Basrah Journal Of Surgery

Case Report

Bas J Surg, March, 15, 2009

NASAL GLIOMA, A CASE REPORT AND LITERATURE REVIEW

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Abstract

Nasal glioma (also known as nasal glial heterotopia) has been used to describe a congenital benign tumor of the nasal region containing neural tissue. It arises from failure of closure of foramen caecum at about the third week of gestation. The patient described in this report is an 18 months old girl who presented with a nasal bridge swelling for three months. The differential diagnosis included nasal encephalocele, nasal dermoid and epidermoid cysts. All are due to failure of ectoderm and neuroectoderm embryologic separation. CT scan and MRI imaging can be used to look for probable concomitant intracranial tumors and the existence of a connection between nasal tumor and the brain. Surgical resection is the usual method of managing such pathologies. In this case, an open rhinoplasty approach was used to resect this mass after the radiologic evaluation was complete.

Introduction

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^{1,2} with glial cells in a connective tissue matrix with or without connection to the subarachnoid space or dura³. It is a rare tumor of neurogenic origin. Embryologically, they are considered to be of similar etiology to an encephalocele without intracranial linkage. The incidence of congenital nasal masses has been reported to range from 1 in 20,000 to 1 in 40000 live births⁴. These congenital malforare classified nasal encephalocele, dermoid, and nasal glioma.

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Case Report

This is an 18 months old healthy girl who was born following a normal pregnancy and delivery with average birth weight. She was presented to our tertiary care centre by her parents who were concerned with what they claim as a sudden swelling on their daughter's nasal bridge that has been there for the past three months. The swelling was increasing in size with change in color. There was no significant past history of nasal trauma and no significant past medical or surgical history. On examination, a mass on the nasal bridge is found. It was 1.5x1.5x1 cm in size. There was no punctum and the skin was slightly reddish. The mass was firm and well circumscribed. It was non compressible and had a negative Fursten sign (do not enlarge with crying). The nasal cavity was clear by rigid endoscopy.

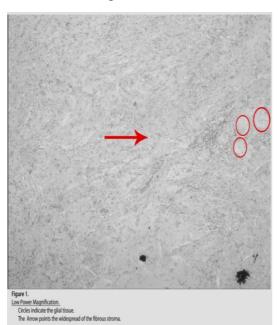
Cranial nerves were grossly intact, and the rest head and neck examination was normal.

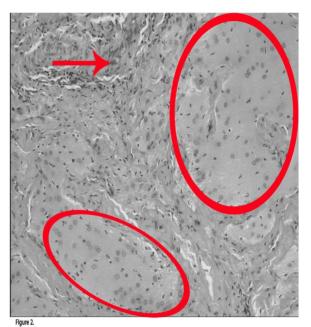
CT scan revealed that the mass had no connection to brain tissue, with no other abnormality detected.

The patient underwent an open rhinoplasty technique which involves an inverted V shaped transcolumellar incision. The skin was elevated of the medial crura and connected to marginal incision along the lateral crura. Skin-soft tissue envelope of the tip was elevated along with that of the nasal dorsum. A firm white nodule was excised as one piece with no difficulty, plus there was no evidence for any superior connection to the mass. Post-operatively the patient was free of any complications and she was discharged on the second post operative day.

The histopathology sections showed a nodular mass composed of fibrillary neuroglial tissue intersected with vascular connective tissue which is consistent of the diagnosis of nasal bridge glial heterotopia.

The patient was followed up for one year post operatively with no complications. She had a complete resolution of her initial signs





<u>High Power Magnification</u>

Circles are surrounding areas of glial tissue.

The arrow is pointing to the fibrous stroma sorrounding the glial tissue.

Discussion

Complete history and physical examination are the essential milestones for the evaluation and management of congenital midline nasal masses as they all may present as an intranasal or extranasal masses. The three most common aetiologies are dermoid cysts, gliomas, and encephaloceles.

Dermoids have an equal sex ratio, they can present with repeated infections or discharge through a visible sinus tract opening in the nasal skin. They are firm, non compressible, non pulsatile masses that do not transilumminate.

Gliomas are also firm in nature, noncompressible, non-transilumminable, with a normal or telangiectatic skin covering.

On the other hand, encephalocele may present as a bluish or a reddish mass. They are usually soft, compressible, and increase in size while crying.

Nasal glioma, is a rare entity, that has to be included as part of the differential diagnosis of a congenital mass at the level of the nasal dorsum. According to Manning et al, an initial biopsy might result in a rapid diagnosis but might also result in a cerebrospinal fistula⁵. Therefore, nasal masses should be evaluated first with radiological evaluation as an essential part of the pre-operative workup, to rule out any intracranial extension, where the surgical approach then should include a neurosurgeon if indicated⁴.

In general, open rhinoplasty is usually performed in patients older than 16 years old as such a procedure at a younger age may lead the nose to grow in an unpredictable manner⁶. An unbalanced nasal shape may result due to damage of the proportions

made during surgery by the nasal growth. Younger children may also not tolerate the emotional trauma of a cosmetic surgery. In this particular case, we believe that an open rhinoplasty was an ideal procedure because imaging studies did not show or detect an intracranial connection of the nasal mass together with the advantages provided by the open approach as the good exposure of the mass and the ability of doing a controlled surgical manipulations and better cosmesis^{7,8}.

The photomicrographic pictures by courtesy of Dr. Sawsan Bu Khamseen, a pathology registrar.

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