brain tissue and also post-operative meningitis or cerebrospinal fluid rhinorrhea. A 1-year-old female child presented with a mass on nasal bridge. Overlying skin was unremarkable. Swelling did not increase in size on coughing. Diagnosis: Dermoid cyst/encephalocele. Computed tomography (CT) scan investigation: CT scan confirmed the diagnosis of nasal encephalocele > nasal dermoid. The mass was excised. Histopathology (histopathological examination [HPE]): The excised specimen was a single, unencapsulated, ovoid, and soft to firm, yellow-colored tissue bit, measuring 2.5 cm × 2 cm × 1 cm. On cut section, there were no cystic areas/spongy appearance/mucoid bits. Hematoxylin and eosin-stained sections revealed a poorly circumscribed mass, showing a population of cells with ovoid or irregular nuclei and a fibrillary stroma - resembling cerebral and glial tissue. These were arranged in a disorganized fashion and were surrounded by fibrous tissue and few skeletal muscle fibers. All HPE findings point toward the diagnosis of NGH. It is important to consider NGH as a differential, in case of childhood swellings in the forehead and nasal root region. Histopathology remains the gold standard for diagnosis.

Key words: Congenital malformations, histopathology, nasal glial heterotopia

INTRODUCTION

Nasal glial heterotopia (NGH) is a benign congenital malformation wherein abnormally located mature brain

(glial) tissue presents as a mass on the forehead or nasal root area.^[1-6] It may also present as allergic nasal polyp or chronic otitis media.^[2-6] Rarity of this condition makes clinical level diagnosis a challenge unless the clinician is sensitized to this possibility and bears a high index of suspicion.^[2,4] The differential diagnoses for NGH are dermoid cyst and encephalocele.^[2,4-6]

The incidence of the triad of "dermoid cyst, encephalocele, and NGH" - at glabellar/root of the nose location - is quite low, approximately 1:20,000-1:40,000 live births.^[2-4,7] However, swellings in this region are of clinical significance due to the possibility of, at least a potential or retained,

| Access this article online | |
|------------------------------------|-------------------------------------------------------------------------------------------------------------------|
| Website: www.mmj.net.in | Quick Response Code  |
| DOI: 10.15713/ins.mmj.41 | |

Address for correspondence:

Dr. Ashwini Gundawar, Department of Pathology, MIMER Medical College, Talegaon Dabhade, Pune, Maharashtra, India. E-mail: gundawar.ashwini@gmail.com

connection to the intracranial cavity. While an encephalocele, by definition, has a patent connection to the intracranial cavity, a dermoid may or may not have a patent intracranial passage.^[7] NGH has no direct communication with intracranial cavity, unlike an encephalocele. However, a potential intracranial connection is always possible and may be located through cribriform plate or other bony deformities in the location.^[1-7]

Therefore, pre-operative aspiration and biopsies are contraindicated in childhood swellings in forehead/nasal bridge area. Instead, pre-operative imaging modality investigations are mandatory. It is also important to note the risk for the removal of functional brain tissue during surgical excision of the mass and post-operative meningitis or cerebrospinal fluid rhinorrhea.^[1,2,4-6]

CASE REPORT

History and Clinical Examination

A 1-year-old child presented in the otorhinolaryngology ear, nose, and throat (ENT) outpatient department (OPD) for a mass on the bridge of her nose that was present since birth and growing slowly [“Figure 1” for a clinical photograph]. On examination, the child was pale, malnourished, and suffered from upper respiratory tract infection. She also had a history of recurrent upper respiratory tract infections and failure to thrive. On local examination, the mass measured 2.5 cm × 2 cm in size, skin over the swelling appeared unremarkable. The swelling was firm on palpation, not compressible and it did not increase in size on coughing. The last elicited sign suggestive of the absence of a patent communication with the intracranial cavity.^[1-3,5,7] A provisional clinical diagnosis of nasal dermoid/encephalocele was made. The patient was posted for surgery, but the operative procedure was delayed by almost 3 months due to recurrent upper respiratory tract infections and her malnourished status.

