Isolated nasal septal neuroglial heterotopia – A case report

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A B S T R A C T

Glial heterotopia is a rare condition in which mature neuroglial tissue is found in the body other than central nervous system (CNS). It manifests as a mass of extra-cranial cerebral tissue unconnected with the brain. Clinically, these masses are firm and incompressible. Histopathologically, they consist of neuroglial cells and astrocytes embedded in fibrous and vascular connective tissue. Radiological investigations such as computed tomography (CT) or magnetic resonance imaging (MRI) should be done to rule out intracranial extension. We report one case of nasal glial heterotopia in a 3-month-baby which has an unusual attachment over the nasal septum. The mass was completely resected endoscopically which was attached to the anterior part of the nasal septum.

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Introduction

Nasal glial heterotopia is also known as nasal glioma. These are rare, benign congenital nasal masses more accurately referred to as sequestered glial tissue. Schmidt was the first to present a comprehensive description of this lesion and coined the term glioma in 1900 [1]. According to Bradley et al. nasal glioma is reported to account for five of 109 congenital nasal masses [2]. Nasal glial heterotopia is frequently diagnosed in newborn infants, but rarely found in adults. It is habitually diagnosed right after birth and may be detected in the neonatal period in 60% of the cases [3]. Nasal glial heterotopia may appear at the root of the nose. These are one of the congenital midline masses, a category which also include dermoids and encephaloceles. An encephalocele is a protrusion of brain substance connected to the rest of the brain by a pedicle with associated osseous defect whereas nasal glial heterotopia has no communication with the subarachnoid space or the central nervous system (CNS) [4]. They may deform the bones of the nasal fossa and extend through the nasal bones, cribriform plate or the foramen caecum. Calcifications are rarely seen. Cystic changes inside the nasal glioma are occasionally seen. They may protrude out of the nostril and rarely attach to the nasal septum. Our case was showing attachment of glioma to the anterior part of nasal septum which is an extremely rare ectopic site inside the nasal cavity.
A 3-month-old girl baby was referred to outpatient department of otorhinolaryngology for right nasal mass. The mass had been present since birth. The baby was born at full term and had a normal vaginal delivery. Her elder sibling had no congenital abnormalities and the family history was unremarkable.

On examination, the baby had no facial and nasal swelling except a polyp like mass blocking the right nasal cavity which was protruding from the right nostril (Fig. 1). It was pale in colour and firm in consistency. It was neither tender, non-pulsatile, non-compressible nor with negative Furstenberg sign. On probing test with a swab stick, mass was attached to the anterior part of the nasal septum. There was no history of cerebrospinal fluid (CSF) leak or meningitis. Computed tomography (CT) (Fig. 2) and magnetic resonance imaging (MRI) (Fig. 3) of the brain showed a right side intranasal mass with no direct connection to the brain and mass showing attachment to the septum. The lesion was completely resected endoscopically. The nose was packed with merocele nasal pack which was removed after 72 h. The post-operative period was uneventful.

Histopathological examination of the nasal mass confirmed the diagnosis of nasal glioma. It consisted of mature glial tissue, astrocytes with fibrous components (Fig. 4). The immunohistochemistry revealed that S-100 and glial fibrillary acidic protein (GFAP) were positive. At 6 months follow-up the patient was doing well with no evidence of residual or recurrence in diagnostic nasal endoscopic examination.

Discussion

The incidence of congenital nasal masses is 1 in every 20 000-40 000 live births [5]. The differential diagnosis of
Fig. 4 – HPE showing (a) glial tissue in fibrovascular stroma, (b) glial elements in Masson’s trichrome, (c) phosphotungstic acid haematoxylin stain and (d) glial fibrillary acid protein stain

