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A Rare Case of Glial Choristoma Arising from the Vomer in Association with Cleft Palate

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Abstract

Heterotopic neuroglial tissue represents normal glial tissue in an abnormal location distant from the central nervous system. It is a rare congenital condition and the majority of these lesions are diagnosed at birth or early childhood. We report a rare case scenario of a growth arising from the vomer associated with cleft palate. The origin of a glial choristoma from the midline of the nasal cavity in association with a cleft palate has not been reported in the literature. Complete surgical excision was performed prior to palatoplasty with no postoperative complications or evidence of recurrence.

Keywords

glial choristoma, cleft palate, vomer

Introduction

The term "Choristoma" refers to a cohesive tumor-like mass consisting of normal cells in an abnormal location.[1] Heterotopic brain tissue, a rare entity, first described by Reid in 1852 has <30 cases being reported in the English literature so far.[2] A variety of terms have been used to describe the lesion. The term glial choristoma is more accurate as it better reflects the lesion's pathologic nature. Glial choristoma is a developmental malformation of heterotopic central nervous system tissue, which is an uncommon developmental abnormality typically presenting at birth or in early childhood.[3,4] The most frequently affected is the nasal region. Palate, tongue, cheek, scalp, and orbit can also be affected but the occurrences are relatively rare.[4] It can be associated with other craniofacial anomalies including cleft palate, micrognathia, glossoptosis, polydactyly, pectus excavatum, bronchopulmonary dysplasia, and cardiovascular anomalies. Although these lesions are rare, they should be included in the differential diagnosis of congenital masses present in early childhood in the oral and maxillofacial region.[3]

This case report will discuss a rare presentation of glial choristoma arising from the vomer associated with cleft of secondary palate and management of the same.

Case Report

A 1-year-old male child was brought to our unit by his mother with a chief complaint of a defect in the child's roof of the mouth with growth since birth. A detailed history revealed that the growth was seen to be mobile when the child cried with no history increase in the size of the growth. No history of nasal regurgitation or recurrent ear, nose, and throat infection associated with cleft palate. Medical history revealed full-term vaginal delivery with no history of neonatal intensive care unit admission. There was no family history of similar complaints.

A general physical examination revealed that the patient was healthy with normal development of milestones and neurologically stable. No abnormalities in the fingers and toes were detected. An extraoral examination revealed a retrognathic mandible with no significant gross facial asymmetry. An intraoral examination exhibited the presence of a cleft of hard and soft palate with bifid uvula and a well-defined growth of 3.0×1.5 cm oval in shape arising from the cleft defect (Figure 1), further evaluation of the swelling was not possible since the child was not cooperative. Since the parents were reluctant to get further investigations done due to financial problems, surgery was planned and performed.

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Intraoperatively a well-defined pedunculated growth of 3.0 × 1.5 cm oval in shape with irregular surface was seen arising from the vomerine bone. Stay suture was placed at the base and the growth was excised (Figure 2) and sent for a histopathological investigation followed by surgical correction of cleft of secondary palate was done using Veau Wardill Kilner V-Y pushback palatoplasty. Postoperative recovery was uneventful and the patient was under intensive care post-surgery.

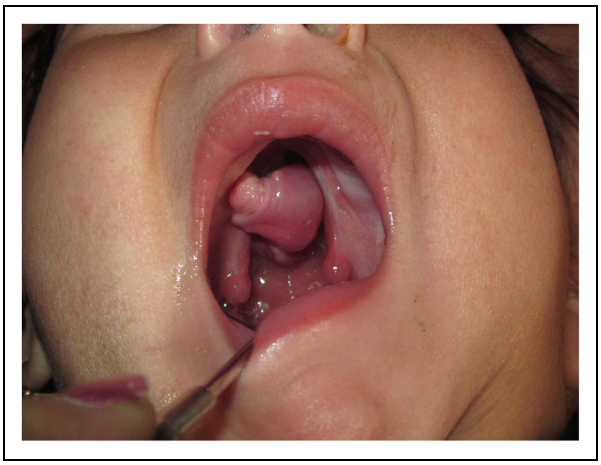

Figure 1. Preoperative clinical presentation.

