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RARE CASE REPORT OF NASAL GLIOMA

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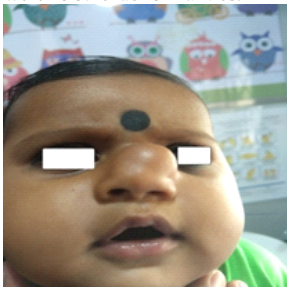
ABSTRACT Congenital midline swellings of nose are encountered rarely, and nasal gliomas are nonmalignant rests of neuroglial tissue that typically present as a craniofacial mass constitute about 5% of such lesions. Various theories have been suggested to explain the pathogenesis. Imaging preferably by MRI is mandated to study the extent and to rule out intracranial extension. Early surgical excision of these masses is advised to minimize nasal and craniofacial distortion. and the approach depends upon the extent of the lesion and availability of various surgical subspecialties, including otolaryngology, neurosurgery, plastic surgery, and ophthalmology. We present the management of one such case of congenital both intranasal and extranasal glioma without any intracranial extension.

KEYWORDS :

1. CASE PRESENTATION

A 5-month-old female baby was brought by parents with complaint of a swelling over the nose on left side since birth. History of occasional nasal obstruction, and the mass was growing slowly. There was no history of epistaxis or any other nasal discharge. The baby was born to a nonconsanguineous parents and born at fullterm by a normal vaginal delivery. The family history was unremarkable. On general physical examination, the baby was playful and hemodynamically stable. On anterior rhinoscopy mass seen over the left nostril and fleshy mass over the dorsum of nose.No change in size of the mass was observed during crying or on jugular vein compression (Furstenberg's test). The right nostril was patent.on palpation mass was 2x3cm in size ,firm in consistency. There were no other abnormalities.



The parents were counseled, and MRI scan was advised to study the nature and extent of the mass. mri scan revealed irregular, lobulatd, hetrogenous altered signal intensity lesion measuring approx 20x12x15 mm in soft tisuue of left side of nose ,extending posteriorly into left nasal cavity ,which is hetrogenously hypertense onT2W,STIR sequences and isointense onT1W sequence fig[1,2,3].No bony defect or intracranial extension or other synchronous lesion was seen. under general anesthesia nasal mass excised . There were no perioperative complications, and the patient was particularly observed for postoperative bleeding, CSF leak, fever or other features of infection. Histopathological analysis of the specimen revealed nonmalignant gliomatous cells with low proliferative activity. No meningeal or dural tissue was identified. The diagnosis of nasal glial hetrothrophia was hence established. The patient remained under followup for five months and did not show any evidence of recurrence of lesion.

Fig 1

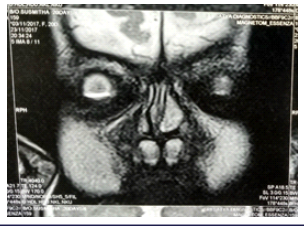


fig 2

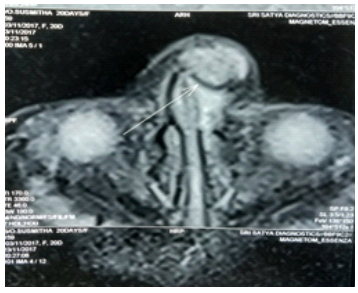
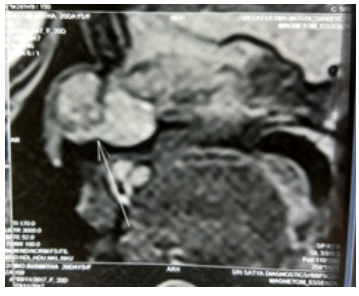


fig 3



2. DISCUSSION

Congenital midline nasal masses are rare anomalies that occur in about one in 20000–40000 live births [1]. Nasal gliomas account for approximately 5% of all congenital nasal swellings. These usually arise during infancy or later childhood with relative peaks of occurrence between 5 and 10 years of age. Although the majority of patients present during the first year of life, a later presentation may be due to a specialist's failure to recognize a subclinical lesion in childhood. Approximately 250 cases have been reported in the literature.3

The term “nasal glioma” is a confusing misnomer as it implies a neoplastic condition, which it is not. It needs to be differentiated from glioma, which is a malignant tumor of the brain [1, 2]. 60% of these gliomas are extranasal, lying external to the nasal bones and cavities; 30% are intranasal lying within the nasal cavity, mouth, or pterygopalatine fossa and 10% are mixed, dumbbell shaped communicating through a defect of the nasal bones [3]. Our case was mixed glioma.

