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CASE REPORT

Endoscopic Approach a Rare Adult Nasal Glioma and Review of the Literature

Mehmet Akdag [1], Ulaş Alabalik [2], Abdurrahim Dusak [3], Bülent Aguloglu [1]

[1]*Department of Otolaryngology, Faculty of Medicine, University of Dicle, 21280, Diyarbakir, Turkey*
[2]*Department of Pathology, Faculty of Medicine, University of Dicle, 21280, Diyarbakir, Turkey*
[3]*Department of Radiology, Faculty of Medicine, University of Dicle, 21280, Diyarbakir, Turkey*

Abstract

Nasal gliomas are rare, benign, congenital tumors that are throught to be the result of abnormality in embryonic development. Three types of clinical presentations have been recognized; extranasal, intranasal and combined. Clinically, these masses are non-pulsatile, gray or purple lesions that obstruct the nasal cavity and can lead to various complications. Histologically, they consist of astrocytic cells, fibrous and vascular connective tissue covered with nasal respiratory mucosa. Treatment of nasal glioma requires a multidisciplinary approach including a radiologist, neurosurgeon and otorhinolaryngologist. Radiological investigation should be performed to determine intracranial extension. This report describes a 60-year-old male undergoing endoscopy for nasal mass and postoperative pathology presenting as nasal glioma.

To the best of our knowledge there are few published reports of adult glioma arising from the nasal cavity. The mass was excised totally using an endoscopic approach.

We describe the clinical and histopathological features of this rare lesion, with a complete review of the relevant literature.

Key words: *Nasal glioma, embryonic development, Nasal mass.*

* Corresponding author: E-mail: drmehmetakdag @hotmail.com
Phone: +90 412 2488001/4492
Fax: **+90 412 248 8440**

Introduction

Congenital midline mass anomalies are very rare. Dermoid cysts, gliomas, encephalocele and hemangiomas are frequently seen. Nasal gliomas are rare, benign, congenital tumors thought to be the result of abnormality in embryonic development. These abnormal embryonic development; during the closure of the nasal and frontal bone due to extracranial location occurs as a result of ectopic glial tissue [1,2]. The incidence of congenital nasal masses is 1 in every 20,000 to 40,000 live births [1]. Nasal glioma is frequently symptomatic in children with no malignant potential, whereas nasal glioma in adults is very rare and may appear as an extranasal (60%), intranasal (30%) or combined (10%) lesion. Some authors therefore refer to the condition as glial heterotopias [4].Nasal glioma was first described by Reid in 1852 [1,3].

Nasal gliomas are often noticed at birth but are rarely detected in adult patients. Intranasal glioma often manifests as a pale mass within the nasal cavity with protrusion from the nostrils.

Few cases of adult nasal glioma have been reported in the world [5-13]. We describe a rare case of nasal glioma presenting in adulthood with right orbital cellulitis, proptosis and visual disturbance. It mimicked nasal polyp and was surprising in terms of the patient's age. The clinical manifestations, diagnosis and treatment of this clinical entity are discussed, and a complete review of the literature is presented.

Case Report

A 60-year-old patient was admitted to the Dicle University hospital in Turkey with swelling around the right eye, high fever, rhinorrhea and nasal obstruction. Minimal hyposmia was also determined. The patient had no antecedent history of orbital symptoms. Initial examination revealed marked right orbital edema, hyperemia and proptosis. Endoscopic rhinoscopic examination revealed a pale, well-defined, hard, gray mass of about 1x1.5 cm in size covered with a mucous membrane with a smooth surface, sero-mucoid discharge in the right nasal passage and nasal congestion in the nasal cavity. Post-nasal discharge was also present.

Systemic examination of the head and neck revealed no other pathological condition. The patient was diagnosed with rhinosinusitis complicated with nasal polyposis, confirmed as compatible with existing clinical preoperative imaging. Interestingly, there was the presence of loss of vision with the patient in a reclining position. Ocular examination revealed marked right proptosis with associated severe impairment of visual acuity (6/10) and loss of color vision. Fundoscopy did not reveal papilloedema.

