CASE REPORT

Infantile Nasal Glial Heterotopia - A Case Report

Bharti Thaker, Subhash Bhardwaj

Abstract

Nasal Glial heterotopia also known as Nasal glioma is a rare, benign lesion due to defective embryonic development. It is considered to be congenital malformation of displaced normal mature, non teratomatous glial tissue without continuity with an intracranial component. The reported incidence is 1 in every 20,000 to 40,000 births. We present a case of 10-month-old infant presented to ENT department with a mass at the root of the nose since birth with gradual increase in size over months. NCCT head showed a soft tissue density mass measuring 1.8×1.8×0.2cm with no intracranial or intranasal extension. Complete surgical resection was done and specimen was sent for histopathological examination. Histopathological examination showed presence of unencapsulated mature glial tissue composed of astrocytes and fibrillary background. There were no inflammatory cells, fibrosis or calcification. Based on radiological and histopathological examination diagnosis of Nasal glioma was made. Patient had an uneventful postoperative period with no recurrence.

Keywords

Nose, Heterotopia, Glioma

Introduction

Nasal gliomas also known as nasal glial heterotopias are rare congenital lesions that arise from defective embryonic development often represent collections of normal glial tissue in an abnormal location distant to the central nervous system with no intracranial connectivity.[1] Schmidt was the first person to coin the term glioma in 1900.[2] Most common sites involved are in and around the nose and nasopharynx. In the nasal region it could be extranasal seen in 60% cases, intranasal (30%) and both (10%). Other rare locations for heterotopic brain tissue are lips, tongue, scalp and oropharynx. The incidence of nasal gliomas is one in 20000-40000 live births. It is seen more in females.[3] The other swellings that occur in midline and forms differential diagnosis with glial heterotopia include haemangiomas, gliomas, dermoid cysts and encephaloceles. Therefore, Radiological examination along with histopathological examination is important for reaching the definitive diagnosis.

Case report

A 10-month-old infant presented to ENT department with a mass at the root of the nose since birth with gradual increase in size over months. There was no history of nasal obstruction, difficulty in feeding, difficulty in breathing. The swelling didn't interfere with the vision of the infant and did not increase on crying. Infant had normal developmental milestones .

Routine haemogram, biochemistry tests and urine examination were unremarkable. NCCT of the head showed a soft tissue mass measuring 1.8×1.8×0.2cm without intracranial or intranasal extension. No bony abnormality was seen in nasal cavity, base of the skull and paranasal sinuses. Surgical excision was done and specimen was sent for histopathological examination.

Gross examination showed a globular unencapsulated soft tissue mass with unremarkable outer surface. Cut section showed a greyish white solid tissue with no areas of haemorrhage or necrosis. Tissue was processed and Haematoxylin eosin-stained smears were examined under microscope.

Histopathological examination showed presence of

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unencapsulated mature glial tissue composed of astrocytes and fibrillary background. There were no inflammatory cells, no fibrosis or calcification. There were no mitotic figures or necrotic areas.

Based on radiological and histopathological examination diagnosis of Nasal glioma was made. At the time of this report after 6 months of surgery patient is in good health without any signs of recurrence till date.

Discussion

The term Nasal gliomas is a misnomer as it is not a true neoplasm. These are locally aggressive lesions usually present at birth or diagnosed in early childhood.[4] When intranasal these may cause nasal obstruction resulting in respiratory distress and dyspnoea in neonates and infancy. Extranasal lesions present as mass lesion usually at the midline nose resulting in disfigurement as in our case. The swellings that occur in midline and share a similar

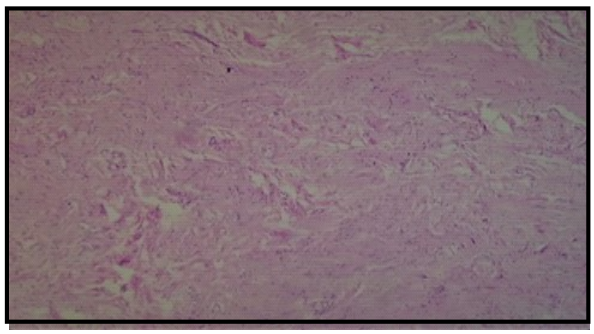

Fig 1. Photomicrograph showing glial tissue intermixed with fibrillary background in a case of nasal glioma. H&E(20x)

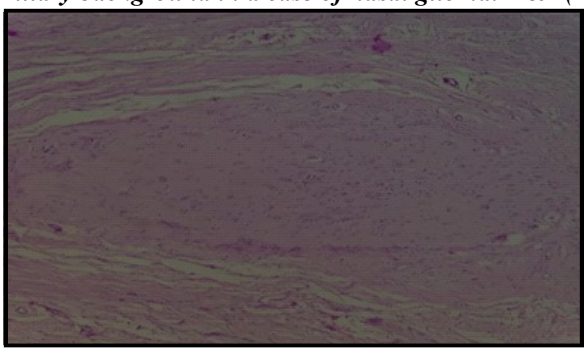

Fig 2. Photomicrograph showing presence of glial tissue (arrow) surrounded by fibrous tissue in a case of nasal glioma. H&E(40x)

embryogenic origin with that of nasal glioma include haemangiomas, gliomas, dermoid cysts and encephaloceles, cystic lymphangioma.[5] These occur due to the failure of separation of neuroectodermal and ectodermal tissues during the development of nose.[6] Radiological and histopathological examination are important in differentiating these entities.

Radiological evaluation with CT or MRI is important during both preoperative and postoperative phases. First imaging can show the exact location of the mass and would rule out continuity with the central nervous system. Nasal gliomas usually have no intracranial connection however in 15 -20% cases these have connection with dura. Treatment in case of nasal glioma with intracranial extension is excision followed by craniotomy. Postoperative radiological follow up is necessary as the recurrence rate due to inadequate primary excision has been reported as 4%-10%.[7] Invasive procedures like FNA or biopsy are not done so as to avoid cerebrospinal fluid leakage, removal of functional brain element as well as to avoid meningitis.[8] The treatment of NGH is complete surgical excision and is curative.[9] Our patient had no postoperative complication 6 months postoperatively and had no recurrence. Early diagnosis followed by complete excision prevents complications with better cosmetic results. Therefore, it is important to keep this entity in mind while diagnosing soft tissue swellings in and around nose especially in infancy.

Conclusion

Congenital midline nasal masses rarely occur. Glial heterotopia of head and neck are more common in and around nose. Early diagnosis and surgical resection is important to prevent deformity of nasal bones and adjacent structures.

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