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Congenital Nasal Glioma: A Rare Case Report

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Abstract

Background: Nasal glioma, also known as nasal glial heterotopia, is a rare congenital lesion that is thought to be the result of abnormal embryonic development. Nasal glioma was reported for the first time by Reid in 1852 and are extremely rare with an incidence of 1 in 20,000 to 40,000 with very few cases being reported in literature.

Case presentation: We hereby report a rare case of congenital nasal glioma in a two year old boy who presented with a mass on the dorsum of the nose since birth with no significant increase in size.

Conclusion: Nasal gliomas are rare masses that are usually benign. They are embryologically related to encephaloceles but are unique without any intracranial connection. Thus, imaging and histopathology are paramount in the diagnosis of nasal gliomas.

Keywords: Congenital, Glioma, Heterotopia, Nasal

Introduction

Nasalglioma, also known as nasal glial heterotopia, represents a collection of normal glial tissue in an abnormal location; the tissue is isolated from the nervous system without intracranial connection.[1] Nasal glioma was reported for the first time by Reid in 1852 , but the term itself was introduced by Schmidt in 1900.[2] Nasal glioma is a rare congenital lesion that is thought to be the result of abnormal embryonic development. Nasal gliomasare extremely rare in neonates with an incidence of 1 in 20,000 to 40,000 with very few cases being reported in literature.Since they have no communication with the central nervous system, nasal gliomas are oftenasymptomatic but can present with respiratory distress depending on the size and location of the tumor.[3] Although the majority of these benign congenital tumors are found in the nasal region and occur on the bridge of the nose, some will be located intranasally, and the remaining few may be seen elsewhere in the facial region.[4] We hereby report a rare case of congenital nasal glioma in a two year old boy.

Case Presentation:

A two year old boy came with a complaint of swelling at the bridge of nose since birth with no significant increase in size. He underwent ultrasound examination followed by MRI and diagnostic nasal endoscopy to rule out intracranial extension. CT PNS revealed a tiny defect likely extranasal glioma. Later he underwent surgical excision of the mass in the Department of Otorhinolaryngology in our Institute and excision biopsy was sent for histopathological examination.

Figure1: Clinical image of the patient with mass on the dorsum of nose at two months age

Figure2: Clinical image of the same patient at two years, presenting with mass on the dorsum of the nose without any significant increase in size

Figure 3: CT PNS of the patient showing tiny defect (2.5mm) in right nasal bone

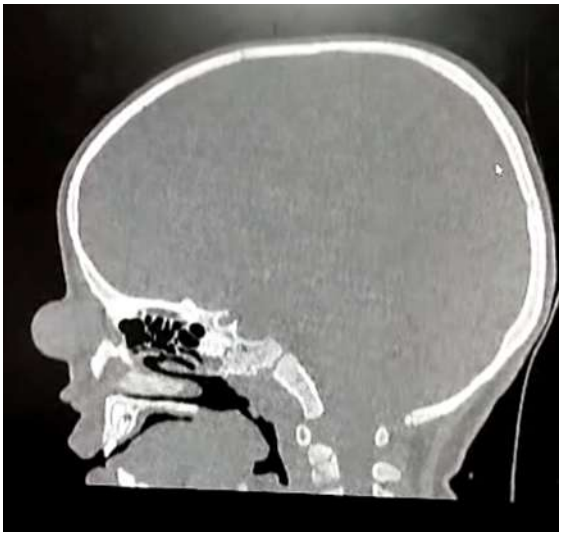

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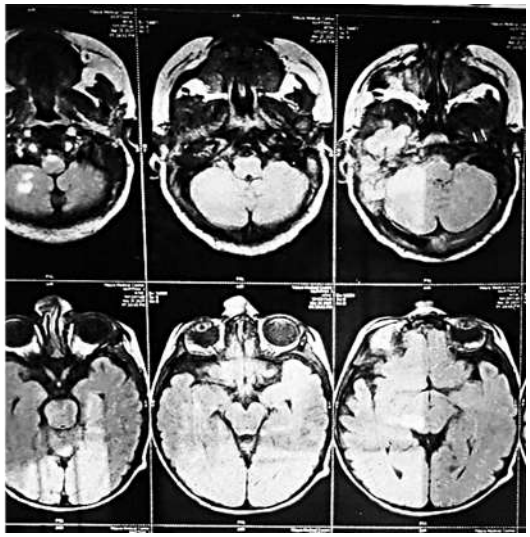
Figure 4: MRI of the patient showing no intracranial extension



Microscopic examination:

