10.5005/jp-journals-10013-1227

CASE REPORT

Glial Heterotopia over the Nasal Septum: A Case Report and Review of the Literature

[1]Santosh Kumar Swain, [2]Satyabrata Dash, [3]Manash Ranjan Baisakh, [4]Rankanidhi Samal

ABSTRACT

Nasal gliomas are rare congenital lesions arising from defective embryonic development, often termed as Nasal glial heterotopias. It manifests as a mass of extra-cranial cerebral tissue unconnected with the brain. Clinically, these masses are firm and incompressible. Histopathologically, they consist of neuroglial cells and astrocytes embedded in fibrous and vascular connective tissue. Radiological investigations, such as computed tomography (CT) or magnetic resonance imaging (MRI) should be done to rule out intracranial extension. The mass was completely resected endoscopically which was attached to the anterior part of the nasal septum. Endoscopic technique provides excellent visualization. We report one case of nasal glial heterotopia in a baby from southern part of Odisha which has an unusual attachment over the septum. A review of the literatures regarding nasal glioma is also presented.

Keywords: Encephalocele, Glial heterotopias, Nasal glioma, Nasal septum.

How to cite this article: Swain SK, Dash S, Baisakh MR, Samal R. Glial Heterotopia over the Nasal Septum: A Case Report and Review of the Literature. Clin Rhinol An Int J 2015;8(1):34-37.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Nasal glial heterotopia is also known as nasal glioma. These are rare, benign congenital nasal masses more accurately referred to as sequestered glial tissue. Schmidt was the first to present a comprehensive description of this lesion and coined the term glioma in 1900.[1] According to Bradley et al, nasal glioma is reported to account for five of 109 congenital nasal masses.[2] Nasal glial heterotopias is frequently diagnosed in newborn infants, but rarely found in adults. It is habitually diagnosed right after birth may be detected in the neonatal period in 60% cases.[3] Nasal glial heterotopias may be extranasal or intranasal. Extranasal gliomas usually appear at the root of the nose.[4] These are one of the congenital midline masses, a category which also include nasal dermoids and encephaloceles. An encephalocele is a protrusion of brain substance connected to the rest of the brain by a pedicle with associated osseous defect, whereas nasal glial heterotopias has no communication with the subarachnoid space or the central nervous system (CNS).[5,6] They may deform the bones of the nasal fossa and extend through the nasal bones, cribriform plate or the foramen cecum. Calcifications are rarely seen. Cystic changes inside the nasal glioma occasionally seen. They may protrude out of the nostril and rarely attach to the nasal septum. Our case shows attachment of glioma to the anterior part of nasal septum which is a rare ectopic site inside the nasal cavity.

[1]Associate Professor, [2]Assistant Professor, [3]Consultant [4]Professor

[1,2,4]Department of ENT, Institute of Medical Sciences and SUM Hospital, Bhubaneswar, Odisha, India

[3]Department of Pathology, Apollo Hospital, Bhubaneswar Odisha, India

Corresponding Author: Santosh Kumar Swain, Associate Professor, Department of ENT, Institute of Medical Sciences and SUM Hospital, Bhubaneswar, Odisha, India, Phone: 06742384166, e-mail: swainsantoshbbsr@yahoo.com

CASE REPORT

A 3-month-old baby girl was referred to ENT outpatient department (OPD) for right nasal mass. The mass had been present since birth. The baby was born at full-term and had a normal vaginal delivery. Her elder sibling had no congenital abnormalities and the family history was unremarkable.

On examination, the baby had no facial and nasal swelling except a polyp like mass blocking the right nasal cavity which was protruding from the right nostril (Fig. 1). It was pale in color and firm in consistency. It was

