Case Report:

Glial heterotopia presenting as nasal polyp - Report of two cases.

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Abstract

This report described two cases of nasal glial heterotopia which primarily presented as nasal polyp. Glial heterotopia one form of congenital midline nasal mass is a rare anomaly usually detected at birth. There are congenital malformation of displaced normal, mature glial tissue, which is no longer in continuity with an intracranial component. The case reports correlates clinical findings with immunohistochemical investigations.

Introduction: Nasal glial heterotopia is considered to be congenital malformation of displaced normal, mature glial tissue, which is no longer in continuity with an intracranial component[1.] This is distinctly different from an encephalocle, which is a herniation of brain tissue and/or leptomeninges, that develops through a defect in the skull, where there is continuity with the cranial cavity[1.]

Glial Heterotopia, one of the congenital midline nasal mass is a rare anomaly usually detected at birth. The reported incidence is 1 in every 20,000 to 40,000 births[1,2.] While nasal glial heterotopia (NGH) is the preferred term, synonyms have included nasal glioma[1,2.]

However, this term is to be discouraged, as it implies a neoplasm or tumor, which it is not. By definition, nasal glial heterotopia is a specific type of choristoma. It is not a teratoma, however, which is a neoplasm comprising all three germ cell layers (ectoderm,endoderm,mesoderm)[1,2.] As a congenital malformation or ectopia, it is distinctly different from the trauma or iatrogenic development of an encephalocele[1,2.]

The most common congenital nasal masses are nasal dermal sinus cysts, nasal encephaloceles, and nasal gliomas. These masses appear to share a similar embryogenic origin. They occur when the neuroectodermal and ectodermal tissues fail to separate during the development of the nose [2.]

They can be extranasal (60% of cases), intranasal (30%), or mixed (10%). Other rare locations for heterotopic brain tissue include the lips, tongue, scalp, nasopharynx and oropharynx[3.] Although rare, these disorders are clinically important because of their potential for connection to the central nervous system[3.]

Patients come to clinical attention early in life (usually at birth or within the first few months), with a firm subcutaneous nodule at bridge of nose, or as a polypoid mass within the nasal cavity, or somewhere along the upper border of the nasal bridge[3.] If the patient presents with an intranasal mass, there may be obstruction, chronic rhinosinusitis, or nasal drainage. If there is a concurrent cerebrospinal fluid (CSF) leak, then an encephalocele is much more likely[3.]

They are categorised into two types based on the anatomic site of presentation:Extranasal (60%) located in subcutaneous bridge of nose;Intranasal (30%) located in superior nasal cavity and mixed (10%) located in subcutaneous tissues and nasal cavity which are larger in size[1,2,3.]

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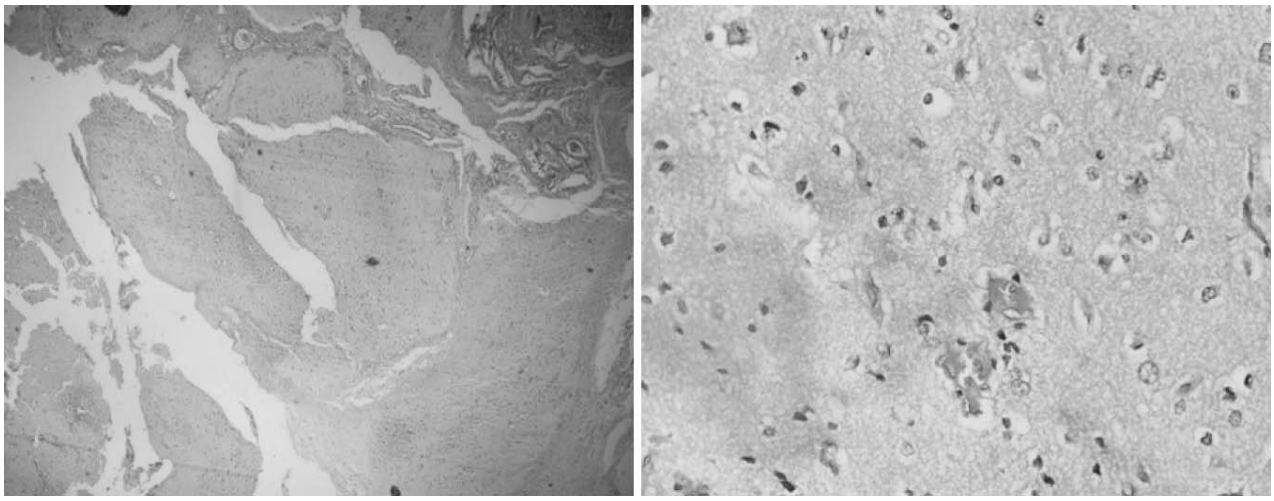