In the Department of Pathology, we have received a single, cut-opened, skin covered mass measuring 1.5cm in the greatest dimension. Cut section is grey-white from which a full thickness section was taken and processed for routine histopathological examination. In general, the histology of the nasal glial heterotopia is characterized by neuropil interlaced with fibrous and vascular connective tissue. Neurons and gemistocytic astrocytes may be seen in some lesions. It may be arranged in a lobular pattern, and cystic structures may be present as well.[5]The histologic picture may vary in places, and may be difficult to identify with hematoxylin-eosin stain alone; thus special stains like Masson trichrome and immunohistochemistry are of great utility when making the diagnosis. The glial tissue can be confirmed by immunoreactivity for glial fibrillary acidic protein (GFAP) or S100 protein.[6] In this case, on histopathological examination, the sections studied from the lesion showed skin lined by keratinized stratified squamous epithelium with underlying normal adnexal structures and focal areas of hyalinised collagen bundles.Deeper dermis is cellular consisting of few mature glial elements, astrocytic giant cells, nerve bundles and neuropil. Furthermore, the paraffin embedded section was sent forImmunohistochemistry and glial tissue showed diffuse strong positivity for GFAP (Glial fibrillary acidic protein). Hence the final diagnosis given on histopathological examination was Nasal Glioma.

Figure 5: Skin lined by keratinized stratified squamous epithelium with underlying normal adnexal structures (H& E 40X)

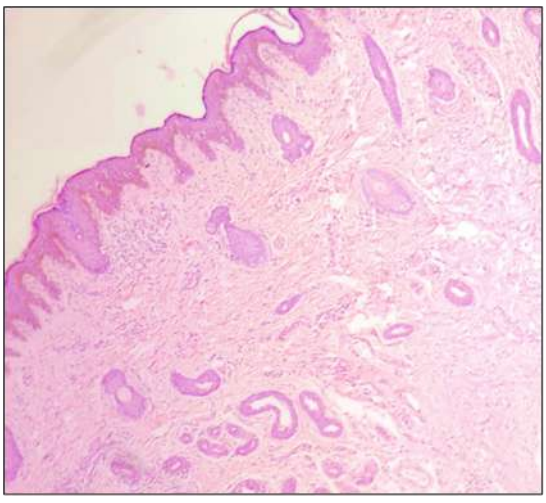


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Figure 6: Dense fibrocollagenous tissue with areas of hyalinization (H& E 100X)

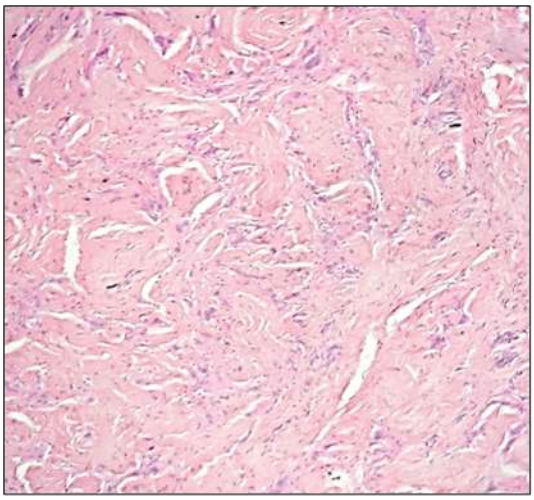

Figure 7: Mature glial tissue with astrocytic giant cells and neuropil (H& E 400X)

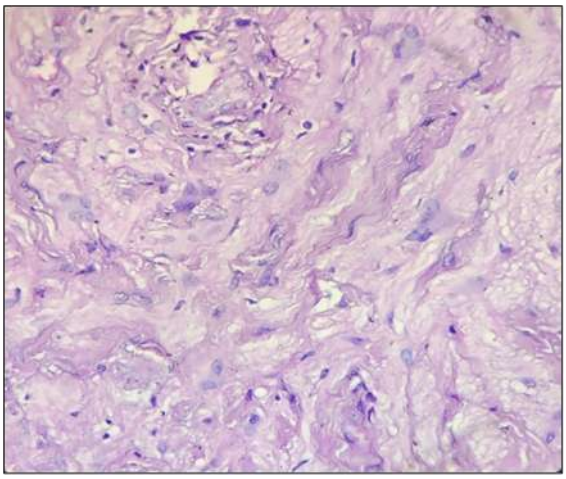

Figure 8: Areas of focal lymphocytic infiltration (H& E 400X)

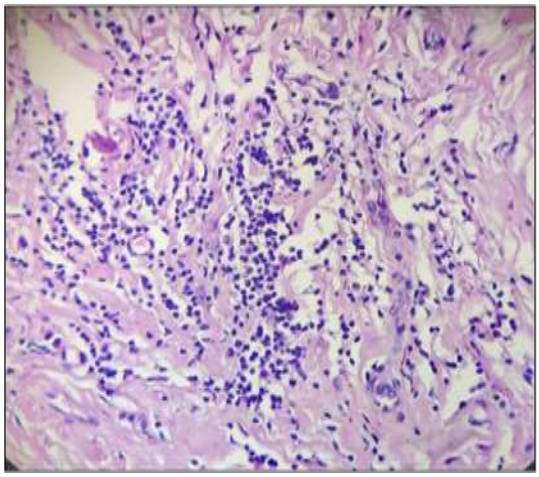

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Figure 9: Masson trichrome stain highlighting glial tissue in red colour and fibrous tissue in blue colour (400X)