Rigid endoscopic examination revealed a pale, nonpulsatile mass obstructing the right nasal cavity emanating from above the middle turbinate. Hypertrophy of the turbinate of left nasal cavity was also present. The mass was not compressible and did not transilluminate or alter in size during Valsalva's maneuver. The Furstenburg test (compression of the ipsilateral internal jugular vein and observation for any increase in the size of the mass) was negative.

Computerized tomography (CT) revealed bilateral different degrees of mucosal thickening in the soft tissue and maxillary sinus, ethmoid cell lines and frontal sinuses (Fig. 1).

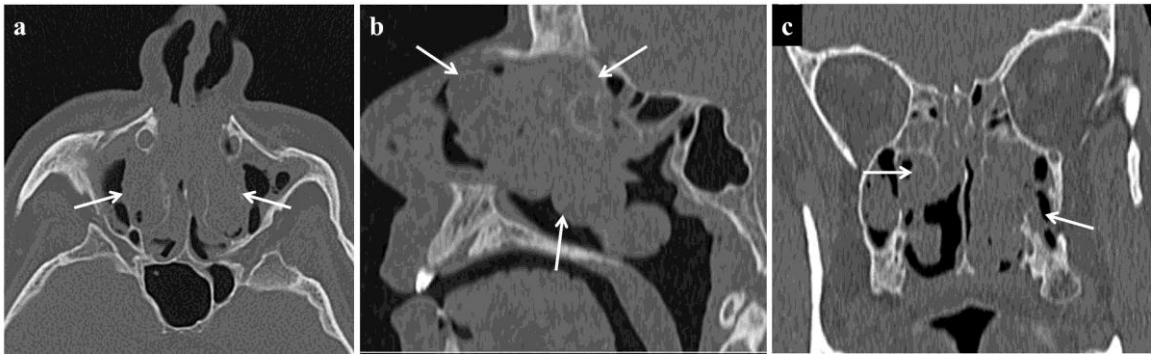

Figure 1 : A 60-year-old man with a nasal mass. The axial (a), sagittal (b) and coronal (c) non-contrast CT images reveal a low-attenuation mass (*arrows*) within the nasal cavity and mucosal thickening. Osteomeatal complexes are occluded.

No obvious dural envelope was seen. Magnetic resonance imaging (MRI) was performed to further evaluate the nasal mass. MR was confirmed the earlier CT findings (sino-nasal polyposis?)(**Fig. 2**).

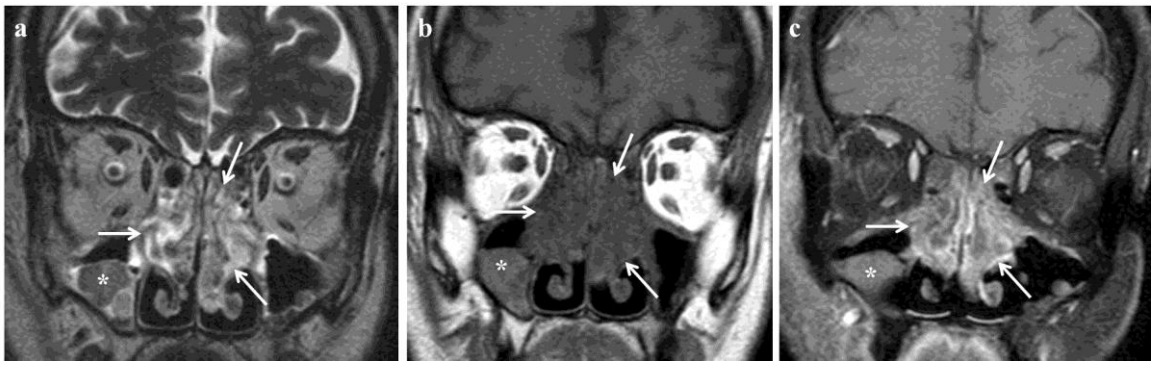

Figure 2 : (a) T2W MR image reveals a heterogeneously hyperintense, (b) T1W heterogeneously hypointense and (c) T1W contrast-enhanced image reveals a heterogeneously enhancing mass within the nasal cavities.

