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Case Report

Nasal Glial Heterotopias: Diagnostic Role of Histopathology

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Abstract: Nasal glial heterotopias (NGH) are rare nonhereditary congenital malformations composed of heterotopic neuroglial tissue. Congenital midline nasal masses are rare and have been reported to occur in 1 in 20000-40000 livebirth. NGH accounts for 5% of them. NGH usually presents during infancy but occasionally in older children and adults. Evaluation should be done with CT scan & MRI to rule out intracranial extension. There have been several cases reported in which NGH were misdiagnosed as capillary hemangiomas, dermoid cyst, teratoma, encephaloceles and even desmoids. A near definitive diagnosis can be carried out by following a proper clinical, sonological and including CT, MRI evaluation, while surgical excision and histopathological confirmation is of gold standard. We report a rare case of midline 3×3×3cm firm, subcutaneous, non-tender, non-pulsatile, non-compressible, non-reducible with posture and pressure swelling, covered with bluish red skin at root of nose. On CT scan and MRI swelling was given differential diagnosis of encephalocele, NGH and dermoid. It was histopathology which gave definitive diagnosis of NGH. The case represents importance of histopathology as gold standard in diagnosis of NGH.
Keywords: Nasal glial heterotopias, Encephalocele, Histopathology.

INTRODUCTION

Nasal glial heterotopias (NGH) are congenital malformations of displaced glial tissue in which intracranial meningeal continuity has become obliterated. Differing from NGH encephalocele has herniation of brain tissue and leptomeninges through a bony defect of skull which maintain intracranial continuity [1]. NGH is also known as nasal glioma representing collection of normal glial tissue in an abnormal location [2]. NGH is rare congenital lesion first described by Reid in 1952 and the term glioma was coined by schimdt in 1900 [3].

The term nasal glioma is a misnomer as it is not a true neoplasm. It actually consists of ectopic nerve tissue containing neuroglial elements, with glial cells in a connective tissue matrix that may or may not have connection to the subarachnoid space or dura [4, 5]. The incidence of congenital mid line nasal masses is 1:20000 to 40000 with male to female preponderance 3:2 [7]. NGH is frequently diagnosed in newborns or infants rarely in adults [2], 250 cases have been reported so far [6].

Histologically gliomas are composed of predominantly mature astrocytes and neuroglial cells with varying degree of stromal fibrosis covered with respiratory epithelium [2, 8]. Excision is curative with no complications. Recurrence is rare.

NGH often misdiagnosed as encephalocele, midline dermoids, hemangiomas. Even after a proper systematic approach involving sonography, CT (computerised tomography) and MRI (magnetic resonance imaging) a near definitive diagnosis can be made [9]. Thus to reduce diagnostic error it is advised that each and every excised specimen should be subjected for histopathological examination and if required immunohistochemistry (IHC) to achieve 100% diagnostic accuracy.

CASE REPORT

A 1 year old infant presented with swelling at root of nose more towards right since birth. The swelling was firm, non-tender, non-pulsatile, non-discharging, non-compressible and non-reducible with posture/pressure. The swelling progressively increased to present size of 3×3×3cm. Patient did not have any history of epistaxis or history suggestive of meningeal irritation and difficulty in respiration. Intercanthal distance was increased. Other biochemical investigations were within normal limit.

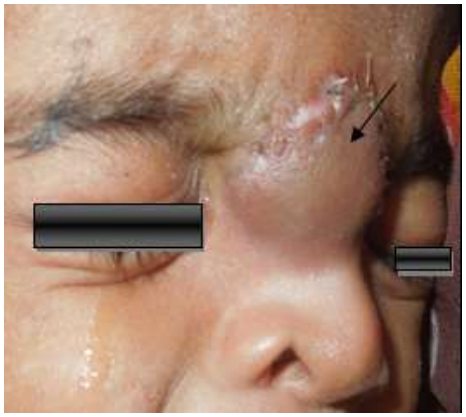

Fig.1: Clinical photograph showing swelling over root of nose

CT scan showed well defined lobulated 2.6×2×3cm heterogeneous isoechoic lesion having soft tissue component with septations within with possible defect in underlying crysta galli. Features suggestive of? encephalocele? dermoid.

MRI brain plain and contrast study revealed well defined altered signal intensity lesion anterior to glabella in midline more towards right side with possible communication to right nasal cavity without any intracranial communication, features suggesting nasal glioma.

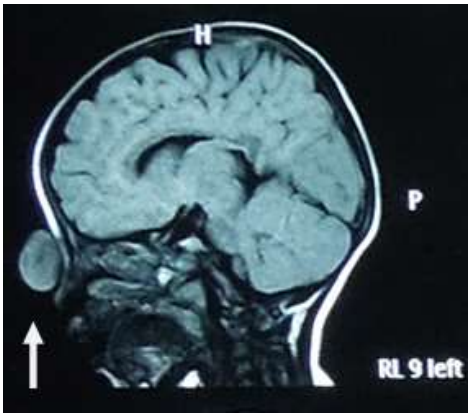

Fig. 2: MRI brain showing well defined altered spinal intensity lesion anterior to glabella, no obvious intracranial extension

After excision the mass was sent for histopathological examination. On gross examination it was well circumscribed soft to firm of size 3×3×3cm. Cut section was homogenous gray white. No areas of haemorrhages, cyst or calcification noted. Microscopy showed fibrillary neuroglial cells in connective tissue matrix, predominantly astrocytes in background of neuropil.

Considering clinical history, radiological and histopathology findings final diagnosis of nasal glial heterotopia was given.