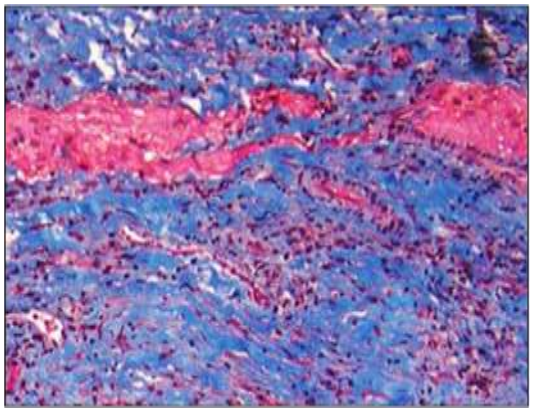
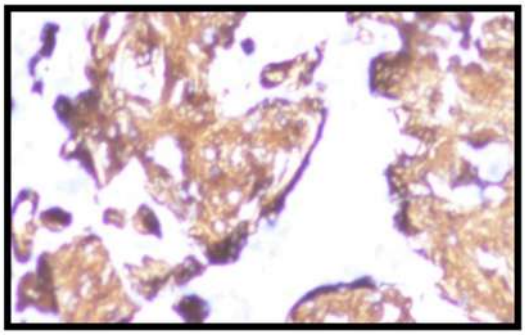


Figure 10: Glial tissue showing diffuse strong positivity for GFAP (Glial fibrillary acidic protein) on immunohistochemistry (400X)



Discussion:

Nasal glioma is a non-hereditary, benign, congenital malformation, embryologically related to encephaloceles.[7] It is a rare lesion occurring once in 20,000 to 40,000 live births with a total of 264 cases reported in the world literature since the first description by Reid in 1852.[8] Encephaloceles and gliomas have a similar embryologic origin, but as the encephalocele is a herniation of cranial contents through a defect in the skull, a glioma is thought to be an encephalocele that has lost the intracranial connection.[9] Gliomas are locally aggressive lesions usually present at birth. 90% of cases are diagnosed before the age of 2 years.[10] Our case also showed a classic presentation at the age of two years. The discrimination of nasal glioma (glial heterotopia) from encephalocele is based on the presence or absence of the connection between the mass and the intracranial tissue. However, even with highresolution computed tomography and magnetic resonance imaging, the connection may be very small and unapparent. However, bony defects may also be seen in association with nasal gliomas while still showing no communication with the brain parenchyma.[11]In our case also MRI ruled out intracranial extension and CT PNS showed a tiny defect(2.5mm) in the right nasal bone. The medical literature characterizes nasal glial heterotopia as being a congenital mass presenting within the first year of life, with rare examples occurring later in life. For congenital lesion, the term nasal glioma is a more accurate description than nasal glial heterotopia.[12] About 60% of nasal gliomas are found extranasally, 30% intranasally, and 10% can be of mixed type seen in both the locations.[13]Extranasal mass can be asymptomatic causing minimal symptoms but intranasal lesions can present with nasal obstruction or nasal deformity or may present as mass protruding from a nostril, or more frequently within the nasal cavity or nasopharynx.[14] In this case, the lesion is present since birth without any significant increase in size , also without any causing any significant distress to the patient which could be the sole reason

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for delayed presentation. Definitive diagnosis is often only possible after complete surgical excision as biopsy or fine needle aspiration of nasal masses is contraindicated because of the increased risk of infections or meningitis.[15] Diagnosis of nasal glial heterotopia is difficult if few glial elements and astrocytic giant cells are seen within dense fibrous connective tissue like in this case. Thus in a young child where the clinical suspicion is high, special stains or immunohistochemistry would ensure a correct diagnosis.

Diffuse strong positivity for GFAP (Glial fibrillary acidic protein)on immunohistochemistrysupported the diagnosis of nasal glioma in our case. Though this lesion has a slow growth rate and is benign without any potential for malignant transformation, delays in treatment may lead to distortion of the septum and destruction of nasal bone due to the locally aggressive nature of the tumour.Postoperative radiological follow-up is fundamental sinceoverall recurrence rates of 4 to 10% have been reported.[16]

Conclusion:

Nasal gliomas are rare masses that are usually benign. They are embryologically related to encephaloceles but are unique masses without any intracranial connection. Thus, imaging and histopathology are paramount in the diagnosis of nasal gliomas. Surgical excision is the mainstay of treatment and regular follow up of the case is mandatory to prevent recurrence and complications.

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