Review sections in the field of the right orbital and periorbital subcutaneous tissue thickness slightly increased density available (cellulitis?). There was considered the periorbital cellulitis with acute rhinosinusitis and received medical treatment firstly. The patient was immediately started on a course of intravenous dexamethasone (once a day [1 mg/kg per day]), and an ophthalmologist opinion was sought. Orbital edema and hyperemia resolved within five days, and visual acuity improved on the right side. Elective bilateral functional endoscopic sinus surgery was planned after orbital symptoms and signs resolved with medical treatment. We excised and cleaned the nasal polypoid masses filling the right nasal passage, the maxillary sinus and ethmoid cells. No bleeding or rhinorrhea were observed in the postoperative period. The lesion was limited to the nasal mucosa, with no extension into the intracranial space. At gross examination, the lesion consisted of two parts, 18 x 15 x 10 mm and 16 x 14 x 10 mm, in size. Both were pale and firm, with solid gray surfaces. There was no evidence of cerebrospinal fluid (CSF) leakage pre-operatively, during surgery or postoperatively. Following complete excision, the specimen was fixed in 10% formaldehyde and submitted for histopathological examination. The fixed specimen was embedded in paraffin, and 4-mm sections were stained with hematoxylin and eosin (HE) or periodic acid-Schiff (PAS) stain. Similarly cut sections were processed for immunohistochemistry using glial fibrillary acidic protein (GFAP) (**Fig. 3**).

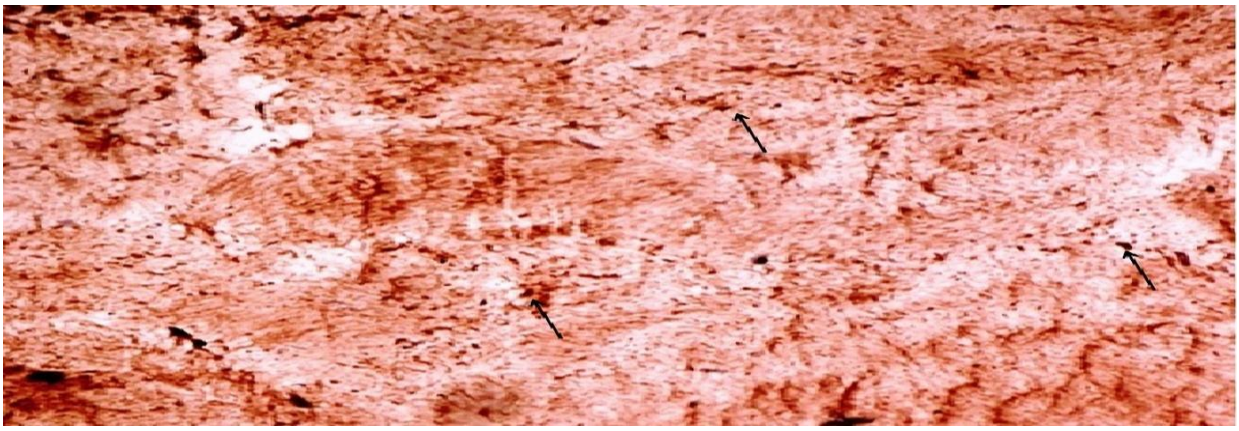

Figure 3: Low magnification. The glial component showed typical features of a reactive gliosis, with enlarged astrocytes with thick and large nucle with normal chromasia. (H&E,40x).

The diagnosis was a histological surprise following excision biopsy for a presumed nasal mass. There was founded rich glial view, fibrotic foci in respiratory epithelial tissue places with Haematoxylin-eosin staining sections taken from in the pathology that the excised specimen. GFAP staining was positive in the sections (**Fig. 4**).