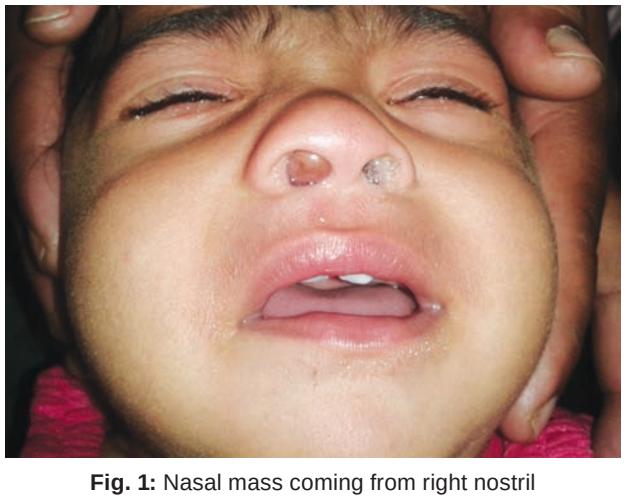


Fig. 1: Nasal mass coming from right nostril

neither tender, nonpulsatile, noncompressible and with negative Frustenberg sign. On probing test with swab stick, mass was attached to the anterior part of the nasal septum. There was no history of cerebrospinal fluid leak or meningitis. Computed tomography (CT) (Fig. 2) and magnetic resonance imaging (MRI) (Fig. 3) of the brain showed a right side intranasal mass with no direct connection to the brain and mass showing attachment to the septum. The lesion was completely resected endoscopically. The nose was packed with merocele which was removed after 72 hours. The postoperative period was uneventful.

Histopathological examination of the nasal mass confirmed the diagnosis of nasal glioma. It consisted of mature glial tissue, astrocytes with fibrous components (Figs 4A to D). At 6 months follow-up, the patient was doing well with no evidence of residual or recurrence in diagnostic nasal endoscopic examination.

