

A Rare Case of Extra Nasal Glioma with Nasal Cleft

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Abstract: Congenital midline swellings of nose are encountered rarely, and nasal gliomas constitute about 5% of such lesions. Nasal gliomas are rare, benign, developmental abnormality of neurogenic origin, generally sporadic, with no familial tendency and are thought to be the result of an error in embryonic development. They are typically slow growing and very rarely associated with other congenital malformations. Extranasal gliomas are usually located at the glabella level, but may present laterally. High-resolution MRI is often useful in demonstrating intracranial stalk. Complete surgical excision of the tumor is the treatment of choice. Etiology of unilateral cleft nasal deformity is a lack of skeletal support of the cleft alar base. Early management of the nasal deformity minimizes nasal asymmetries and allows the nose to grow in a symmetric fashion. We present an infrequent case of nasal glioma associated with unilateral nasal cleft in a five month old child.

Keywords: Congenital Midline Masses, Nasal Glioma, Nasal Cleft

INTRODUCTION

Nasal gliomas are rare, benign, congenital lesions accurately referred to as 'ectopic sequestered glial tissue' also called nasal glial heterotopias arise from abnormal embryonic development with a potential for intracranial extension.

They were first described by Reid in 1852, although comprehensive description and coining of the term was by Schmidt in 1900 [1].

The reported incidence is 1 in every 20,000 to 40,000 births [2]. Nasal gliomas occur when the neuroectodermal and ectodermal tissues fail to separate during the development of the nose [1].

Notably these lesions can be foci of infection and is of paramount importance given the tendency for intracranial communication in these lesions, hence leaving the patient at risk for intracranial infections. It is prudent to have preoperative imaging with a thin-cut axial and coronal CT scan and multiplanar MRI. Biopsy of the lesion with an intracranial connection can lead to meningitis or cerebrospinal fluid leak and is contraindicated. Treatment is complete surgical excision.

Nasal clefting (medial or lateral) is a rare entity often associated with other congenital anomalies. We present an exceptional case of congenital extranasal nasal glioma with nasal cleft, association of which has been scarcely reported in the medical literature.

CASE REPORT

A five-month-old male presented to our out-patient with swelling at the root of the nose and nasal cleft on right side since birth. Parents gave history of the swelling initially being trilobed and later when child was two months old one lobe dried and fell off. The swelling has gradually increased in size since birth with no other complications. The only child was product of non-consanguineous marriage and born at full term by normal vaginal delivery. Mother had an uneventful pregnancy. Family history was unremarkable and child had attained all developmental milestones at appropriate age.

On general physical examination, baby was playful, interested in its surrounding and with no other abnormalities other than those mentioned earlier. Baby was pale. Systemic examination was normal. On examination, swelling was right paramedian in position, bilobed with short stalk at root of nose with larger lobe measuring 2.5x1.7x1 cm and smaller 2x1x0.8 cm. It was reddish blue, non-pulsatile with hairs present.

Consistency was firm and was non-compressible with skin not pinchable. There was slight bony indentation at the base. Transillumination was negative. No change in size of the mass during crying or on internal jugular vein compression (Furstenberg's test). No intranasal mass was noted and both nostrils were patent. Alar notching in the area of soft triangle on right side of the nose was evident with no clinically notable skeletal excess or deficiencies. Columella was short and broad and telecanthus was noted.

Ultrasound showed a well defined iso to hypoechoic subcutaneous mass with homogenous echo texture and no vascularity / calcification / hemorrhage within the mass and a diagnosis of dermoid cyst was made. CT scan revealed a large well defined isodense subcutaneous polypoidal mass measuring 3.4 x 1.6 cms in the mid line with right deviation at the orbitonasal junction. Axial CT showed an underlying nasal bone defect on the right side but coronal reconstructed CT image showed no facioskeletal bony defect with intact cribriform plate. In MRI, a polypoidal mass was seen as hyperintense on T2 and hypointense on T1. No intracranial or intranasal extension or other synchronous lesion was seen. Glial heterotopia was suggested.

Hemoglobin was 7.8 and was corrected with transfusion. With neurosurgeon on standby child was taken for surgery under general anesthesia. Horizontal skin incision was taken proximal to the normal and abnormal cutaneous junction, dissected up to the frontonasal junction where it ended abruptly and the bilobed swelling was removed in-toto and sent for histopathological examination. A bony indentation was evident but with no defect. Closure was done in layers. Post-operatively child had surgical site infection and was treated promptly. Child was particularly observed for postoperative bleeding, CSF leak. On gross examination of the specimen, the cut surface of the tissue showed grey-white, smooth appearance with cystic areas. Microscopy examination revealed skin covered mass with large islands of glial tissue with admixture of fibrovascular tissue and vascular spaces.

Also seen were numerous neurons with some showing axonal projections. The diagnosis of nasal glioma was hence established. Sutures were removed on seventh day. Cleft nose correction was contemplated at a later stage and the same was conveyed to the parents and were advised for a regular follow up. However they failed to show up.