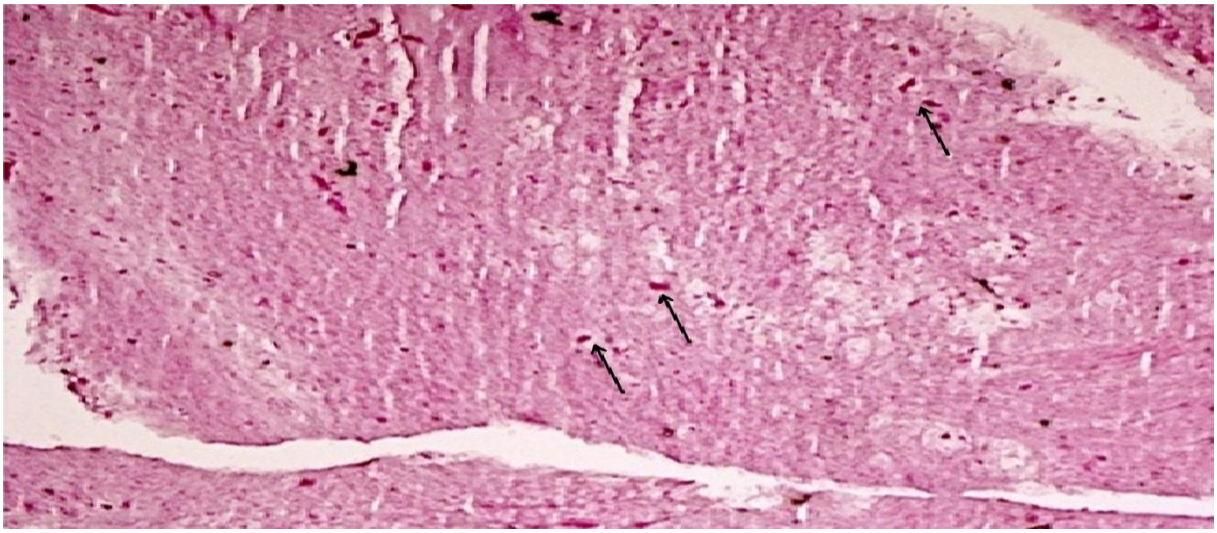

Figure 4: Intense staining of glial elements in response to Glial fibrillary acidic protein (GFAP). GFAP staining stain confirmed the glial nature of the lesion. (Immunoperoxidase-antiperoxidase staining, (40x)

Discussion

Although our case was mostly similar to cases reported in the literature, it was difficult to diagnose without histopathological examination. Nasal glioma is not familial, no sex predilection, and is generally not associated with another developmental abnormalities[2] whereas nasal glioma is commonly seen in men. Most published cases [5, 6, 8, 9, 10, 12, 13] (**Tab. 1**) and our own case was male interesingly.

Diagnosis of nasal glioma is facilitated by the appropriate use of cross-sectional imaging studies. CT is useful in visualizing bony defects in the anterior skull base [14], while MRI provides

complementary information regarding the fluid or soft tissue characteristics of the mass [15]. MRI also provides a three-dimensional view and can determine the presence or absence of any intracranial extension [14]. However, pre-operative radiological evaluation should not lead to a false sense of security if no bony defect is identified. No defect was observed in imaging in our case. The mass formation in the nasal cavities was not connected to the intracranial structures. One of the interesting features of our case is that mucosal thickening was bilateral in the imaging system. Nasal mass-associated anomalies are usually unilateral. Another interesting finding was the lack of clarity between the skull base and nasal space in the imaging system. Other important issues, such as congenital anomalies, are frequently encountered during childhood.