Investigations

Imaging studies

Computed tomography (CT) scan – (brain) was done preoperatively for this patient. The report showed the following features:

1. Soft tissue density seen in the subcutaneous tissue over nose anteriorly with no significant fat density or calcifications seen inside it
2. Small bony defect seen in the midline between the nasal bones with a soft tissue bit herniating through it
3. Small oval defect seen in anterior crista galli
4. There was no large intracranial mass seen.

Final diagnosis on CT scan was, nasal encephalocele, > nasal dermoid.

The mass appeared to have no patent intracranial connection on CT scan.

Mass was excised and submitted for histopathological examination (HPE).

Pathological findings

The excised mass received in the department of pathology for HPE. On gross examination, it was a single, ovoid, soft to firm, yellowish piece of soft tissue, measuring 2.5 cm × 2 cm × 1 cm, with a glistening and firm cut surface.

Microscopic examination of hematoxylin and eosin-stained sections revealed a poorly circumscribed mass showing a disorganized population of cells composed of ovoid or irregular nuclei and a fibrillary stroma resembling glial and cerebral tissue. These two elements interdigitated with and were surrounded by fibrous tissue.



Figure 1: Pre-operative photo of the child with the swelling nasal glial heterotopia

Special stains were done to highlight the glial tissue. Masson's trichrome showed islands of red glial fibers located within green collagen tissue bundles. Mallory's phosphotungstic acid hematoxylin stain revealed, deep blue/purple stained islands of glial tissue with pale pink collagen bundles.

The histopathological features were consistent with the diagnosis of NGH^[4,8,9] [Figure 2].

DISCUSSION

NGH along with dermoid cyst and encephalocele is the triad of differentials for glabellar and nasal bridge swellings in pediatric population. All three occur as a result of the failure of the frontal lobe of the brain to retract inside the cranium as the facial bone sutures close in the 3rd and 4th weeks of embryonic life.^[2-5] Of these three most likely diagnoses for a swelling in this region, the most common is the nasal dermoid and the least common is the NGH. Both present as firm, non-compressible, non-transilluminating subcutaneous

swellings, with no cough impulse similar to the mass in our patient.^[1-7]

The need for mandatory pre-operative imaging modality investigations is stressed on by all authors (CT/magnetic resonance imaging) due to the risk of a potential or patent intracranial connection that a swelling in this location may have.^[1-8] The child, in our case, underwent a CT scan to rule out a possible communication, between the mass occupying the glabellar and nasal bridge region, and the intracranial cavity. The CT scan revealed that there appeared to be no obvious patent passage between the mass and the intracranial cavity. However, a nasal bone defect was present and a stalk of soft tissue was seen to be passing through this nasal bone defect and was attached posteriorly to the mass in consideration. The presence of the nasal bone defect and this stalk skewed the CT scan diagnosis in favor of an encephalocele rather than a dermoid, as the possibility of a potential connection with the intracranial cavity could not be ruled out completely.

