**Nasal Glioma: A rare congenital midline lesion in paediatric population**

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**Type of Publication:** Case Report

**Conflicts of Interest:** Nil

**Abstract**

Nasal glioma (glial heterotopia) is a rare benign congenital midline lesion. Patient may present with a swelling in nostril or nasal congestion. We are reporting a case of nasal glioma that was diagnosed in department of Radiodiagnosis, Pushpagiri Institute of Medical Sciences and Research Centre in Kerala. We emphasize on the developmental anatomy, imaging and differentials of the same.

**Keywords:** Nasal glioma, congenital midline lesion, nasal dermoid, encephalocele, case report.

**Introduction**

Nasal glioma or nasal glial heterotopia, is a rare congenital midline lesion composed of dysplastic glial cells. They occur sporadically with no familial tendency or gender predilection. A nasal glioma may be connected to the brain by a stalk in upto 15% of cases, but stalk does not contain a fluid filled tract, hence is distinct from a nasal encephalocele. A near definitive diagnosis can be made by clinical examination with the aid of imaging, but gold standard is surgical excision and histopathological confirmation. Only a few cases of nasal glioma have been reported in literature so far, especially from Kerala settings.

We present the case of a 3year old patient with nasal glioma with an emphasis on MR findings. We highlight the use of MR findings in facilitating physicians to make an early diagnosis and to differentiate this condition from other diagnoses, so that it will be helpful in guiding and deciding on subsequent clinical management.

**Case Report**

A 3year old male patient without any surgical history presented to ENT OPD with complaints of nasal congestion since birth which was associated with mouth breathing, snoring and nasal discharge for which he was on nasal spray for past 6 months. Parents noticed a swelling within the right nostril for past 4 months, which was gradually increasing in size. On clinical examination, a pink soft tissue mass was present anterior to right inferior turbinate. Right nasal bone splaying was present with normal vestibule and columella. Roof of the right nasal cavity was not clearly visualised.

MRI brain and PNS was done (GE-Signa HDxt 1.5T) which revealed well defined smoothly marginated lesion in right nasal cavity measuring 1.4 x 0.6 x 0.7 cm. The lesion exhibits T1W hypointense (Fig 1) and T2W hyperintense (Fig 2) homogenous signal. The lesion was seen to arise from the roof of right nasal cavity and extended down to fill almost the entire anterior half of right nasal cavity, causing significant luminal narrowing as well as deviation of nasal septum to the left side. No definite defect in cribriform plate or communication with intracranial CSF were noted. No blooming was appreciated on gradient sequence. On post contrast study, only thin smooth peripheral enhancement was demonstrated without any enhancing solid components within the lesion (Fig 3).

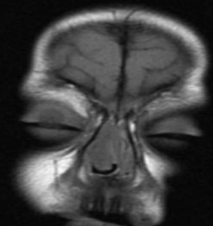


Fig. 1: Coronal T1W. A polypoid T1W hypointense signal arising from the roof of right nasal cavity and extending down to fill the nasal cavity.

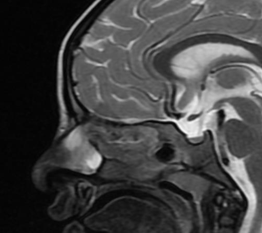


Fig. 2: Sagittal T2W. Hyperintense lesion on T2W. No definite communication with intracranial CSF

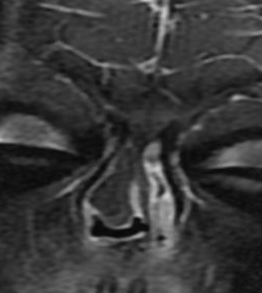


Fig 3: Post contrast Coronal T1WFS image shows thin smooth peripheral enhancement of the lesion.

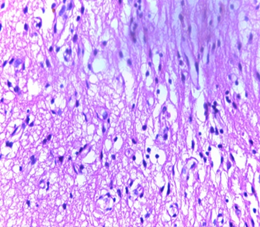


Fig 4: Microscopic section from tumour showglial cells (mature) and fibrovascular connective tissue

The patient underwent endoscopic excision of right nasal mass under general anaesthesia. It was pathologically proven as nasal glioma.

