

Case of Nasal Glial Heterotopia: A Diagnostic Dilemma

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ABSTRACT

Nasal glial heterotopia (nasal glioma) is a mass of mature brain tissue isolated from cranium, located mostly on the nasal bridge. We present a case of nasal glial heterotopia in a 1-year-old female child. She came with a swelling over nasal dorsum since birth which was fluctuant, non-tender. It did not increase on crying or coughing. Computed tomography scan showed soft tissue density over nose with extension in between nasal bones, no intracranial extension but a small oval defect in crista galli anteriorly. Based on the clinical picture we made a provisional diagnosis of congenital nasal dermoid. Child was taken for excision of mass under general anesthesia once her blood parameters were normal. The mass was approached through a central midline incision over nose. The adherent posterior part of the mass was dissected meticulously from the nasal bones without any residual damage. Histopathological examination of the mass revealed it to be nasal glial heterotopia.

Key words: Mass, nasal glial heterotopia, nasal dorsum

INTRODUCTION


Nasal glial heterotopia (nasal glioma) is the term used to describe a mass of mature brain tissue isolated from the cranium.^[1,2] Majority of these benign congenital tumors are found on the bridge of the nose, some are seen intranasally, and very few are seen elsewhere on the face.^[3-7]

Nasal glial heterotopia is a rare congenital lesion resulting due to abnormal embryonic development. It is frequently seen in newborns and infants and is rarely found in adults. Histologically, these tumors

are composed of astrocytes and neuroglial cells interspersed within the fibrovascular connective tissue. It is lined by the epithelium or respiratory mucosa.^[8] Nasal glial heterotopias have no communication with the central nervous system. However, from computed tomography (CT) findings, 15–20% of patients have a defect in the cribriform plate.^[9,10] We present a case of nasal glial heterotopia in a 1-year-old child which was adherent to the nasal bones.

CASE REPORT

A 1-year-old female child of 8 kg came with complains of progressively increasing swelling at the dorsum of the nose since birth. On examination, the swelling was of size 2 cm × 2 cm present over the root of nose and dorsum. It was a non-tender, fluctuant swelling with no increase of size on coughing or crying. The overlying skin was normal, and no sinus or punctum noted. Although the overlying skin was freely mobile, the swelling appeared to be adherent to underlying

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nasal bones. The child was also suffering from upper respiratory tract infection. Her blood indices were hemoglobin 9 mg/dl, total leukocyte count - 13500, differential leukocyte count - 30/60/5/5, and platelets - 44,300. Prothrombin time 15.1, international normalized ratio - 1, peripheral blood smear showed anisosis, poikilocytosis, microcytosis, presence of pencil cells, few target cells, and occasional schistocytes with mild to moderate hypochromasia. Her blood group was A positive. Her C-reactive protein was positive, and titer was 48 mg/dl. Her dengue iggg/igm/ns1 was non-reactive and typhoid tests were also negative. On further investigation, her CT brain showed soft tissue density in subcutaneous tissue over nose anteriorly. No significant fat density or calcification is seen within it. Small defect seen in midline between nasal bone and soft tissue is seen herniating through it. Small oval defect is seen in crista galli anteriorly. No large intracranial mass was seen. Findings likely suggest possibilities of encephalocele, nasal dermoid. Neurosurgeons opinion was also sought who mentioned that the swelling was superficial with small extension below nasal bone, no intracranial extension, most likely dermoid. Child underwent treatment for viral urti and oral hematinics were also administered. Once her urti was treated, and hemoglobin was improved, she was taken up for excision of mass under general anesthesia. Under aseptic precaution, a midline skin incision was given over dorsum of nose extending from glabella to supratip area. Lateral flaps were elevated, and cyst identified. Meticulous dissection is done all around the cyst. The cyst was found to be attached to nasal bone inferiorly. The attachment was excised, and the cyst delivered. The cyst was 15 mm × 20 mm oval in shape with smooth surface. Betadine irrigation was given in the wound and wound closed using 6-0 vicryl. Pressure bandage was applied. Child was given IV antibiotics. Dressing removed on 2nd post-operative day and child was discharged and followed up after 5 days for suture removal. Histopathological examination of the mass showed areas of gliosis separated by fibrocartilaginous tissue and also seen in the skeletal muscle fibers at periphery. No granuloma or malignancy noted. Moreover, the impression is Nasal glial heterotopia [Figures 1-3].