FIG.I, II (Case1): H&E stain showing glial cells,small vessels and inflammatory cells in fibrillary background.(100x & 400x).

Here, we are presenting two cases of nasal polyp which on microscopic examination and immunohistochemically diagnosed as Nasal Glioma.

Case Report:

A 17 year old boy (case1) and a 35 year old lady (case2) presented with history of right sided chronic nasal obstruction and intermittent nasal discharge. They had no other complaints. The family history and sibling history were unremarkable.

Clinically their general condition appeared normal. The right nasal cavity examination in both cases showed a pale white mass filling the right nasal cavity.

On indirect laryngoscopic examination the mass was seen attached to nasal septum which did not bleed on touch and was non-pulsatile. The left side of nose was normal. Rest of the ENT examination was within normal limit.

FIG. III : (Case 2) : H& E stain showing polypoid tissue lined by columnar epithelium and subepithelial tissue showing oedema and fibrillary glial tissue.(100x).

Haemogram and urine examination were done which were within normal limit. CT scan showed an intranasal mass not connected to intracranial cavity or bony defect in floor of anterior cranial fossa. Other examination findings were unremarkable.

Functional endoscopic sinus surgery was performed for the visualization of the nasal polyp and its extent. Polypectomy was done under general anaesthesia and postnasal pack applied.

After surgical excision gross examination of the masses revealed pale unencapsulated tissue, measuring 2 x 1.5 cm and 0.7 x 0.5 cm respectively. The cut surface was greyish white, glistening.

Histopathological examination of the nasal mass showed fibrocollagenous tissue with fibrillary neuroglial tissue with a prominence of glial fibres and few neural cells. Chronic inflammatory infiltrates comprising of lymphocytes and plasma cells with few congested blood vessels also seen (Fig. I, II, III).

Immuno-histochemical typing was performed and the cells were positive for S100 protein(Fig. IV, V). There were no mitotic figures or necrotic areas. At the time of this report after one year of surgery, patient is in good health and has shown no signs of recurrence till date.