congenital nasal masses includes dermoid, hemangioma, glioma and encephalocele. Nasal glioma and encephaloceles are the results of abnormal embryologic development. During second gestational month, a diverticulum of the dura extends through the pre-nasal space and comes in contact with the superficial ectoderm in the area that will become the nose. As the naso-frontal area develops, the nasal processes of the frontal bone grow and surround the dural projection and form the foramen caecum. Under normal circumstances, the dural projection involutes into a fibrous structure, which fills the foramen caecum. Nasal glioma results from aberration of this normal developmental process. Nasal glial heterotopia is the congenital mass composed of mature brain tissue isolated from the cranial cavity or spinal canal. It is a rare condition, thought to be derived from either entrapped neuroectodermal tissue during the closure of the covering of the brain or a nasal encephalocele which is covered by dura, pia and arachnoid and later disconnected from the intracranial cavity during subsequent development. Nasal glioma occurs sporadically with no familial tendency or sex predilection [6]. The symptoms seen are nonspecific for nasal cavity mass: nasal obstruction, nasal discharge and chronic otitis media. These lesions usually present as a red or bluish mass at or along the naso-maxillary groove, or as an intranasal mass. They are characteristically firm, non-compressible, do not increase in size with crying or coughing and do not transilluminate. They may be associated with a widened nose or with hypertelorism, secondary to growth of the mass. Imaging studies are needed before excision of this mass to differentiate from encephalocele. An encephalocele is a protrusion of brain substance connected to the rest of the brain by a pedicle with associated osseous defect whereas nasal glial heterotopia has no communication with subarachnoid space or the central nervous system [7]. Biopsy or aspiration of these nasal masses is contraindicated because of the risk of meningitis or injury of functional brain tissue within an encephalocele [8]. Nasal gliomas are made of neuroglial
elements consisting of glial cells in a connective tissue matrix with or without a fibrous connection to the dura. Usually they consist of fibrillary neuroglial tissue with a prominent network of glial fibres, gemistocytic type astrocytes in background of neuropil, representing classic neuroglial tissue. It may be arranged in a lobular pattern and cystic structures may be present as well [9]. There is no fluid filled space connected to the subarachnoid space. About 90% of the reported nasal gliomas do not contain neurons, explained by either inadequate supply of oxygen to provide support the neurons or failure of the neurons to differentiate from the embryonic neuroectoderm in an intranasal glioma or both [10]. The presence of abundant neurons raises the possibility of an encephalocele. The glial nature of the cells can be confirmed by immunohistochemical demonstration of S-100 protein and GFAP. These two proteins can demonstrate neuroglial cells with high specificity, and help to distinguish nasal gliomas from other tumours such as meningiomas and granular cell tumours [11]. In our patient, both S-100 and GFAP were positive. Intranasal glioma most commonly arises from the lateral nasal wall near the middle turbinate [12] whereas our case showing attachment to the anterior part of the nasal septum which is extremely rare.

The diagnosis of heterotopia versus encephalocele needs the knowledge of radiologic and operative findings of the patients [13]. CT scan and conventional radiographic studies have limitations in detecting small bony defects or narrow stalk like connections across the skull base. MRI is considered as the investigation of choice for evaluation of congenital nasal masses, especially those having intracranial connection [14]. Nasal endoscopy should be done in all cases to get precise location, origin and pulsatility [15]. The preferred treatment of nasal glioma is complete surgical excision, although recurrences after surgery rarely occur. Intranasal endoscopic approach is strongly recommended for the removal of intranasal gliomas [16]. With suspected bony defects, an intranasal endoscopic approach is considered as the procedure of choice [17]. Early surgical intervention is essential in view of the potential risk of cerebrospinal fluid leak and meningitis. Delaying of the treatment may cause distortion of the septum and nasal bone or infection in the nasal cavity.

Conclusion

Nasal glial heterotopia is rare, benign and a congenital lesion. Ectopic presentation of glial tissue over the nasal septum is extremely rare as in our case. Evaluation should involve mandatory pre-operative imaging with a thin cut coronal and axial CT scan and/or multi-planar MRI to exclude any intracranial connection before doing an invasive procedure. Treatment requires endoscopic excision in case of intra-nasal glioma. Conservative endoscopic excision is done as it is slow growing, rarely recurrent and has no malignant potential. It is very important to close follow-up of these patients because of the possibility of postoperative CSF rhinorrhea and infection.