DISCUSSION

Nasal glial heterotopia is a rare condition, due to entrapment of neuroectodermal tissue during the closure of the covering of the brain, or a nasal



Figure 1: Pre-operative picture



Figure 2: Intraoperative picture of child after excision of mass

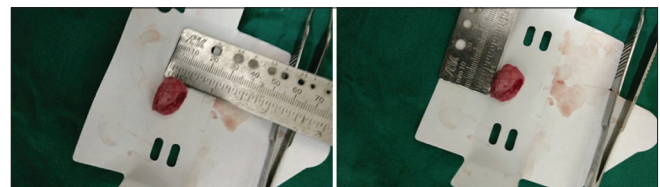


Figure 3: Excised mass

encephalocele which is lined by meninges and later disconnected from the intracranial cavity during subsequent development.^[11,12] The distinction of nasal glial heterotopia from encephalocele is based on the presence or absence of the connection between the mass and the intracranial tissue. However, the connection may be very small to identify even with high resolution computed tomography and magnetic resonance imaging. Furthermore, bony defects may

be seen in association with nasal gliomas while still showing no communication with the brain parenchyma because of developmental anomaly.^[11,13] The distinction may be made by the presence of meningitis and/or cerebrospinal fluid rhinorrhea either before or after surgical manipulation.^[3,11,12]

Clinically, nasal glial heterotopia may involve the external nose (60%), the internal nose (30%), or both sites (10%). Extranasal tumors manifest as a tumor mass usually on the bridge or side of the nose.

Most of these will not show any connection to the brain.^[11] Intranasal glial heterotopia present with nonspecific symptoms for nasal cavity masses: Nasal obstruction, polyps, rhinorrhea, and chronic otitis media. Complete surgical excision is the treatment of choice, although recurrences after surgery occur rarely.

Biopsy or fine needle aspiration of childhood nasal masses is contraindicated as it risks removal of functional brain tissue and increases the risk of meningitis.^[1] The histology of nasal glial heterotopia is characterized by mature glial cells (astrocytes and oligodendrocytes) in stromal connective tissue, lined by the respiratory epithelium.^[14]

Pathologically nasal glial heterotopia and encephalocele cannot be differentiated yet.^[15] Encephalocele is protrusion of the brain parenchyma connected to the rest of the brain by a pedicle and is associated osseous defect.^[12] Nasal glial heterotopia is thought to be basal or frontal encephalocele that has lost its intracranial meningeal connection.^[8] Therefore, nasal glial heterotopia has no connection with the subarachnoid space or central nervous system.

Differentiating these conditions is mainly based on clinical and radiological features. The Furstenberg test examines whether the mass enlarges on compression when the ipsilateral jugular vein. This test is positive in encephalocele.^[14] CT is useful for the visualization of bony defects in the anterior skull base, whereas magnetic resonance imaging gives complementary information regarding the fluid or soft tissue characteristics.

In our case child presented with a mass over the nasal dorsum since birth, which was slowing increasing in size. Since there was no increase in size on coughing or crying and CT scan showed no connecting stalk, a provisional diagnosis of nasal dermoid was made. However, CT scan did show a defect in anterior part of cribriform plate without brain herniation. Histopathology of the mass revealed it to be nasal glial heterotopia.

CONCLUSION

From our experience with this patient, we put forward the fact that nasal gliomas should be considered in the differential diagnosis of a nasal mass in children.

A systematic approach should be employed for the diagnosis involving clinical and radiological evaluation to obtain a near-definitive diagnosis; however, surgical excision and histopathological confirmation are of the gold standard. Importance of histopathology in solving diagnostic dilemma prompted us to report this rare case.

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