Fig. 1: Extranasal swelling at the root of nose (right Paramedian) with right nasal cleft



Fig. 2: White arrow shows junction of normal and telangiectatic hairy skin and Black arrow pointing the cleft nose

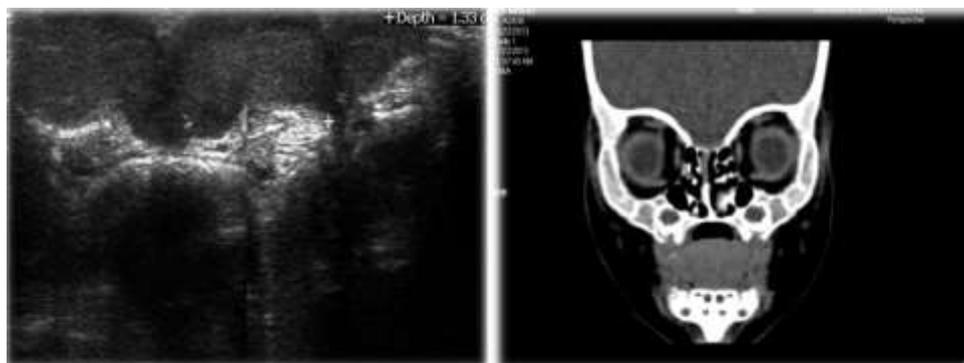


Fig. 3: USG on left showing well defined iso to hypoechoic subcutaneous mass with homogenous echotexture and axial CT on right showing a large well defined isodense subcutaneous polypoidal mass

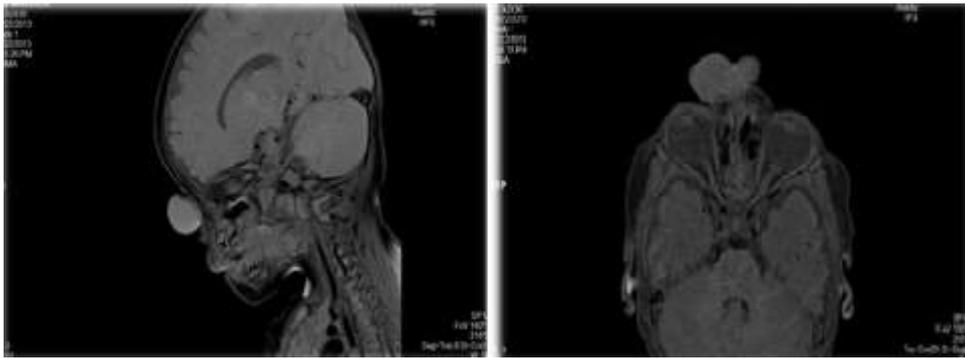


Fig. 4: On MRI polypoidal mass is seen as hyperintense on T2 and hypointense on T1 and does not show any intracerebral extension as there is no apparent tract or a cribriform plate defect



Fig. 5: Specimen sent for HPE was partially skin covered irregular tissue bit with two bulbous ends & on microscopy glial tissue in upper half and admixture of fibrovascular tissue in lower half is seen



Fig. 6: Post-Op day 3. Note the more evident telecanthus and broad nose after removal of nasal glioma



Fig. 7: Post-Op day 7. After suture removal. Oedema and erythema noted.

DISCUSSION

Nasal glioma is ectopic nerve tissue that contains neuroglial elements. The male-to-female ratio is 3:1 [3]. 60% of these gliomas are extranasal, 30% are intranasal and 10% are mixed, dumbbell shaped communicating through a defect of the nasal bones with a connecting band [4].

Only around 250 cases have been reported and no familial / hereditary predisposition or malignant potential has been described [5]. Some cases of nasal glioma associated with other malformations, such as agenesis of the corpus callosum and cleft palate, have been reported [5, 6].

The diagnosis is substantiated with high-resolution axial and coronal CT scan (bony anatomy) and thin section, high-resolution multiplanar MR imaging delineates soft tissue in detail and is requisite in determining possible intracranial extension. Nasal endoscopy performed to detect intranasal component is of vital importance. Along with histological findings of the specimen, the glial nature of the cells can be further confirmed by immunohistochemical demonstration of S100 protein and GFAP.

Although benign, they can cause significant local damage and cosmetic deformity by compressing and destroying the nasal cartilage with potential risk of cerebrospinal fluid leak or infection and secondary visual involvement. However since they have a slow growth rate (consistent with the patient's body growth), a comprehensive preoperative evaluation should be pursued and an appropriate surgical method be planned. Nasal glioma to be managed by a multidisciplinary approach is of paramount importance. Entire gliomatous tissue must be removed in order to prevent recurrence.

Extranasal gliomas with no obvious CNS connection may be excised via an external incision, using either a vertical elliptical midline incision or a horizontal incision over the dorsum of the nose ceding equally good aesthetic outcome [7]. A frontal craniotomy will be required in case of intracranial extension to prevent any postoperative CSF rhinorrhea or intracranial infectious complication (meningitis) or recurrence.

Our patient had both a nasal glioma and a cleft nose. Nasal clefts may range from a small groove to a large furrow extending all the way to the medial canthus. CT scanning is helpful in determining the bony anatomy.

For the cleft nose, timing for surgical intervention and its extent varies considerably. Current trend is towards early surgical correction. Hugh G.

Thomson has reported three cases of nasal glioma associated with ipsilateral naso-ocular cleft over duration of 28 years.

Nasal gliomas constitute one of the important midline nasal masses and this case report emphasis on the need for its correct recognition along with associated other congenital anomalies.

ACKNOWLEDGEMENT

We acknowledge Departments of Neurosurgery, Paediatric Surgery, Anaesthesia and Pathology, RRMC & H for the support.

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