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Case Report

Nasal neuroglial heterotropia: a rare mimicker of dermoid cyst: a case report

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ABSTRACT

We report an uncommon case of nasal neuroglial heterotopia (NGH, glioma) presenting as midline nasal mass. Review of literature, imaging challenged and surgical management options are discussed. Unlike the more common nasal dermoid, 20% of NGH have intracranial communication which can cause post-operative life-threatening complications like CSF leak and meningitis.

Keywords: Frontonasal mass, Midline nasal mass, Nasal glial heterotopia, Nasal glioma

INTRODUCTION

Congenital midline nasal masses are rare, presenting in 2-5 per 100,000 live births with 5% to 7% of them being nasal gliomas.[1] Differential diagnoses of midline mass include nasal dermoid cysts and less commonly nasal glial heterotopia (NGH) or encephaloceles.[2] NGH (nasal glioma) is a congenital, relatively rare displacement of cerebral tissue in extracranial sites. They are non-neoplastic benign with collections of heterotopic tissue of neurogenic origin. Approximately 20% retain intracranial connectivity. In a recent first systematic review of 72 original publications along with a case report revealed 152 NGH, incidence being more in males with ratio of 3:2. Majority were diagnosed in childhood (92%) with 84% diagnosed and treated before 3 years of age. NGH was the most frequently located intra-nasally (45%), followed by extra-nasally (36%), least being mixed (19%). The extra-nasal type is noted after birth as a firm, non-compressible mass on the nasal dorsum, often with a reddish/ bluish appearance.[3]

The development of the frontonasal region (anterior neuropore) is complex. The development of NGH occurred in the process of cribriform plate fusion. During embryogenesis, a defect of anterior neuropore closure results in herniation or sequestration of tissue through the foramen cecum (located anterior to cribriform plate and posterior to frontal bone) or fonticulus frontalis (located between frontal and nasal bones). Hence NGH can have intracranial extension due to their central nervous system origin.[4] At times, though no patent intracranial connection, in 15%-20%, its remnant forms a fibrous stalk to the meninges and intracranial space. Identifying connection between these mass and anterior cranial fossae, as well as its content characteristics is imperative, in decision making for therapeutic intervention. Failing this, can lead to life threatening complications of CSF leak and meningitis.[5]

Histopathologically, NGH is characterized by fibro-connective tissue with focal infiltration by bundles of glial tissue. This is typically immune-reactive for glial fibrillary acid protein, S-100, synaptophysin, and epithelial membrane antigen.[6,7] Encephaloceles can also contain variable amounts of glial tissue. The presence of leptomeninges helps distinguish encephalocele from NGH. If no ependymal tissue is present, then the diagnosis is based on clinicopathological and imaging findings.[7,9]

CASE REPORT

A one-year-old male child presented with an swelling over the dorsum of nose which has been present since birth with no other associated symptoms. His perinatal history was uneventful and the family history was unremarkable.

A 1×1 cm globular firm, nonpulsatile swelling located at the root of nose just to the right of midline with normal healthy skin over was noted (Figure 1). There was no change in size of swelling on crying and Frustenberg's test was negative. Nasal endoscopy revealed no intranasal extension. CT scan of paranasal sinuses including skull base revealed well-defined soft tissue density present in the midline of fronto-nasal region causing deformity of the nasal bones, displacement of the nasal septum to the right (Figure 2A) with no bony defect in the cribriform plate (Figure 2B).

With a working diagnosis of dermoid cyst, excision biopsy of the lesion was done. Intraoperatively too, it was seen that there was no intracranial extension (Figure 3 A and B) The histopathological examination, however revealed glial tissue with fibrosis and vascular proliferation, suggestive of glioma (Figure 4). Postoperative period was uneventful.

Figure 1: Swelling over root of nose.