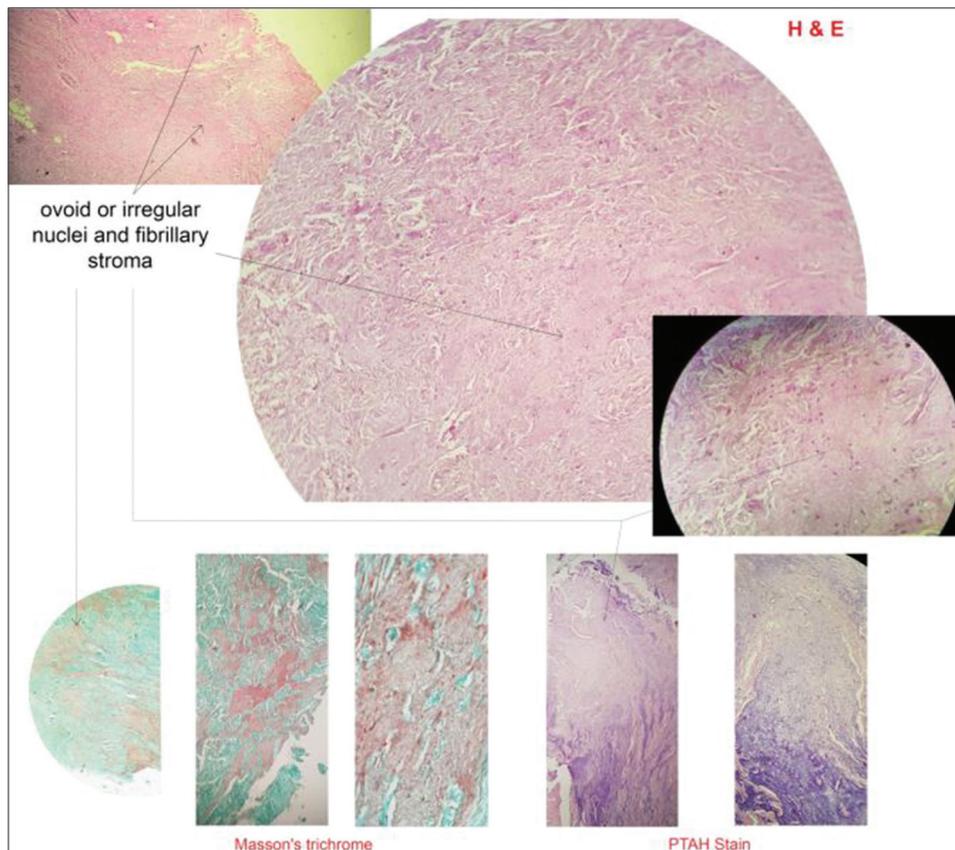


Figure 2: Microscopic pictures of routine (Haematoxylin and Eosin) stained sections and special stains for confirmation: Masson's Trichrome and PTAH

As per literature, this twist is a fairly common occurrence due to the incomplete ossification and anatomical variations in the area in children <1 year old.^[7]

Our patient was a female child of 18 months of age. Literature references mention that the most common age group for presentation, of a swelling in this region, is early infancy including the newborn.^[1-6] The average size for such swellings mentioned in literature was 2.4 cm. This size matches with the size of the mass present in the current case.^[4]

Literature mentions three types of NGH as per the location of the lesion. These are extranasal (60%), intranasal (30%), and combined (10%).^[1,2,4,10] In our case, it was the most common type, the purely extranasal kind, and in the most common location – the midline.

Our patient, therefore, matches – the most common age group for presentation (birth to early infancy), the average mass size (2.4 cm), most common site – nasal bridge and the extranasal location, and most common of the three varieties mentioned in literature. These children have a tendency to present with upper respiratory tract infection such as symptoms, rhinorrhea, obstruction, and otitis media.^[1-6] This child too had episodes with similar complaints during the admission period. In addition to the bony defect in nasal bone, the CT scan also showed a middle ear density which was inferred as incomplete pneumatization/ infection. These kinds of bony deformities and defects in this region are very commonly associated with NGH.^[7] Therefore, this child was a classic book picture like case of NGH.

However, despite fitting into the classic features for the said diagnosis, the clinical diagnosis in this particular girl's case was "dermoid cyst" and the radiological diagnosis was "More in favor of encephalocele than a dermoid." This situation may be explained by the rarity of this condition, "Nasal glial heterotopia." This condition is so rare that the "Department of Otorhinolaryngology – Head-and-Neck Pathology," in an institute of the caliber of "Armed Forces Institute of Pathology, Washington, DC, USA" saw a total of only 10 cases of NGH, diagnosed, and identified as such, over a 30-year span.^[4] In all the cases quoted in the scientific literature, the actual diagnosis of NGH was cinched only on HPE of the excised mass. This also shows succinctly that an HPE remains the gold standard for an accurate diagnosis.