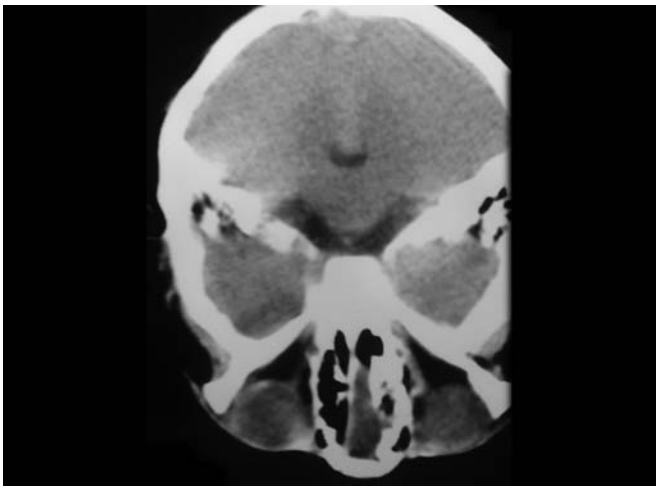


Fig. 2: Computed tomography scan of nasal mass

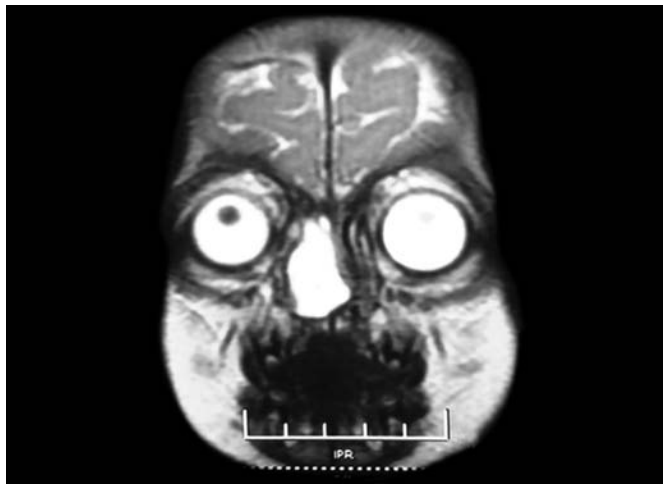
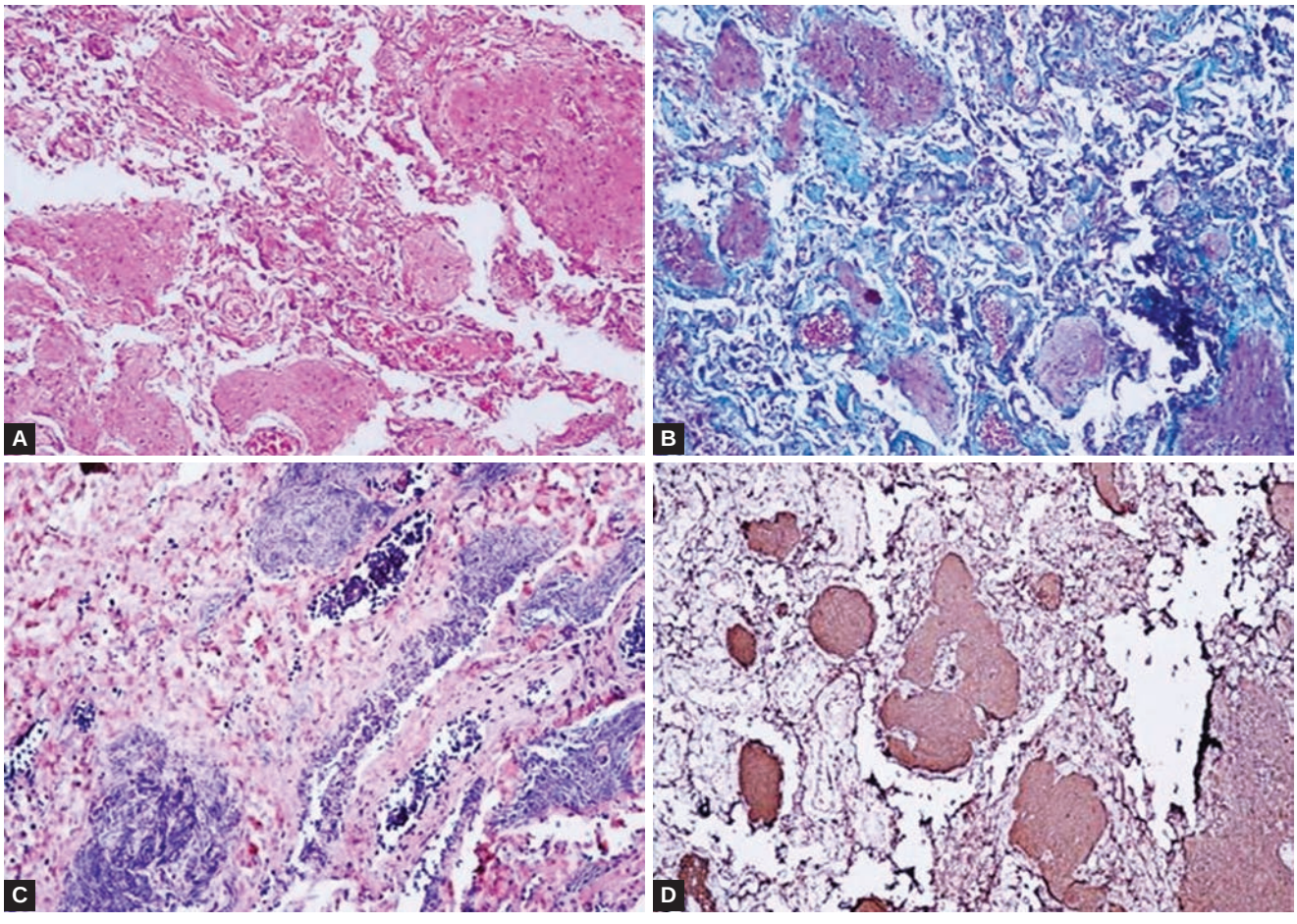


Fig. 3: Magnetic resonance imaging of brain showing no connection to nasal mass



Figs 4A to D: Histopathological examination: (A) glial tissue in fibrovascular stroma, (B) glial elements in Masson's trichrome, (C) phosphotungstic acid hematoxylin stain and (D) glio fibrillary acid protein stain