**Discussion**

**Nasal glioma****:** Nasal glioma is one of the rarest anomalies that radiologists encounter in day-to-day practice. In a study conducted by A. James Barkovich, out of 16 cases of congenital nasal mass in children, only one case was pathologically proven as nasal glioma. (1)

Developmental anatomy of primitive frontonasal region

Primitive frontonasal region (anterior neuropore) develops in the third week of fetal life. Small fontanella between the inferior frontal bone and nasal bone is fonticulus frontalis. Anterior skull base opening through a midline opening anterior to the crista galli of the ethmoid bone is the foramen cecum. The apex of dural diverticulation extends through pre nasal space till the subcutaneous plane. Failure of involution at these sites where surface and neural ectoderm approximate each other can lead to anomalous development including: nasal dermal sinus, encephalocele, and nasal glioma. (4) Gliomas occur due to abnormal closure of fonticulus nasofrontalis which later leads to an ectopic rest of glial tissue being left extracranially. Formation of encephaloceles is also similar to this but retains an intracranial connection which is not necessary in case of a glioma. (2)

**Imaging**

Neuro imaging is of utmost importance especially to exclude a possible intracranial extension. Ultrasound is used to assess the content of the lesion – whether it is a solid or cyst. Doppler flow studies of nasal glioma show low velocity arterial flow during the end-diastolic phase. (2)

Classically, radiologist must look for widening of the nasal septum, a bony defect in the cribriform plate, erosion of bones and an increased interorbital distance in plain CT study. (1) However, the differentiation between nasal glioma and encephalocele can be challenging. In these cases, MRI with high-resolution imaging and IV contrast administration is advised to assess for contiguity of heterotopic glial tissue with the cranial cavity because this finding will significantly change the surgical approach and risk. The most valuable imaging plane is sagittal. (3) Nasal glioma has intermediate signal intensity on T1- weighted images, and show mild peripheral contrast enhancement. Lesion exhibits high signal on T2-weighted images as a result of gliosis. The nasal septum is not deformed in most of these lesions but may appear truncated anteriorly with large nasal gliomas. Nasal glioma usually is not connected to the brain. These lesions contain non neoplastic glial tissue, unlike intracranial gliomas. For this reason, nasal brain heterotopia or nasally trapped brain might be a better term rather than nasal glioma. (5)

Histopathologically, the diagnosis is confirmed by observation of glial cells (mature) and ﬁbrovascular connective tissue (Fig 4). Foci of calciﬁcations may also be present. The presence of leptomeninges, ependyma, choroid plexus would support a diagnosis of encephalocele. (7)

**Types of nasal gliomas:**

* Extra nasal (most common): most commonly occur slightly off the midline
* Intranasal (Second most common)
* Combined (rarest). (2)

Our case was of a typical intra nasal type of nasal glioma.

**Differential diagnoses**

The differential considerations of nasal glioma include nasal encephalocele and nasal dermoid. (3)

1. Nasoethmoidal encephalocele

This is characterized by the herniation of brain substance into the nasal cavity through a congenital defect in the skull, just anterior to the crista galli. If the encephalocele is large enough, midline structures can be splayed and can lead to hypertelorism. (2) The best imaging technique for describing an encephalocele's contents is magnetic resonance imaging (MRI). Using MR imaging, the brain tissue in an encephalocele is also more clearly characterised, helping with surgical planning and prognosis. Although MR imaging provides the clearest definition of the intracranial link, high-resolution CT may also be employed to show bone architecture. (8)

1. Nasal dermoid

Nasal dermoids occur due to lack of regression of a diverticulum of dura that extends through the foramen ceacum between the developing nasal cartilage and nasal bone. There may be an associated sinus or opening on the skin surface and can extend intracranially. The lesion exhibits T1W and T2W hyperintense signal and fat suppression. Minimal peripheral enhancement post gadolinium is also seen. (9)

**Conclusion**

This case illustrates the classic clinical presentation and typical imaging findings of nasal glioma in a child. Early diagnosis of nasal glioma and differentiating it from encephalocele and nasal dermoid with MRI helps in the clinical management.

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