Resection of NGH is usually curative.^[4,6] Our patient is now lost to follow up and so the final post-operative

outcome remains unknown. Intense efforts by the social worker department to trace her were, alas, fruitless. We may, however, assume complete cure without any untoward outcomes, as the immediate post-operative period was uneventful and the child did not return to the ENT OPD with any further complaints.

CONCLUSIONS

1. Although a rare condition, it is important to consider "nasal glial heterotopia" as a differential, for congenital midline mass in the glabellar/bridge of nose region, in the pediatric population, especially in early infancy
2. Histopathology remains the gold standard for diagnosis of this condition as imaging techniques cannot yet differentiate between an encephalocele and NGH.

REFERENCES

1. Hagiwara A, Nagai N, Ogawa Y, Suzuki M. A case of nasal glial heterotopia in an adult. *Case Rep Otolaryngol* 2014;2014:1-4.
2. Krishna LG, Uppoor R, Rao KN, Harish K. Heterotopic central nervous tissue-nasal glioma: A case report. *Indian J Radiol Imaging* 2005;15:511-6.
3. Chintale SG, Jatale SP, Kirdak VR, Shaikh KA. Congenital midline nasal mass: Four cases with review of literature. *Bengal J Otolaryngol Head Neck Surg* 2017;25:160-5.
4. Penner CR, Thompson LD. Nasal glial heterotopia: A clinicopathologic and immunophenotypic analysis of 10 cases with a review of the literature. *Ann Diagn Pathol* 2003;7:354-9.
5. Swain SK, Dash S, Baisakh MR, Samal R. Glial heterotopia over the nasal septum: A case report and review of the literature. *Clin Rhinol* 2015;8:34-7.
6. Kardon DE. Nasal glial heterotopia. *Arch Pathol Lab Med* 2000;124:1849.
7. Rahbar R, Shah P, Mulliken JB, Robson CD, Perez-Atayde AR, Proctor MR, *et al.* The presentation and management of nasal dermoid: A 30 year experience. *Arch Otolaryngol Head Neck Surg* 2003;129:464-71.
8. Thakur S, Parab S. Case of nasal glial heterotopia: A diagnostic dilemma. *MIMER Med J* 2017;1:37-40.
9. Pernick N. Home Nasal Cavity, Paranasal Sinuses, Nasopharynx Glial Heterotopia. Michigan USA, 2004-2018 Glial Heterotopia; 2018. Available from: <http://www.pathologyoutlines.com/topic/nasalglialheterotopia.html>. [Last accessed on 2018 Nov 03].
10. Chandran SK, Hirsch MB. Congenital midline nasal

masses. In: Sataloff RT, editor. Surgical Techniques in Otolaryngology-Head and Neck Surgery: Pediatric Otolaryngologic Surgery. 1st ed. Philadelphia, PA, New Delhi, London, Panama: Jaypee Brothers Medical Publishers; 2014. p. 53. Available from: https://www.books.google.co.in/books?hl=en&lr=&id=1t_fBAAAQBAJ&oi=fnd&pg=PA53&dq=Congenital+midline+nasal+masses&ots=aZ4FqudBdo&sig=UC3bHraFY2eaXBYN1AFuoJ5m-LU#v=onepage&q=Congenital%20

midline%20nasal%20masses&f=false. [Last accessed on 2018 Dec 12].

How to cite: Gundawar A, Joshi S. Nasal glial heterotopia in a 1 year old child – A case report: Histopathology is the ultimate gold standard for diagnosis. MIMER Med J 2019;3(1):15-19.

Source of Support: Nil. **Conflict of Interest:** None declared.

This work is licensed under a Creative Commons Attribution 4.0 International License. The images or other third party material in this article are included in the article's Creative Commons license, unless indicated otherwise in the credit line; if the material is not included under the Creative Commons license, users will need to obtain permission from the license holder to reproduce the material. To view a copy of this license, visit <http://creativecommons.org/licenses/by/4.0/> © Gundawar A, Joshi S. 2019