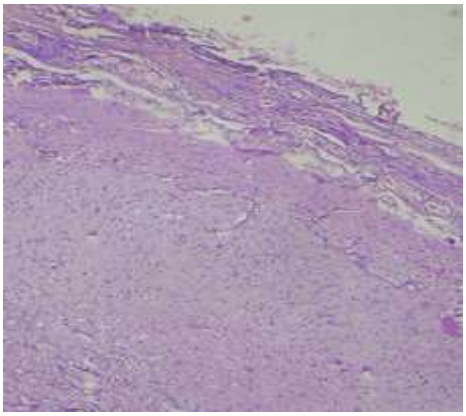

Fig. 3: Well encapsulated normal appearing neural tissue beneath respiratory epithelium (H & E 100X)

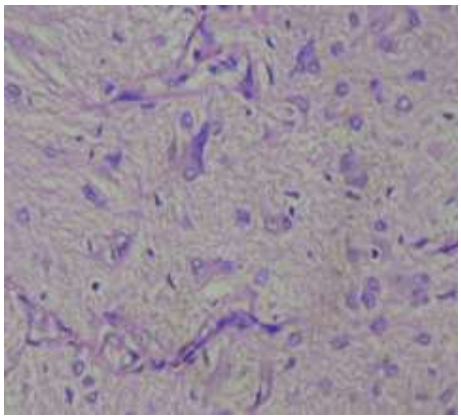

Fig. 4: Normal neural & glial tissue (H & E 400X)

So in our case histopathology has solved the diagnostic dilemma due to difference in CT and MRI opinion.

DISCUSSION

Congenital midline nasal masses are rare anomalies. They have been reported to occur in about one in 20000–40000 live births [6, 10]. Nasal gliomas account for approximately 5% of all congenital nasal swellings [11] and 0.25 % of all tumours in nasal cavity [12].

The term "nasal glioma" is a confusing misnomer as it implies a neoplastic condition, which it is not. It needs to be differentiated from glioma, which is a malignant tumor of the brain, nasal glioma representing collection of normal glial tissue in an abnormal location.

Nasal gliomas are CNS masses of neurogenic origin, which have lost their intracranial connections and present as an obvious extranasal or intranasal mass at birth without associated surgical symptoms of encephalocele [10]. Neuroimaging is essential for the characterization of these lesions, in order to determine the exact location of the lesion and most importantly to exclude a possible intracranial extension or connection [13]. CT scan or MRI forms are more essential for the investigation as fine needle aspiration cytology or

excision biopsy have found to be associated with a significant risk of meningitis or CSF leaks [14]. CT scan demonstrates bony defects, and MRI scans are preferable due to their superior soft tissue enhancement [15].

Types of nasal gliomas:

• Extra nasal (60%): They lie external to the nasal bones and nasal cavity, most commonly occur slightly off the midline at the bridge of the nose.

• Intranasal (30%): They are found within the nasal or nasopharyngeal cavity, the oral cavity or rarely the pterygopalatine fossa.

• Combined (10%): They consist of a communication between the extranasal and intranasal components occuring via a defect in the nasal bones or around the lateral edges of the nasal bones [9].

To understand the development of congenital midline nasal masses, knowledge of the normal embryological development of the nose is important.

The most known embryological theory was described by Grünwald in 1910 and is called the "prenasal space" theory which explains embryopathogenic continuum proposed among dermoids, gliomas, and encephaloceles [16].

The possible theories of development of nasal gliomas [11, 17, 18]:

- Sequestration of glial tissue of the olfactory bulb entrapped during cribriform plate fusion.
- Ectopic neural tissue cells
- Encephaloceles with lost intracranial connection and meningeal continuity
- Inappropriate closure of the anterior neuropore (fonticulus frontalis).

Surgical excision of the tumor is the treatment of choice. Endoscopic excision is preferred in intranasal cases. Our case was extra nasal at root of nose more towards right.

Inadequate primary excision results in a 4 - 10% recurrence [19]. Evidence in the form of recurrence of nasal glioma questions whether they are benign neoplasms or simply malformations [20].

Histologically nasal glioma consist of fibrillary neuroglial tissue with a prominent network of glial fibres, gemistocytic type astrocytes predominantly in background of neuropil, representing classic neuroglial tissue with varying degree of glial fibrosis [9]. There have been several cases reported in which nasal gliomas were misdiagnosed as capillary hemangiomas, dermoid [16]. In one case there was confusion even on histopathology having difference in opinion among two pathologist as desmoids tumour versus nasal glioma [12]. Here comes the role of immunohistochemistry (IHC) as GFAP (glial fibrillary acidic protein positivity) is diagnostic. Thus histopathology if required aided with IHC is gold standard.

In our case histopathology has resolved diagnostic dilemma, CT scan and MRI being of differing opinion.

In NGH no fluid filled space is connected to the subarachnoid space. In general, the lesions present as a red or bluish lump at or along the nasomaxillary suture, or as an intranasal mass that are characteristically firm, non compressible, do not increase in size with crying, and do not transilluminate [9]. They may be associated with a widened nose or with hypertelorism that is secondary to growth of the mass. If they are left untreated, they can cause deformity of nasal bones and adjacent structures [7].

CONCLUSION

Nasal gliomas should be considered in the differential diagnosis of a nasal mass in an infant. A systematic approach should be employed for the diagnosis involving clinical, sonological & including CT/MRI evaluation in order to obtain a near definitive diagnosis; however surgical excision and histopathological confirmation is of gold standard [9]. Importance of histopathology in solving diagnostic dilemma prompted us to report this rare case.

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