DISCUSSION

The incidence of congenital nasal masses is one in every 20,000 to 40,000 live births.[7] The differential diagnosis of congenital nasal masses include dermoid, hemangioma, glioma and encephalocele. Nasal glioma and encephaloceles are the results of abnormal embryologic development. During second gestational month, a diverticulum of the dura extends through the pre-nasal space and comes in contact with the superficial ectoderm in the area that will become the nose. As the nasofrontal area develops, the nasal processes of the frontal bone grow and surround the dural projection and form the foramen caecum.[8] Under normal circumstances, the dural projection involutes into a fibrous structure, which fills the foramen cecum. Nasal glioma results from aberration of this normal developmental process. Nasal glial heterotopia is the congenital mass composed of mature brain tissue isolated from the cranial cavity or spinal canal.[9] It is a rare condition, thought to be derived from either entrapped neuroectodermal tissue during the closure of the covering of the brain or a nasal encephalocele which is covered by dura, pia and arachnoid and later disconnected from the intracranial cavity during subsequent development.[10,11] Nasal glioma occur sporadically with no familial tendency or sex predilection.[12] The symptoms seen are nonspecific for nasal cavity mass: nasal obstruction, nasal discharge and chronic otitis media. These lesions usually present as a red or bluish mass at or along the nasomaxillary groove, or as an intranasal mass. They are characteristically firm, noncompressible, do not increase in size with crying or coughing and do not transilluminate. They may be associated with a widened nose or with hypertelorism, secondary to growth of the mass. Imaging studies are needed before excision of this mass to differentiate from enecephalocele.[13] An encephalocele is a protrusion of brain substance connected to the rest of the brain by a pedicle with associated osseous defect, whereas nasal glial heterotopias has no communication with subarachnoid space or the central nervous system.[14] Biopsy or aspiration of these nasal masses is contraindicated because of the risk of meningitis or injury of functional brain tissue within an encephalocele.[15] Nasal gliomas are made of neuroglial elements consisting of glial cells in a connective tissue matrix with or without a fibrous connection to the dura. Usually they consist of fibrillary neroglial tissue with a prominent network of glial fibers, gemistocytic type astrocytes in background of neuropil, representing classic neuroglial tissue. It may be arranged in a lobular pattern and cystic structures may be present as well.[16] There is no fluid filled space connected to the subarachnoid space. About 90% of the reported nasal gliomas do not contain neurons, explained by either inadequate supply of oxygen to provide support the neurons or failure of the neurons to differentiate from the embryonic neuroectoderm in an intranasal glioma or both.[17,18] The presence of abundant neurons raises the possibility of an encephalocele. The glial nature of the cells can be confirmed by immunohistochemical demonstration of S-100 protein and glial fibrillary acidic protein (GFAP). These two proteins can demonstarate neuroglial cells with high specificity, and help to distinguish nasal gliomas from other tumors, such as meningiomas and granular cell tumors.[19,20] In our patient, both S-100 and GFAP were positive. Intranasal glioma most commonly arises from the lateral nasal wall near the middle turbinate[21] whereas our case showing attachment to the anterior part of the nasal septum.

Computed tomography and conventional radiographic studies have limitations in detecting small bony defects or narrow stalk like connections across the skull base.[22] Magnetic resonance imaging is considered as the investigation of choice for evaluation of congenital nasal masses, especially those having intracranial connection.[23] Nasal endoscopy should be done in all cases to get precise location, origin and pulsatility.[24] The preferred treatment of nasal glioma is complete surgical excision, although recurrences after surgery rarely occur. Intranasal endoscopic approach is strongly recommended for the removal of intranasal gliomas.[25] In cases with suspected bony defects, an intranasal endoscopic approach is considered the procedure of choice.[26] Early surgical intervention is essential in view of the potential risk of cerebrospinal fluid leak and meningitis. Delaying of the treatment may cause distortion of the septum and nasal bone or infection in the nasal cavity.

CONCLUSION

Nasal glial heterotopias are rare, benign and a congenital lesion. More rarely, ectopic presentation of glial tissue over the nasal septum is presented in this case. Evalution should involve mandatory preoperative imaging with a thin cut coronal and axial CT scan and/or multiplanar MRI to exclude any intracranial connection before doing an invasive procedure. Treatment requires endoscopic excision in case of intranasal glioma. Conservative endoscopic excision is done as it is slow growing, rarely recurrent and has no malignant potential. It is very important to close follow-up of these patients because of the possibility of postoperative CSF rhinorrhea and infection.

REFERENCES

1. Schmidt MB. Ubner seltene Spaltbildungen in Bereiche des mittleren Stirnforsatzes. Virchows Arch Pathol Anat 1900; 162:340-370.

