

Nasal Glial Heterotopia-A Case Report

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Abstract: Nasal glial heterotopia is a rare congenital non-neoplastic lesion that usually occurs during infancy. We report a case of nasal glial heterotopia in a 4-month-old female child in which the initial clinical diagnosis of Nasal Teratoma was entertained but histomorphologic features are consistent with Nasal glial heterotopia.

Keywords: Glial Fibrillary Acid Protein (GFAP), CT (Computed tomography), MRI (Magnetic resonance Imaging).

Introduction

Nasal glial heterotopia is a congenital malformation composed of displaced normal mature glial tissue without an intracranial communication [1]. It is the most common congenital nasal mass lesion seen during embryonic development resulting from failure of the neuroectodermal and ectodermal tissues to separate during development of the nose [2]. Although, this lesion is rare but clinically they are important because of their potential for connection to the central nervous system [3]. It is most often diagnosed at birth or in early childhood with few cases reported during adulthood [2]. It is extra-nasal in 60% of cases, intra-nasal 30% or mixed in 10% [3]. Other reported rare locations include the lip, tongue, scalp, nasopharynx and oropharynx. Patients usually present with polypoid mass seen at birth or within few months after birth. The reported incidence is 1 in every 20,000 to 40,000 birth [4]. The mass is usually soft to firm subcutaneous nodule at the bridge of the nose, within the nasal cavity or anywhere along the upper border of the nasal bridge [3].

Nasal glial heterotopia mass lesions could be complicated by the obstruction, chronic rhinosinusitis or nasal discharge. In rare cases, the incidence of cerebrospinal fluid leaks which suggest intracranial connection and hence a different entity encephalocele. This must be ruled out by CT scan and or MRI. Histologically, it is composed of mature astrocytes and neuroglial fibres intermixed with fibrovascular connective tissue and stromal fibrosis. Occasionally, neurons may be present. These cells (astrocytes, neuron) are S-100 positive and glial fibrillary acid protein (GFAP) positive confirming their neuroglial origin.

We report a 4-month-old with this rare tumour to enunciate the challenges in managing the condition and the significant role of histology in making the diagnosis.

Case Report

A 4-month-old female child presented with an abnormal swelling at the inferolateral aspect of the left nostril displacing the left nostril superomedial. The mass was noticed at birth. It was initially a thumb-sized lesion but progressively increased in size with the growth of the patient. There was no history of swelling in any other part of the body or history of difficulty in breathing. The child was a

product of a term pregnancy delivered via spontaneous vaginal delivery to a 32-year-old P₃+0 (3 alive) woman. The pregnancy was booked at 4 weeks of gestation age and the mother had regular antenatal visits. She had 2 doses of Tetanus toxoid vaccine during the pregnancy. There was no history of consumption of alcoholic beverages, cigarette smoking, use of herbal concoction, exposure to radiation, usage of unprescribed drugs and fever during index the pregnancy. The index patient is the third child of 3 children in a monogamous family. There was no history of such abnormality in other siblings.

On examination, the mass was attached to the overlying skin and underlying nasal bone. It measured approximately 3.0cmx3.0cm. It was not tender and had no differential warmth. It was soft to firm and pedunculated. The mass did not transilluminate and both nostrils were patent (Figure A).

MRI done revealed no intracranial extension and other ancillary investigations done were within normal limit. The working diagnosis was Nasal Teratoma.

Subsequently, she had elective surgery in which the tumour was excised along with a rim of nasal bone under general anaesthesia. The polypoid shaped specimen measuring about 3cm x2.5cm was sent for histology. The columella was lengthened with V-Y advancement flap and the left nostril was repositioned to lie in anatomical position. Estimated blood loss was about 46mls. (Figure B).



Figure A



Figure B

Histology revealed sheets and islands of neuroglial tissue with the overlying normal epidermis and superficial dermis and an assessment of Extranasal glial heterotopias was made.