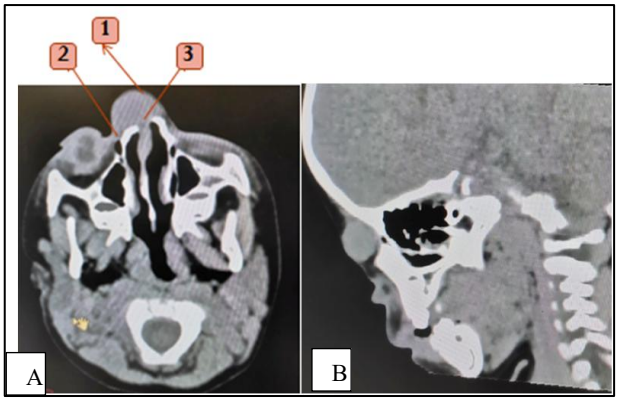

Figure 2 (A and B): CT scan of nose and PNS-axial view. CT scan nose and PNS- sagittal view showing no bony defect.

1.Well defined midline soft tissue density lesion in frontonasal region. 2. Mild medial displacement of the nasal bone and 3. Mild widening of internasal sutures.

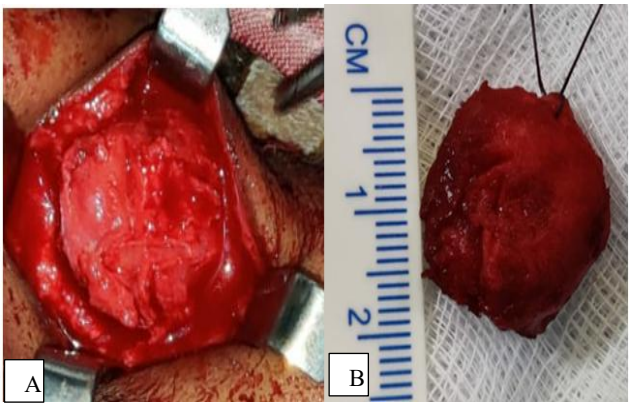

Figure 3 (A and B): Exposure of nasal mass via vertical. Excised specimen incision.

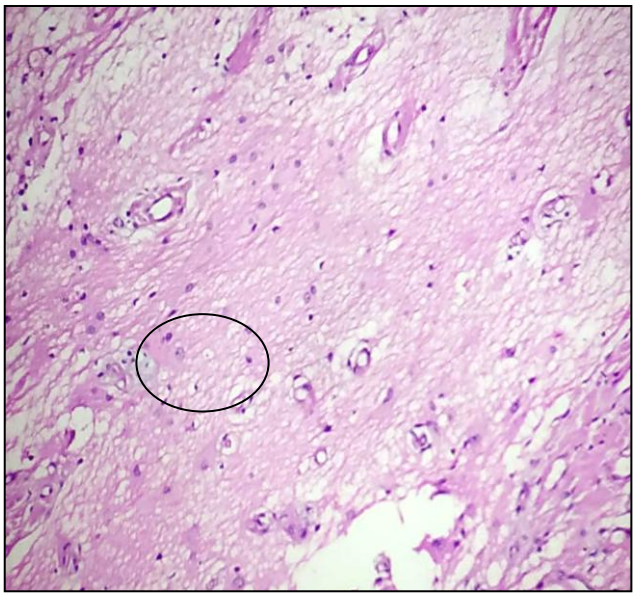

Figure 4: Glial tissue with fibrosis and vascular proliferation (H and E stain under 40× magnification).

