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RARE CASE OF NASAL GLIAL HETEROTOPIA- CASE REPORT

Pathology

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ABSTRACT

Nasal glial heterotopia or nasal glioma is a rare congenital anomaly presenting as midline masses primarily in infancy or early childhood. Here we present a rare case of nasal glioma in a 9th month old child presented as midline swelling near root of nose. Clinical impression was dermoid cyst. The mass was excised. Histopathological examination showed glial tissue,thus making the diagnosis of nasal glioma.

KEYWORDS

nasal gliomas,Dermoid, encephalocele

INTRODUCTION

Nasal glioma is a rare anomaly presenting as congenital midline masses usually detected at birth. Its incidence is 1 in every 20,000 to 40,000 births[1].Glial heterotopias has male gender predilection. Nasal glioma has no familial tendency. It presents mostly as external nasal mass in 60% of cases. It can be located as an internal nasal mass (30%), mixed lesion (10%)[2].Most of the intranasal lesions arise from the wall of middle turbinate or rarely from septum and presents as pale masses. [2]. Glioma may extend into the orbit, frontal sinus, oral cavity or nasopharynx in rare cases [3].A few nasal gliomas(15%-20%) have a fibrous stalk connecting to the central nervous system [4].

Nasal dermal sinus cysts, nasal encephaloceles, and nasal gliomas are the most common congenital nasal masses. These masses have a similar embryogenic origin. When the neuroectodermal and ectodermal tissues fail to separate during the development of the nose,these usually occur [5].Lips, tongue, scalp, nasopharynx and oropharynx are rare locations for heterotopic brain tissue[6].

These masses have the potential for connection to the central nervous system so they become clinically important, although their incidence is low. Biopsy of the lesion with an intracranial connection can lead to complications such as meningitis or cerebrospinal fluid leak.Surgical excision is the treatment of the choice.

CASE STUDY

A 9 months old female baby presented with a swelling near root of the nose more towards right side. On examination, the swelling was measuring approximately 3X2 cm. It was non tender, firm to hard and non pulsatile. The other hematological and biochemical investigations were within normal limits. She had no other complaints. There was no relevant family history. A diagnosis of nasal dermoid was made clinically.

NCCT scan showed well defined soft tissue lesion near inner canthus of right eye with defect in underlying right nasal bone and no intracranial extension. Surgical excision was done under local anaesthesia and material sent for histopathological examination.

After surgical excision of mass gross examination showed a skin covered soft tissue piece measuring 1.5x1x1cm.Cut surface is greywhite.Histopathological examination of the nasal mass shows stratified squamous epithelium lined tissue.Subepithelium shows large fibrillary tissue arranged in nests and small sheets(Figure1&2)

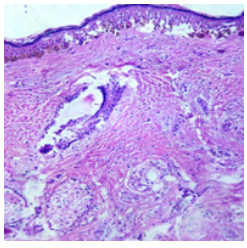

Figure 1-Photomicrograph showing epidermis with underlying glial tissue and skin appendages,H&E(100X)

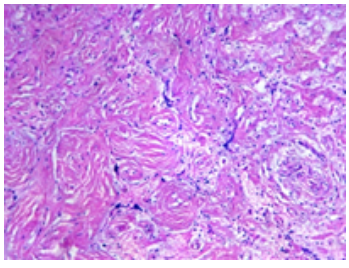

Figure 2- Photomicrograph showing sheets of fibrillary glial tissue, H&E, 400X

The cells were stained positive for glial fibrillary acidic protein (GFAP) and S-100(Figure 3&4)

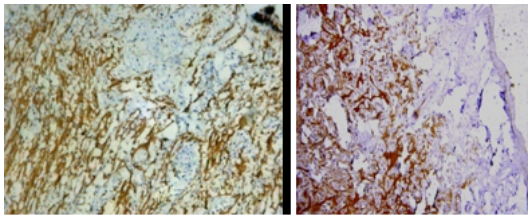

Figure 3&4- Photomicrograph showing diffuse glial fibrillary acidic protein and S100 positivity in glial tissue respevtively, 400X