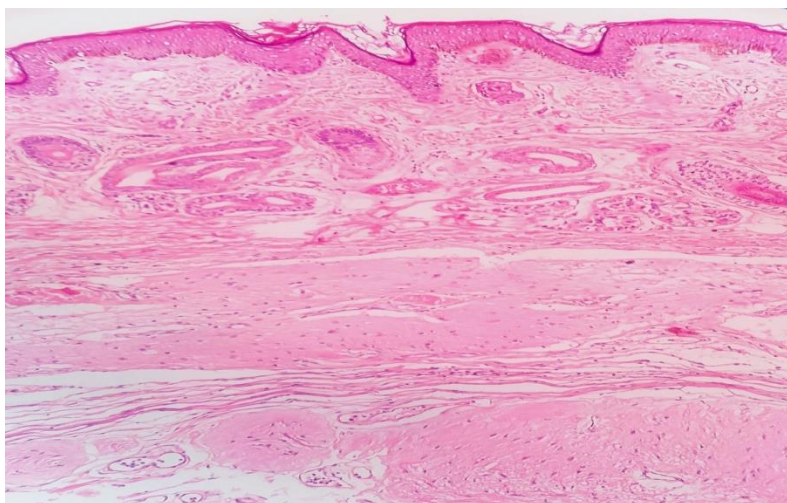


Figure 1. Histology reveals skin tissue with the sub epithelium composed of sheets and island of neuroglial fibres in a background of fibrocollagenous tissue

Discussion

Nasal glial heterotopias are rare congenital benign masses of neurogenic origin with intranasal or extra nasal locations or both [1]. It is found at birth and only rarely in adults with an incidence of 1 in 20,000 to 40,000 births [1]. It is generally found at birth and only rarely found in adults. The male to female sex distribution is 3:1. There is no known familial or hereditary predisposition and there is no malignant potential [5].

The differential diagnosis of congenital nasal masses is nasal dermal sinus cyst, nasal encephalocele and nasal glioma. These congenital lesions share similar embryonic origin which results from the failure of neuroectodermal and ectodermal tissues to separate during the development of the nose. The nasal glial heterotopia is also called nasal glioma. They are not true neoplasm. They originate from ectopic glial tissue left extracranially following abnormal closure of the nasal and frontal bone during embryonic development [1]. Thus, many authors recommended using the term glial heterotopia instead [6]. They usually present with intranasal or extra nasal mass with or without symptoms.

About 60% of glioma are extra nasal, 30% are intranasal and 10% are mixed [3]. Intranasal glial heterotopia usually presents with nasal obstruction or septal deviation. The extra nasal glial heterotopias are firm, incompressible round to globular often along the naso-maxillary suture. The overlying skin may have telangiectasia and may be confused with haemangioma. There are about 15% to 20% nasal glial heterotopias with a fibrous stalk connecting them to the subarachnoid space of the brain [1].

Histologically, nasal glial heterotopia is composed of astrocytes and neuroglial cells in a background of fibrovascular tissue. The glial nature of the cells is confirmed with immunohistochemical markers consisting of S100 and GFAP. Proper management of nasal glial heterotopia requires a multidisciplinary approach including a facial plastic surgeon, otorhinolaryngologist, radiologist, neurosurgeon and pathologist [7].

A complete medical history, full physical examination and any other associated congenital abnormalities should be ascertained. Nasal endoscopy should be done to delineate the site and extent of any intranasal mass. The intranasal connection must be excluded using CT scan or MRI due to risk of meningitis or cerebrospinal leakage [7]. The preferred form of imaging is MRI [7].

MRI imaging is better for delineating soft tissue details and hence excludes possible intracranial connection. It also minimizes the level of ionising radiation exposure especially in infant [5].

Surgical excision is the mainstay of treatment and the excised specimen should be sent for histopathological diagnosis. The extent of the surgery is dictated by the exact size, location and content of the lesion. Early surgical intervention is strongly advised because of possible complications such as cosmetic problems, brain abscess and meningitis [7].

Conclusion

Nasal glial heterotopias are non neoplastic, rare congenital lesion with possible intracranial involvement and complications. Appropriate clinical evaluation, cross-sectional imaging such as MRI or CT scan of the brain must be done to exclude an intracranial connection before surgical procedure. Conservative surgical excision is the mainstay of the management and the specimen should be submitted for histopathological diagnosis.

Conflict of interest: I have no financial conflict of interest.

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