DISCUSSION

Congenital midline frontonasal masses are rare but important anomalies that present in children. Abnormal closure of the frontal and nasal bones during embryologic development results in incomplete separation of ectodermal and neuro-ectodermal elements. Nasal masses with actual or potential central nervous system connections are nasal dermal sinus cysts, encephaloceles and neuroglial heterotopia (NGH or glioma).[1,3-5] Intranasal NGH or gliomas present with airway obstruction in the newborn and in older children as nasal obstruction, epistaxis or CSF rhinorrhea, lesions being seen with middle turbinate or higher above. It occasionally mimic nasal polyps. Extra-nasal NGH presents as mass at glabella level, nasal deformity or widening. It is firm non-compressible, not trans-illuminate, not increasing in size with crying and Furstenberg sign negative. Expansion of the nasal lesion with compression of the internal jugular vein (Furstenberg's sign) can indicate an intracranial connection. Extra-nasal frontonasal mass which is discolored and soft, is more likely to be a hemangioma than neuroglial heterotopia.[7,8]

CT and MRI are the imaging of choice for these lesions, both being complementary.[4,5,8-10] Contrast enhanced MRI is the preferred imaging modality to evaluate a midline frontonasal mass for excellent soft-tissue characterization as well as identifying intracranial communication, if present. Thin-section, high-resolution multi-planar MRI with contrast-enhanced images delineates cartilaginous skull base, sagittal view being most valuable. NGH are hypo or iso-intense to gray matter on T1-weighted images, and hyper-intense on T2-weighted images. Diffusion-weighted imaging helps differentiate glioma from epidermoid cysts, the latter having increased signals. Absence of enhancement, differentiates NGH from venous malformation. Herniation of meninges alone (meningocele) or brain and meninges (encephalocele) are also apparent.[3-5,8,9] CT interpretation in young children is challenging as anterior skull base is only partially ossified, rest being cartilaginous and membranous. The cribriform plates, being non-ossified, appear radiolucent during the first year of life with ossification of crista galli beginning at 1 year of age and completing by 5 years. Foramen cecum is located between frontal bone and crista galli and normally measures up to 2 mm in diameter as well.[5,10] On CT images, NGH appears as a large, well-defined, non-enhancing soft tissue mass that is intranasal or over the nasal dorsum. Thin-section, high-resolution axial and coronal 'Limited CT' is to be done following MRI, if latter reveals intracranial connection. This avoids un-indicated CT induced ionizing radiation exposure to the child.[3-5,9,10]

NGH is diagnosed by presence of fibroconnective tissue with focal infiltration by bundles of glial tissue, which is typically immune-reactive for glial fibrillary acid protein, S-100, synaptophysin, and epithelial membrane antigen. Though variable amounts of glial tissue are also seen in encephaloceles, presence of leptomeninges distinguishes it from NGH.[6,7] Biopsy of a pediatric midline frontonasal mass is contraindicated. The treatment of choice is surgical excision and biopsy. Urgent surgical intervention is indicated if there is evidence of intracranial connection or CSF rhinorrhea to avoid meningitis and in neonates presenting as airway obstruction secondary to intranasal mass, as increased work of breathing can lead to even respiratory failure, neonates being obligate nasal breathers.[3-5] In recent years, reports of complete removal of intranasal and mixed NGH using endonasal endoscopic techniques and stereotactic navigation system along with skull base plasty to cover a bony defect or to treat a cerebrospinal leak have been published.[3,5,8]

Clinical significance

We report an uncommon case of nasal neuroglial heterotopia (NGH, glioma) presenting as midline nasal mass. Review of literature, imaging challenged and surgical management options are discussed. Unlike the more common nasal dermoid, 20% of NGH have intracranial communication which can cause post-operative life-threatening complications like CSF leak and meningitis

CONCLUSION

Congenital frontonasal/nasal mass, including NGH, can have intracranial extension as they are developmental anomaly of the anterior neuropore. Contrast enhanced MRI is the initial imaging of choice, as it identifies crucial intracranial communication along with soft tissue characterization. 'Limited CT' is reserved for those with intracranial connection to identify bony defects details for surgical intervention. Though surgical excision was traditionally by external or trans-nasal approach, complete removal using endonasal or pre-nasal endoscopic techniques under stereotactic navigation system along with skull-based plasty and multidisciplinary involvement is advised.

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