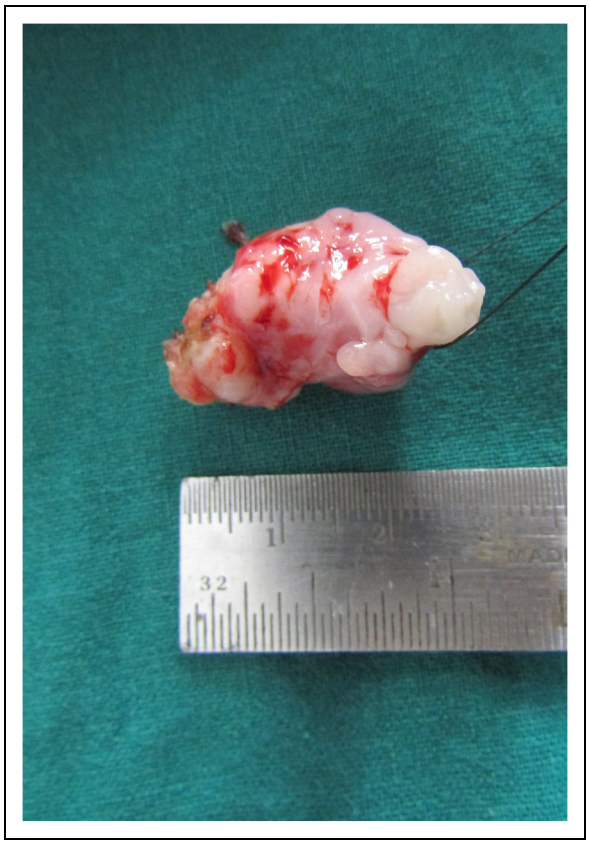

Figure 2. Intraoperative-excised specimen.

Follow-up visits showed wound healing to be satisfactory with no signs of dehiscence or recurrence (Figure 3).

Histologically mature glial tissue scattered in a background of fibrovascular stroma was noted. The overlying epithelium is parakeratinized and is of varying thickness (Figure 4). The glial tissue consists of a delicate eosinophilic fibrillary network consisting of astrocytes, which are stellate shaped (Figure 5) in a basophilic background, other areas are seen in the form of clumps or small islands with cystic lumen formation. The cells are round with vesicular nuclei and eosinophilic cytoplasm. Mature bony trabeculae, lobules of adipose tissue, minor salivary gland, and blood vessels are also evident. Aggregates of chronic inflammatory cells are seen in the stroma and within a minor salivary gland.

Discussion

Cleft of primary and/or secondary palate is a common congenital anomaly, incidence being ~1 in 700 live births.[5] The real etiology is still unknown, but environmental and genetic factors are involved in the pathogenesis. These patients suffer from development defects, maxillofacial shape secondary changes, surgical trauma, language, hearing, and other dysfunction; which decreases a patient's quality of life and brings consequential psychological trauma for both the patients and parents. Surgical intervention is necessary to guarantee adequate growth and functional development.[6]

Neuroglial choristomas (NGC) are very rare malformations, also called heterotopic brain, defined as a mass-like lesion composed of mature brain tissue isolated from the cranial cavity or spinal cord.[2] Some authors have divided NGC into lesions that are paraneuroaxial and those remote from the neuroaxis. In some literature, this malformation has also been referred to as glial choristoma, which has been used in our case to describe the condition. The embryological origin of heterotopic neuroglial tissue is not clear and numerous theories to explain their occurrence have been proposed but the general agreement is that they are developmental rather than neoplastic. Berblinger