Table 1: *Clinical and Demographic Information of Adult Nasal glioma in Literature*

Patient Number	Year	Authors	Age	Gender	Surgical Approach	Radiological Work-up	Pathological Work-up	Location	Intracranial Extension
1	1963	KR, Smith, Ogura	54 25	M, F	bifrontal craniotomy	Pneumo-encephalogram,	hematoxylin and eosin(HE)& Electron microscopy	Roof of left frontal sinus, anterior part of the cribriform	Yes
2	1987	Pollard K,	25	M	craniofacial resection	CT	S-100 and GFAP	Nasal	Yes
3	1987	G. Altissim	81	M	Intranasal endoscopic resection	CT	S-100 and GFAP	Right nasal	No
4	1998	Pasquini E	17	M	Intranasal endoscopic resection	MR (Gadolinium)	HE	Right nasal	No
5	1999	Ducic Y	42	M	Transfacial lateral rhinotomi	CT/MRI	HE	Anterir cranial region	Yes
6	2005	Chau HN	37	M	craniofacial resection	Three dimensional reconstruction of CT scan	GFAP	Right nasal	Yes
7	2008	Michael T. Tetzlaff	37	M	craniofacial resection	CT	HE, Ki 67	Right nasal	Yes
8	2010	M.K. Kasliwal	40	M	Intranasal endoscopic resection	MRI	GFAP	Left Meckel's cave region	No
9	2010	A Majithia	41	F	Conservativ e therapy	CT/MRI	HE	Left nasal	Yes

*CT: Computerized tomography ** MR: Magnetic resonance imaging ***GFAP: Glial Fibrillary Acidic Protein

Congenital nasal anomalies should therefore be borne in mind in differential diagnosis of patients with adult polypoid masses. When there is no erosion of the lateral nasal wall with loss of vision, such as in our case, may suspected congenital anomalies. Majithia *et al.* reported a rare case of a nasal glioma detected incidentally in an adult presenting with visual loss [11]. The critical distinction between nasal glial heterotopia and encephalocele is that the latter retains a patent connection with the central nervous system, whereas the glioma is connected by a fibrous stalk or disconnected from the CNS entirely[7, 16]**.** Therefore, methods of imaging should be refer. This matter supported our study. Our scan of preoperative imaging in the literature revealed four CT, two MRI, two combined CT and MRI and one pneumoencephalogram. We performed both CT and MR.

Nasal glioma is rarely diagnosed without histological examination. Histologically, nasal gliomas are composed of astrocytes, and neurologically of vascular and soft tissue cells embedded in fibrous masses. There are no real capsules. There were consist of multinucleated and germitositik astrocytes and rarely can be observed neurons. Encephalocele should be considered in the differential diagnosis of histologically detected heavy amounts neurons. Glial tissue as determined by immunohistochemical staining for GFAP and S100 protein. These two proteins showed high specificity and identification of neurological cells, allows the separation of granular cell tumor and meningiomas[17]. We also reviewed histopathological analyses that using different techniques in literature. Some used only light or electron microscopy. Some also used glial cells specific for S-100 protein (n-3), or GFAP (n-4). In our case, GFAP was used in the preparation of the specimen.

Nasal gliomas in adults may treated by surgical excision. Preoperative differential diagnosis is essential prior to surgery. Location, size and the connection intracranial structures of the lesion should be determined prior to the operation. With recent advances in endoscopic equipment and techniques, proper exposure and complete excision of an intranasal glioma can now be achiev and removed [8]. This makes it possible to perform a comprehensive preoperative evaluation and design the most appropriate surgical plan. Endoscopic examination can also be easily performed for follow-up [18]. The literature contains reports of nine patients treated with surgical resection, including the lateral rhinotomy approach (n=1), craniofacial resection (n =3), bifrontal craniotomy (n=2) and intranasal endoscopic resection (n =3).However, from 1971 to the present, nasal endoscopicsurgery have been preferred to conventional surgery such as rhinotomymethods in adult nasal gliomas.Because there was less cosmetic damage and morbidity in intranasal endoscopic surgery. Overall, the surgical approach should be based on the location and size of the mass.

In conclusion, nasal glioma should be considered in the differential diagnosis of nasal masses in the presence of nasal polypoid mass with orbital complications. The present case was similar to previously reported cases, revealing a characteristic and consistent clinical and morphological picture. Recognition of this clinical picture may allow preliminary diagnosis by the otolaryngologist, but final diagnosis should be based on the characteristic histopathology.

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