Neonatal Nasal Glioma: A Case Report

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Citation

S Singhal, R Virk, A Dass, S Bansal, Amanjit. *Neonatal Nasal Glioma: A Case Report*. The Internet Journal of Otorhinolaryngology. 2005 Volume 4 Number 2.

Abstract

A congenital midline nasal mass is a rare anomaly usually detected at birth. The most common congenital nasal masses are nasal dermal sinus cysts, nasal encephaloceles, and nasal gliomas. Nasal glioma is a developmental abnormality of neurogenic origin. We report a case of a one day old neonate in which the nasal glioma was excised endoscopically.

INTRODUCTION

Congenital midline nasal masses are rare. The reported incidence is 1 in every 20,000 to 40,000 births[1]. The most common are dermoid cysts, teratoma, 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. Nasal glioma is a rare congenital malformation, first described by Reid in 1952, although the term "glioma" was coined by Schmidt in 1900[2]. The term nasal glioma is a misnomer because as such this mass is not a true neoplasm.

It is 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. The male-to-female ratio is 3:2, and about 250 cases have been reported so far[3, 4]. No common associations with other malformations or familial predisposition have been described. Treatment of glioma with intracranial extension is excision by craniotomy. Lateral rhinotomy approach used to be standard for the gliomas localized to the nasal cavity but off late more surgeons are using endoscopes for excision.

We describe a case of a one day old child in which endoscopic excision of the nasal glioma was done successfully.

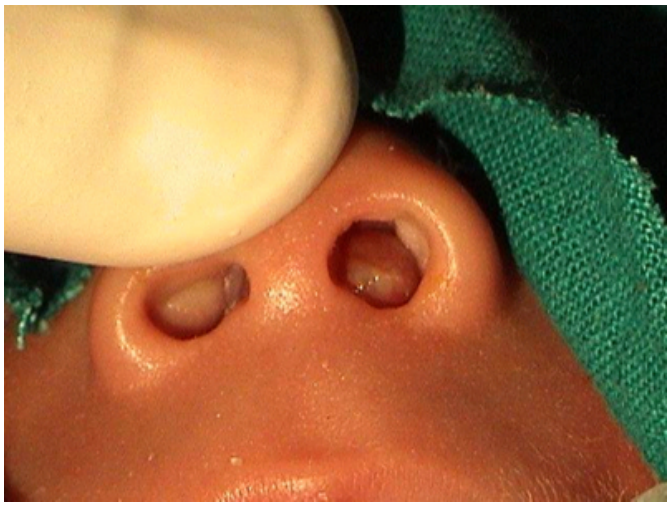
CASE REPORT

An urgent call was received from the labor room for a newly born male baby for respiratory difficulty and mass protruding from the left nostril. External examination of the nose was normal. Anterior rhinoscopy revealed a mass having a purple hue and occupying the whole of the left nasal cavity protruding through the left external nares like a polyp(Fig.1). It was soft, polypoidal, non pulsatile and was moving in and out with respiration. There was no change in size of the mass during crying or on jugular vein compression. The right nostril was patent. There were no other abnormalities.

Computerized Tomography (CT) revealed a well rounded soft tissue density mass attached to the lateral wall of the nasal cavity filling the anterior left nasal cavity. There was no intracranial extension or any other intracranial mass. All the routine hematological and biochemical investigations were normal.

Figure 1

Figure 1: Photograph of the patient showing the mass in the left nasal cavity.



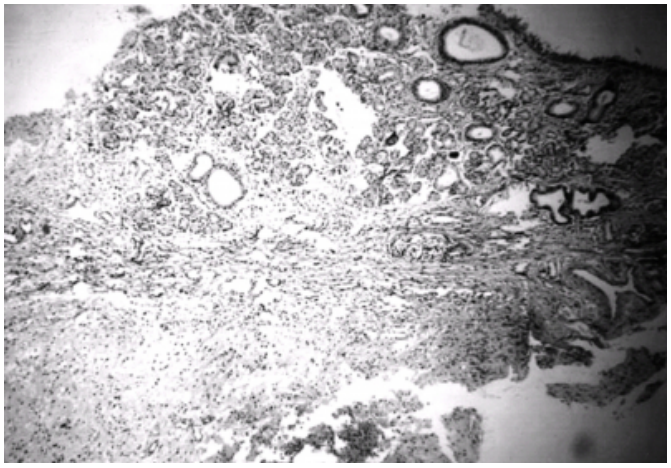
Excision of the mass was performed under endotracheal anaesthesia using a paediatric nasal endoscope. The mass was seen arising from the lateral wall of the nose anterior to

the attachment of the middle turbinate. It was removed in toto and the nasal cavity was packed with MEROCEL™ . The mass measured 1.5cm X 1cm X 0.5cm. The nasal pack was removed after 48 hours. There was no bleeding, no CSF leak, no fever and no sign of infection. The patient was discharged on the 4th post operative day.

