Nasopharyngeal Glioma: A unique presentation in infants

Nitin Anand • A.K. Rai

Abstract

Despite the complex embryological development of the nose and surrounding structures, significant developmental nasal anomalies are rare. Of the various anomalies like-nasopharyngeal cysts, hairy polyps, dermoids, haemangiomas, fibromas, mucocoeles, lipoma, aplasia. We are presenting a rare case of Heterotopic Brain Tissue in the nasopharynx. This 1.5-month-old patient was operated through transpalatal route and mass excised. Histopathologically it consisted of various central nervous tissue elements. Seven months post surgery patient is thriving well.

Keywords Glioma Nasopharynx infants

Introduction

Occurrence of extracranial brain tissue is rare. Of the recorded cases, most have been located anatomically within or adjacent to the nose and have been referred to as nasal glioma^(1,2,3,4,5). In cases where it maintains its continuity with intracranial contents, the term encephalocele is used.

They are divided according to the site of presentation as occipital, sincipital (fronto nasal) and basal. The most common are occipital lesions (75%). Next are sincipital masses that are external lesions apparent at birth as subcutaneous masses over nose, glabella, medial canthus or lower forehead.

Least common are basal lesions (10%), which present as intranasal masses. Of these basal lesions, nasopharynx is one of the rare sites affected^{6,7,8}.

Here we report another rare case of Heterotopic brain tissue in nasopharynx.

Case Report

A 1.5 months old male infant, weighing 2.41 kg, presented to ENT department with complaints of difficulty in swallowing, failure to thrive and a mass in oral cavity. Birth history was uneventful. On further enquiring parents gave history of gradual increase in size of oral mass and on-off difficulty in breathing.

On examination an ill-defined bulge of soft palate was seen obscuring most of the oropharynx (Photo 1). Child had noisy breathing and mild respiratory difficulty. On palpation mass behind soft palate appeared to be firm in consistency. There were no other external congenital anomalies found. Contrast enhanced computed tomography was advised. It revealed a soft tissue shadow in the nasopharynx pushing soft palate anteriorly. It extended superiorly from roof of nasopharynx till the tongue base inferiorly. There was no bony defect in the base of skull revealing any connection with intracranial contents (Photo 2).

Patient was taken up for excisional biopsy under general anesthesia. Patient was tracheostomized preoperatively to ensure adequate airway. It was seen firmly adherent to soft palate and roof of nasopharynx but there was no evidence of any anatomic connection with the base of brain.

Three weeks postoperatively the tracheostomy tube was removed. The child remains well seven months later with no evidence of any recurrence or any neurological deficit.

Pathology of the specimen

The specimen was a rounded polypoid structure with a core of homogenous grayish white tissue on cut section.

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Fig. 1 patient with bulge in soft palate.

Fig. 2 C.T. Scan showing mass in nasopharynx with intact base of skull.

Fig. 3 a&b Histological picture of the specimen showing astrocytes.

The tissue was fixed in 10% formalin embedded in paraffin and stained with hematoxylin and eosin.

Histological examination revealed cell clusters with fine fibrillar stroma. The cells were often star shaped and on appropriate staining were confirmed to be astrocytes (photo -3A&B). The cells had same characteristics as reported by Niels L. Low in 1956⁵ -a finely granular, faintly eosinophilic poorly outlined cytoplasm and large epithelioid nuclei with fine chromatin network. Cells showed no mitoses. Bands of dense collagenous fibrous tissue were seen dividing the neuroglial cells into clusters. The cleft like spaces were seen occasionally with the lining of cuboidal or low columnar cells, which were taken to be as ependymal cells.

Discussion

Considering the embryological aspects of the gliomas we have three theories in literature.

(1) They are due to faulty closure of the anterior neuropore⁹. At the third week of gestation period the ectodermal anterior neuropore develops which fuses before the base of skull does. A space between the frontal and nasal bones, known as fonticulus frontalis, completely fuses in the area of the future cribriform plate with the foramen caecum, separating intracranial contents from the extracranial structures. Failure of closure in this area may result in herniation of intracranial structures producing encephalocele. With faulty closure of the anterior neuropore, there may be no herniation but development of extracranial rests of CNS tissue, which forms gliomas. These may remain attached to dura with a fibrous stalk.

(2) Another theory suggest that they are of blastomatous origin due to extracranial separation of embryonal neuroglia¹⁰.

(3) They arise from neuroglial cells in the nasal mucosa and grow through the fronto-ethmoidal suture¹¹.

The gliomas commonly present shortly after birth and rarely in adult life¹². They are firm, incompressible and non-pulsatile with no intracranial connection of mass on CT scanning. However at operation a fibrous connection may be seen in 20 percent of cases¹³. Histologically Sussenguth⁹ mentioned that no typical astrocytes were found in the lesions, which are in contrary to other reports^{14,15}. In our case the cells in the lesion resembled astrocytes but no true nerve cells were recognized. Absence of invasive pattern and lack of mitosis rules out the possibility of any glial neoplasm. Though Lee et al⁽¹⁶⁾ described a case of 6 weeks old infant in which oropharyngeal melanotic ectodermal tumor coexisted with heterotopic brain tissue.

In 1984, V.Kasantikul found that only 10 cases had been reported in literature presenting as a symptomatic oropharyngeal heterotopic brain tissue¹⁷. A significant number of patients had associated cleft palate through which nasal / nasopharyngeal masses protruded into the oropharynx causing symptoms of difficulty swallowing and difficulty breathing^{5,17} but in our case there was no such association. Though mild respiratory difficulty was present, the tracheostomy done was more as a elective procedure rather than emergency. In review of literature it has been suggested that it is better to go for tracheotomy if respiratory obstruction

is severe as most of the patients survive following surgical resection of this benign obstructing lesion.

Conclusion

Many cases have been reported in literature regarding extracranial presence of brain tissue. Nasopharynx is one of the rare sites among forehead, dorsum nose, intranasal, palate and oropharynx, where this brain tissue can be present. Here we report another case of heterotopic brain tissue in nasopharynx. A six weeks old infant presented with bulge in oral cavity. On CT scan a mass was seen in nasopharynx extending to oropharynx. Mass was excised and on histopathological examination brain tissue with astrocytes, ependymal cells and some neurons were seen. Infant after seven months of operation is thriving well.

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