The possible theories of development of nasal gliomas include the following: (a) sequestration of glial tissue of the olfactory bulb entrapped during cribriform plate fusion; (b) ectopic neural tissue

cells; (c) encephaloceles with lost intracranial connection and meningeal continuity; (d) inappropriate closure of the anterior neuropore (fonticulus frontalis) [1, 4]. The most widely accepted hypothesis is the "encephalocele theory," which states that both the encephalocele and glioma develop secondary to a failure of regression of the forebrain dural protrusion through the foramen cecum or fonticulus frontalis.2 This failure of regression is postulated to result from a sustained connection between neural and surface ectoderm, due to insufficient apoptosis at the final closure site of the rostral neuropore.4 In encephaloceles, the connection to the brain is maintained, while in gliomas this connection is obliterated, leaving a nest of tissue.]. In 15–20% of cases, a fibrous stalk exists to connect them to the intracranial space

Histologically, these lesions are made up of astrocytic neuroglial cells interlaced with fibrous and vascular connective tissue [5] that may be covered with skin or nasal respiratory mucosa. True capsule is absent, and mitosis is rare. The glial nature of the cells can be further confirmed by immune-histochemical demonstration of S100 protein and GFAP.

CT scan or MRI forms the mainstay of investigation as fine needle aspiration cytology or excision biopsy carries a significant risk of meningitis or CSF leaks [6]. CT scan demonstrates bony defects, and MRI can demonstrate the characteristics of the soft-tissue mass and its possible intracranial connection. On CT, the mass is usually isodense to brain tissue. On MRI, the lesion is isointense to hypointense relative to gray matter on T1-weighted sequences and hyper-intense on T2-weighted and proton density sequences [5, 7]. Magnetic resonance imaging also has an advantage of minimizing the level of exposure to ionising radiation, particularly in infants.

The clinical differential diagnosis of nasal gliomas includes several disorders, which can present as nasal masses [1, 8]. Some of such important lesions include: (a) nasal dermoids, which constitute the most common congenital nasal anomaly and are cavities or sinus tracts possessing epithelial lining and variable numbers of skin appendages, including hair follicles, sebaceous glands, and eccrine glands; (b) encephaloceles which constitute the lesions caused by herniation of neural tissue through defects in the skull. They may contain meninges (meningocele) or brain matter and meninges (encephalomeningocele), or they may communicate with a ventricle (encephalomeningocystocele). Encephaloceles are etiologically similar to nasal gliomas as per one of the theories; (c) hemangioma which are the most frequent benign vascular tumors in infancy.

The treatment of choice of nasal gliomas is complete surgical excision [1]. Gliomas are benign but incomplete excision results in a 4% to 10% recurrence rate. The approach depends upon the location and extent of the lesion [1] and levels of expertise available. Extranasal gliomas can be excised via lateral rhinotomy, external rhinoplasty, midline nasal incision, or a bicoronal incision. For both intranasal and extranasal gliomas, removal of the stalk is imperative not only to decrease the rate of recurrence but also to minimize the chances of a cerebrospinal fluid leak and subsequent meningitis. If intracranial extension is evident, than frontal craniotomy, multidisciplinary team approach may be required [9] in specialized neurosurgical or craniofacial centers to ensure complete and safe excision of the lesions.

3. CONCLUSION

Nasal gliomas and other anterior craniofacial masses are uncommon lesions. . It is mandatory to rule out intracranial extension by cross-sectional imaging, preferably by MRI before performing any invasive procedure. Nevertheless, when present, these masses are uniformly managed surgically, with early operative intervention believed to correlate with more favorable aesthetic outcomes. Advances in imaging technologies have provided clinicians from various specialties with an opportunity to facilitate earlier diagnoses and timely surgical consultations

REFERENCES

1. Nada V, Dejan V, Dragan D, Ljiljana J. Nasal glioma. Archive of Oncology. 2006;14(1-2):57–59.
2. Ducic Y. Nasal gliomas. Journal of Otolaryngology. 1999;28(5):285–287. [PubMed]
3. Cerdá-Nicolás M, Sanchez Fernandez de Sevilla C, Lopez-Ginés C, Peydro-Olaya A, Llombart-Bosch A. Nasal glioma or nasal glial heterotopia? Morphological, immunohistochemical and ultrastructural study of two cases. Clinical Neuropathology. 2002;21(2):66–71. [PubMed]
4. Swift AC, Singh SD. The presentation and management of the nasal glioma. International Journal of Pediatric Otorhinolaryngology. 1985;10(3):253–261. [PubMed]
5. Singhal SK, Virk RS, Dass A, Bansal S. Amanjit: neonatal nasal glioma: a case report. Internet Journal of Otorhinolaryngology. 2006;4:p. 2.
6. Clarós P, Bandos R, Clarós A, Gilea I, Clarós A, Real M. Nasal gliomas: main features, management and report of five cases. International Journal of Pediatric Otorhinolaryngology. 1998;46(1-2):15–20.[PubMed]
7. Jartti PH, Jartti AE, Karttunen AI, Paakko EL, Herva RL, Pirila TO. MR of a nasal glioma in a young infant. Acta Radiologica. 2002;43:141–143. [PubMed]
8. Dasgupta NR, Bentz ML. Nasal gliomas: identification and differentiation from hemangiomas. The Journal of Craniofacial Surgery. 2003;14(5):736–738. [PubMed]
9. Rahbar R, Resto VA, Robson CD, et al. Nasal glioma and encephalocele: diagnosis and management. Laryngoscope. 2003;113(12):2069–2077. [PubMed]