The histopathological diagnosis was nasal glial heterotopia consistent with nasal glioma (Fig2). Endoscopic examination after one and three months revealed a normal nasal cavity with no evidence of tumor.

Figure 2

Figure 2: Photomicrograph showing nasal mucous membrane and mucous glands separated from glial tissue by fibrous septa. Stain: Hemotoxin & Eosin, Magnification: 40 X



DISCUSSION

Nasal gliomas usually arise during infancy or later childhood. 60% of cases are extranasal, lying external to the nasal bones and cavities, 30%are intranasal lying within the nasal cavity, mouth, or pterygopalatine fossa and 10% are mixed communicating through a defect of the nasal bones. Other rare locations for heterotopic brain tissue include the lips, tongue, scalp, nasopharynx, and oropharynx[5]. Our case was of an intranasal glioma.

The exact pathogenesis of nasal gliomas has not been positively established. However, the most widely held theory is that nasal glioma represents an encephalocele that becomes sequestrated from the brain and cranial vault early in gestation. Therefore, in most instances nasal glioma is not a true neoplasm. They are characterized by unencapsulated collections of astrocytic eosinophilic neuroglial cells in a connective tissue matrix[6].

An intranasal glioma is usually attached to the nasal septum or to the upper part of the middle turbinate. Clinically, these masses are soft, pale, and polypoidal. They can protrude through the nostrils and can be confused with a nasal polyp. Signs and symptoms include nasal obstruction, epistaxis, and cerebrospinal fluid (CSF) rhinorrhoea. They can be associated with deformities of the adjacent bones and obstruction of the nasolacrimal duct. A large mass can lead to hypertelorism, broadening of the nasal bridge, airway obstruction, and epiphora [7,8]. Although benign, they can cause significant local damage and cosmetic deformity by compressing and destroying the nasal cartilage. The diagnosis of this mass may pose a problem as Fine needle aspiration cytology (FNAC) or an excision biopsy carry a significant risk of meningitis[7].

The availability of CT scan and MRI facilitate the diagnosis of a glioma. CT can demonstrate bony defects, and MRI can demonstrate the characteristics of the soft-tissue mass and its possible intracranial connection. On CT, the mass is usually isodense to brain tissue. On MRI, the lesion is isointense to hypointense relative to gray matter on T1-weighted sequences and hyperintense on T2-weighted and proton density sequences[9]. However a final diagnosis can only be obtained after complete excision.

Surgical excision of the tumor is the treatment of choice. Inadequate primary excision results in a 4 -10% recurrence[4] . Intracranial extension of the mass can be shown by CT and MRI and by direct visualization of the origin of the mass during nasal endoscopy. Trans nasal endoscopic resection of the intranasal glioma must be accepted as the treatment of choice[10] . Endoscopic approach allows precise surgery with minimal trauma to the surrounding tissues. Moreover it has an aesthetic advantage eliminating a post operative surgical scar. However in unskilled hands, there may be a risk of incomplete excision. Post operative follow up can also be done easily with an endoscope. Intracranial nasal gliomas require a craniotomy to obtain a clear surgical margin.

Nasal gliomas should be considered in the differential diagnosis of a nasal mass in an infant. Transnasal endoscopic surgery should be considered as a procedure of choice for the removal of intranasal glioma without any intra cranial connection.

CONCLUSION

Congenital midline nasal masses rarely occur. Nasal gliomas arise in infancy or early childhood. If left untreated they can cause deformity of nasal bones and adjacent structures. Transnasal endoscopic surgery should be the treatment of

choice in nasal gliomas not having an intra cranial connection. Successful endoscopic excision in such a young patient prompted us to report this case.

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