DISCUSSION

The term nasal glioma is a misnomer because such a mass is not a true neoplasm. It is actually made up of ectopic nerve tissue that contains neuroglial elements, with glial cells in a connective tissue matrix with or without connection to the subarachnoid space or dura [7]. The male-to-female ratio is 3:2. Approximately 150 cases have been reported and no familial predisposition has been described [8]. Some cases of nasal glioma associated with other malformations, such as agenesis of the corpus callosum and cleft palate, have been reported [9,10]. Only in 15% of the cases, nasal gliomas remain connected with the intracranial structures by, a pedicle of glial tissue, usually through a defect in the cribriform plate [7,10]

It is frequently diagnosed in newborns or infants as in our case although it is rarely found in adults. The usual age of presentation of the nasal gliomas is in infancy or early childhood as in this case the girl was 9months old. Clinically, these masses are soft, pale, and polypoid. They can protrude through the nostrils and mimic a nasal polyp. In our case swelling was present near inner canthus of right eye externally which is most common site of nasal glioma. Nasal gliomas can cause remodelling and deformities of the adjacent bones and commonly cause hypertelorism as in our case there was defect in underlying right nasal bone. Obstruction of the nasal passage and nasolacrimal duct can lead to respiratory distress and epiphora on the affected side. Complications like CSF rhinorrhoea, meningitis or epistaxis can also develop in these patients [10,11,12]

Types of nasal gliomas:

Extra nasal (60%): lie external to the nasal bones and nasal cavity and most commonly occur slightly off the midline at the bridge of the nose

Intranasal (30%): found within the nasal or nasopharyngeal cavity, the oral cavity or rarely the pterygopalatine fossa.

Combined (10%): consists of a communication between the extranasal and intranasal components which occurs via a defect in the nasal bones or around the lateral edges of the nasal bones [13]

Our case is an extra nasal type of nasal glioma with underlying defect in nasal bone and no intracranial extension. Nasal glioma pathologically resemble reactive gliosis so they are classified as heterotopias and not as neoplasia. Histologically, nasal gliomas are unencapsulated nests of glial cells as seen in our case nests and sheets of glial tissue are seen. They usually contain large aggregates of astrocytes (fibrous or gemistocytic) and fibrous connective tissue enveloping the blood vessels. Multinucleated giant cells are often seen. No microscopic invasion, mitotic figures or metastases have been noted.[12] this was also true in our case. Gliomas do not contain neurons, because of [7,11,12] low levels of oxygen in the mass and the lack of differentiation from embryonic neuroectoderm.

One of the differential diagnosis of nasal glioma is nasal dermoid as in this case clinically it was diagnosed as dermoid but later on histopathological examination showed glial tissue thus making a diagnosis of nasal glioma. Nasal dermoids are thought to result from a lack of regression of a diverticulum of dura that extends through the foramen ceacum between the developing nasal cartilage and nasal bone. Depending on the patency of the diverticulum and its contents, the lesion can be a sinus, dermoid cyst, nasal glioma or encephalocele. Nasal dermoids may be sporadic or familial. The mean age of presentation is 3 yrs with no sexual predeliction. They may be found at any location between the glabella and the base of the columella [14]

Complete surgical excision of the mass is the definitive treatment. Entire mass must be removed in order to prevent recurrence. Intranasal lesions are approached via lateral rhinotomy or by endoscopic techniques.

CONCLUSION:

Nasal gliomas constitute one of the important midline nasal masses in infancy or early childhood and this case report emphasis on the need for its correct recognition. Nasal glioma can be a differential diagnosis for nasal dermoid as in this case. Since the prognosis of the patient for nasal dermoid and nasal glioma varies the correct diagnosis of this condition is necessary so that right treatment will be provided to the patient. At present, nasal glial heterotopia needs ENT, neurosurgery, radiology and pathology multidisciplinary combination approach.

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