2. Bradely PJ, Singh SP. Congenital nasal masses diagnosis and management. Clin Otolaryngol 1982;7:87-97.
3. Haafiz AB, Sharma R, Faillace WJ. Congenital midline naso-frontal mass. Clin Paediatr 1995;34(9):482-486.
4. Hengerer AS, Newberg JA. Paed otolaryngology. 2nd ed. Saunders; 1990. p. 718-728.
5. Yeoh GP, Bale PM, de Silva M. So-called nasal glioma: case report with immunohistochemical study. Tumori 1998;84: 398-402.
6. Goodwin RE. Intranasal heterotopic brain. Ear Nose Throat J 1978;57:538-542.
7. Highes GB, Sharpino G, Hunt W, Tucker HM. Management of the congenital midline nasal masses: a review. Head Neck Surg 1980;2:222-233.
8. Sessions RB. Nasal dermal sinuses: new concepts and explanations. Laryngoscope 1982;92:7-28.
9. Chan JK, Lau WH. Nasal astrocytoma or nasal glial heterotopias. Arch Pathol Lab Med 1989;113:943-945.
10. Tashiro Y, Sueishi K, Nakao K. Nasal glioma: An immunohistochemical and ultrastructural study. Pathol Int 1995;45: 393-398.
11. Yeoh GP, Bale PM, de Silva M. Nasal cerebral heterotopias: the so-called nasal glioma or sequestered encephalocele and its variants. Pediatr Pathol 1989;9:531-549.
12. Gorenstein A, Kern EB, Facer GW, Lows ER Jr. Nasal gliomas. Arch Otolaryngol 1980;106:536-540.
13. Barkovich AJ, Vandermarck P, Edwards MS, et al. Congenital nasal masses: CT and MR imaging features in 16 casees. Am J Neuroradiol 1991;12:105-116.
14. Penner CR, Thompson LDR. Nasal glial heterotopias: a clinicopathologic and immunophenotypic analysis of 10 cases with a review of the literature. Annals of Diagnostic Pathology 2003;7(6):354-359.
15. Kardon DE. Nasal glial heterotopias. Arch Pathol Lab Med 2000;124:1849.
16. Abdel-Rahman N, AI Awadi Y, Ali Ramadan A, et al. Cerebral heterotopias of the temperofacial region: case report. J Neurosurg 1999;90:770-772.
17. Harley EH. Pediatric congenital nasal masses. Ear Nose Throat J 1991;70:28-32.
18. Smith KR Jr, Schwartz HG, Luse SA. Nasal gliomas: a report of five cases with electron microscopy of one. J Neurosurg 1963;20:968-982.
19. Uzunlar AK, Osma U, Yilmaz F, Topcu I. Nasal glioma: report of two cases. Turk J Med Sci 2001;31:87-90.
20. Kindblom LG, Angervall L, Haglid K. An immunohistochemical analysis of S-100 protein and glial fibrillary acidic protein in nasal glioma. Acta Pathol Microbiol Immunol Scand 1984; 92:387-389.
21. Yokoyama M, Inouye N, Mizuno F. Endoscopic management of nasal glioma in infancy. Int J Pediatr Otorhinolaryngol 1999;51:51-54.
22. Hughes GB, Sharpino G, Hunt W, Tucker HM. Management of the congenital midline nasal mass: a review. Head Neck Surg 1980;2:222-233.
23. Barkovich AJ, Vadermarck P, Edwards MS, Cogen PH. Congenital nasal masses: CT and MR imaging features in 16 cases. Am J Neuroradiol 1991;12:105-116.
24. Rahbar R, Resto VA, Robson CD, et al. Nasal glioma and encephalocele: diagnosis and management. Laryngoscope 2003;113:2069-2077.
25. Agyrdyr BV, Derin AT, Ozbilim G, Ozeaglar HU. Endoscopic management of the intranasal gliom. Turkey J Pediatric Surgery 2004;39(10):1571-1573.
26. Wu CL, Taso LY, Yang AD, Chen MK. Endoscopic surgery for nasal glioma mimicking encephalocele. Skull Base 2008; 18(6):401-404.