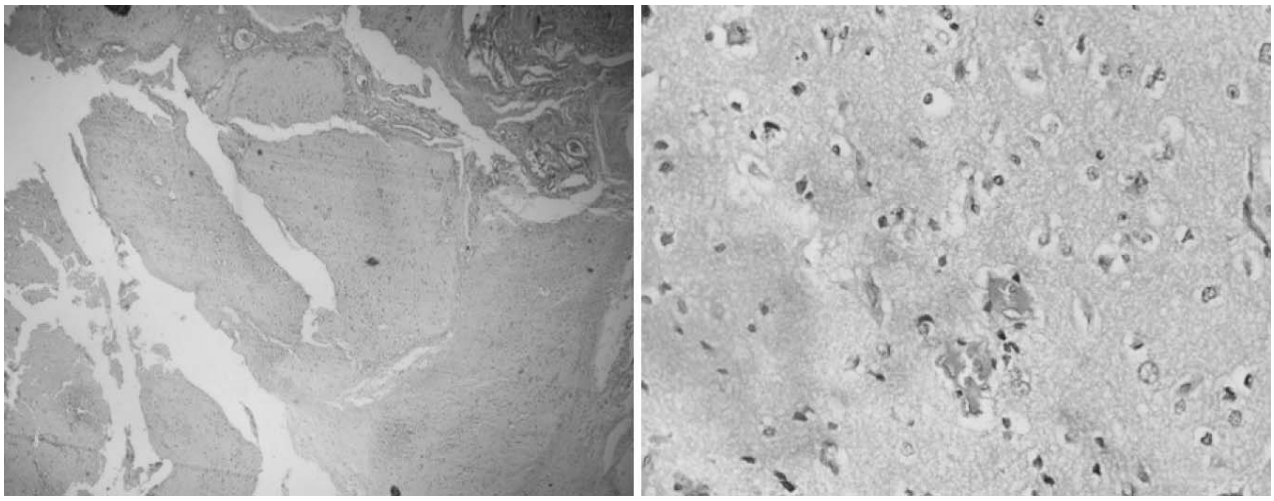

FIG. IV, V: Expression of S 100 protein in glial heterotopic tissue (Immunoperoxidase- antiperoxidase staining , 100x).

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 connective tissue heterotopias.Encephalocele and gliomas have similar embryological origin but encephalocele is due to herniation of cranial contents through a defect in skull; a glioma is thought to be an encephalocele which has lost intracranial connection. Neurons have been identified in matrix with or without connection to the subarachnoid space or dura [4.]

The male-to-female ratio is 3:24. Approximately 150 cases have been reported and no familial predisposition has been described[5.] Penner C.R and Thompson L observed 10 cases in a study period of thirty years ranging from1997-20006.Some cases of nasal glioma associated with other malformations, such as agenesis of the corpus callosum and cleft palate, have been reported [5,6,7.]

Only in 15% of the cases, nasal gliomas remain connected to the intracranial structures by a pedicle of glial tissue, usually through a defect in the cribriform plate . The usual age of presentation of the nasal gliomas is in infancy or early childhood[6,7.]

But in these cases the boy was asymptomatic till 14 years of age and even the lady did not show any remarkable symptoms till at a late age of 35 years. Clinically, these masses are soft, pale, and polypoid. They can protrude through the nostrils and mimic a nasal polyp[6,7.] Nasal gliomas can cause remodelling and deformities of the adjacent bones and commonly cause hypertelorism[7,8.]

Obstruction of the nasal passage and nasolacrimal duct can lead to respiratory distress and epiphora on the affected side[7,8.] Complications like CSF rhinorrhoea, meningitis or epistaxis can also develop in these patients [7,8,9.] Nasal gliomas are classified as heterotopias and not as neoplasia since pathologically they resemble reactive gliosis[8,9.]

They present as a firm, skin covered, reddish coloured, and non-pulsatile, usually slowly growing, polypoidal lesions. Histologically, nasal gliomas are unencapsulated nests of glial cells. 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 reported so far[8,9.]

Reactive changes and local calcifications as seen in some nasal gliomas may reflect the relatively poor blood supply to these heterotopias [9,10.] Neurons have been identified in 10%-60% of cases in the series reported. About 90 reported nasal gliomas do not contain neurons, because of low levels of oxygen in the mass and the lack of differentiation from embryonic neuroectoderm [9,10.]

Neuro-imaging is essential for the characterization of an intranasal glioma to determine its exact location and, more important, to exclude possible intracranial extension[6,9,10.] The definitive treatment is complete surgical excision. Entire mass must be removed in order to prevent recurrence[9,10,11.] Intranasal lesions are approached via lateral rhinotomy or by endoscopic techniques[6,10,11.]

Conclusion

Nasal gliomas constitute one of the important midline rare nasal masses and these case reports emphasise on the need for its correct recognition. Nasal glioma can be a differential diagnosis for nasal polyp as in these cases. Since the prognosis of the patient for nasal polyp and nasal glioma varies, the correct diagnosis of this condition is necessary so that right treatment can be provided to the patient.

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