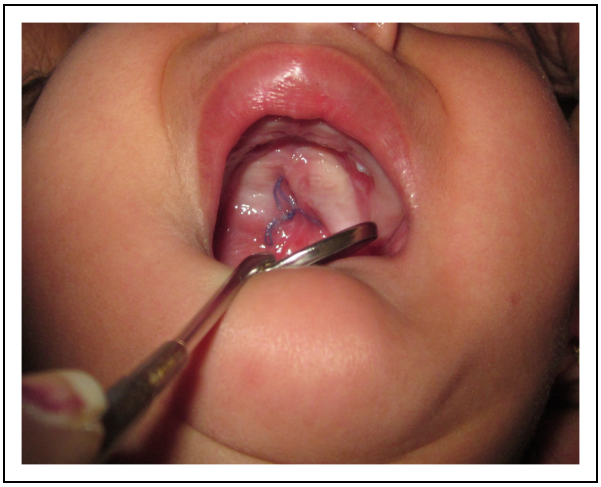

Figure 3. Postoperative-satisfactory wound healing.

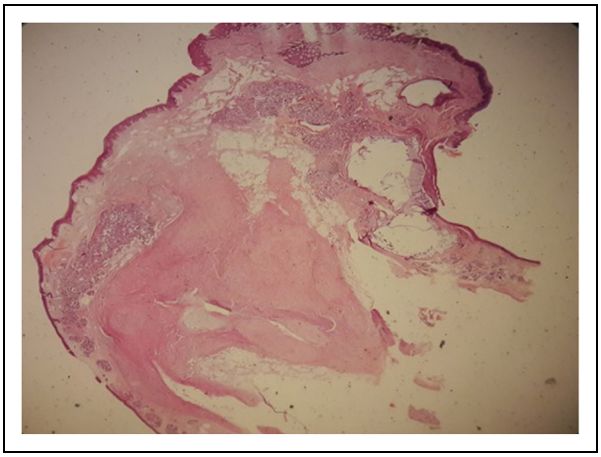

Figure 4. Hematoxylin and eosin stain (1.25×) of the lesion.

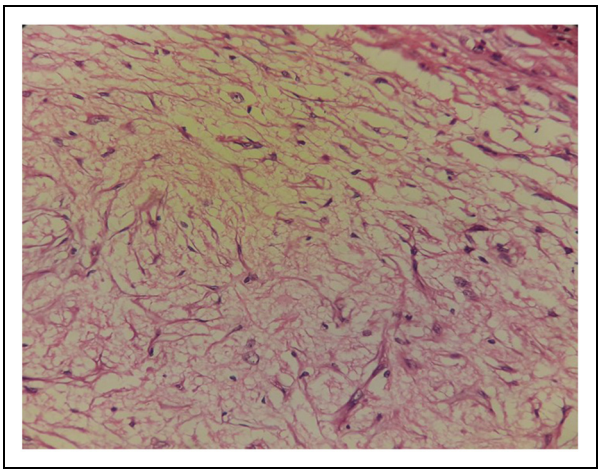

Figure 5. Hematoxylin and eosin higher-power view showing the lesion(40×).

postulated that this arose from tissue with only limited ability to differentiate cut off in early intrauterine life; he was the first to stress the blastomatous nature of the tumors and to apply to them the term "choristoma" in the sense of a blastomatous growth of embryonally separated primitive tissue.[7] The pathogenesis of paraneuroaxial NGC, especially those in the nasal cavity, are believed to be herniations of neural tissue before complete closure or through arrested closure of the osseous cranium or overgrowth of the developing neural lobe that may prevent closure of the cranial opening.[4,8] However, the absence of meningeal cells and the failure to find the site of herniation led other authors to consider all NGC to have arisen from neural crest cells in the head and neck with the ability to undergo neuroglial development. They grow around or incorporate adjacent structures such as the muscles, salivary glands, and fat.[8] On the other hand, encephaloceles are a herniation of dysplastic neural structures through a skull base defect.[2]

NGC occurs most frequently in the nasal cavity (nasal glioma), oral cavity; especially the palatopharyngeal area (57%) and the tongue (37%), and less commonly in the orbit, scalp, and areas remote to the head and neck.[8] As per the literature, an association of glial choristoma arising from the vomer with an associated cleft palate has not been reported. Hence this case is rare and unusual and is of significant importance. Since these tumors are rare, they present a diagnostic and prognostic dilemma. The differential diagnosis should include tumors derived from one of the germ layers: ectodermal (dermoid and epidermoid cyst), mesodermal (hemangioma, lipoma, fibroosseous lesion), neural (neurofibroma, glioma, encephalocele), and teratoma with derivatives from multiple germ layers.[3]

An interesting fact is the high percentage of the oropharyngeal glial choristomas occurring concurrently with cleft palate. A review of 17 glial choristomas of the oropharynx found that up to 6 cases were accompanied by cleft palate. Thus, it was inferred that the presence of heterotopic brain tissue in this area could have disrupted the normal fusion of palatal shelves. The presence of cleft palate, therefore, might be the result rather than the cause of ectopic glial tissue.[9] The same has not been reported in the literature for choristomas arising from vomer or nasal cavity, while it can be speculated to have the same theory as it hinders the fusion of palatal shelves due to the growth present in the region.

Previous reports documenting severe complications caused by glial choristomas of the palatopharyngeal area were not uncommon. The most predominant presenting symptoms were dysphagia and dyspnea. Mechanical obstruction of the airway in newborns may be life threatening.[9] However, in the present case, no such complication was noted.

Histologically, the lesion is composed of mature central nervous system tissue elements with a wide variation in the amount of connective tissue. The constituents include glial cells and sometimes ependymal cells, choroid plexus, and cell elements derived from neuroectodermal structures are noted. A proliferation of the fibrous connective tissue, which is believed to represent the degenerated glial tissue, is frequently observed around the nests of glial cells.[4] Most reported brain heterotopias including nasal gliomas mainly consist of mature neuroglial elements and true nerve cells are rarely seen.[3]

In the present case, the glial tissue was composed of a mass of delicate eosinophilic fibrillary network consisting of astrocytes, which are stellate shaped in a basophilic background. The glial cells are round with vesicular nuclei and eosinophilic cytoplasm. Mature bony trabeculae, lobules of adipose tissue, minor salivary gland, and blood vessels are also evident. The occurrence of mucous gland, adipose tissue, striated muscle, and bone has been reported in oronasopharyngeal cases. Neuroglial tissue is positive for glial fibrillar acidic protein, S-100 protein, vimentin, and CD57. Vimentin is the most primitive, intermediate filament-related protein found in astrocytes and glia while CD57 was initially raised against human natural killer cells but subsequent studies demonstrated specificity to neural tissue and neurogenic tumor.[10]

No syndromic predisposition or etiological factors have been identified in these patients although patients may have other craniofacial anomalies including cleft palate, micrognathia which was relatable with the present case.[2]

An adequate preoperative workup after physical examination is important especially to determine whether the mass has an intracranial connection because meningitis is a serious complication if this is not recognized and necessary precautions are not taken. In general, most of nonnasal gliomas do not have a central nervous system connection. A computed tomography (CT) scan is necessary to determine the location and extent of mass and to rule out any bony erosion or skull base defect. If a CT scan excludes any intracranial extent, excisional biopsy or incisional biopsy should be performed.[3] In our case, however, a CT scan could not be done due to the unwillingness of the parents and financial constraints.

In conclusion, the present case of glial choristoma originating in the midline from the vomerine bone associated with a cleft palate is a rare instance. The management of which is complete surgical resection and correction of the cleft palate with attention to the integrity of vital structures and the lesion must be distinguished from encephaloceles and tumors especially since such entities may have intracranial attachment.

Declaration of Conflicting Interests

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