Case Report / Olgu Sunumu

Ganglioglioma in the nasal cavity: a case report

Burun boşluğunda gangliogliom: Olgu sunumu

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Ganglioglioma is a tumor containing both astrocytic and neuronal components. It may occur any where in the central nervous system and spinal cord but is only encountered rarely. Nasal glial heterotopia (also known as "nasal glioma"), is a rare developmental abnormality seen in a wide age group. Gangliogliomas may also manifest as a nasal glial heterotopia, and neurogenic tumors should be considered in the presence of a nasal mass. In this article, we present a case of ganglioglioma located in the right-nasal cavity. The mass was excised totally through an endoscopic approach. The ganglioglioma developed on a nasal glial heterotopia base. To our knowledge, a ganglioglioma arising from the nasal cavity has not been described previously in the literature.

Key Words: Benign tumor; ganglioglioma; nasal cavity, surgery.

Gangliogliomlar hem astrositleri hem sinir elemanlarını içeren tümörlerdir. Santral sinir sistemi ve omuriliğin her yerinde oluşabilir ancak nadir görülürler. Nazal glial heterotopi (nazal gliom olarak da bilinmektedir) geniş yaş grubunda görülen gelişimsel anomalilerdir. Gangliogliomlar nazal glial heterotopi olarak karşımıza çıkabilir ve burun kitlesi varlığında sinir kaynaklı tümörler mutlaka önemsenmelidir. Bu makalede sağ burun boşluğuna yerleşen gangliogliom olgusu sunuldu. Kitle endoskopik yaklaşımla tam olarak çıkarıldı. Gangliogliom nazal glial heterotopi zemininde gelişmişti. Bilgilerimize göre burun boşluğundan kaynaklanan gangliogliom literatürde daha önce bildirilmemiştir.

Anahtar Sözcükler: İyi huylu tümör; gangliogliom; burun boşluğu, cerrahi.

Neurogenic tumors in the nasal cavity are rare. It is a type of a congenital tumor, nasal glial heterotopia (also called nasal glioma), is extracranial cerebral tissue, with no functional relationship to the brain, that can occur extranasally and intranasally as well as on the face.[1,2] In contrast to the majority of gliomas, gangliogliomas are tumors containing both astrocytic and neuronal components. Although most gangliogliomas are observed in the brain, some may be found in unusual locations, such as the pineal gland, hypothalamus, and optic chiasma.[3] There are no reports describing a ganglioglioma in the nasal cavity. We report a case of a ganglioglioma in the right nasal cavity based on nasal glial heterotopia.

CASE REPORT

A 20-year-old female was admitted to our clinic with a complaint of a sudden, intense yellow-colored bloody discharge from her right nostril. A review of her history revealed a dacryocystorhinostomy operation for nasolacrimal

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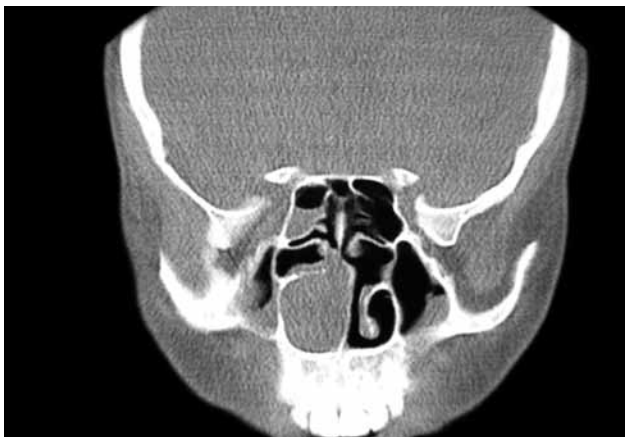

Figure 1. *Coronal section paranasal sinus computed tomography demonstrated a mass in the right nasal cavity, eroding the os concha inferior and air-fluid level in right ethmoid sinuses.*

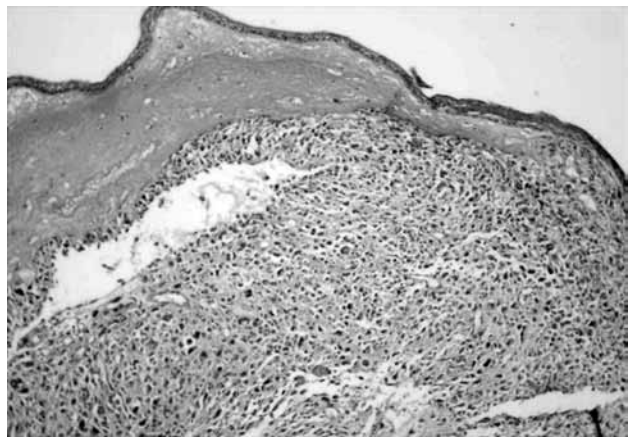

Figure 2. *Photomicrograph of hematoxylin and eosin stained section showing tumoral infiltration under the squamous and prismatic epithelium (Glial heterotopia) (H-E x 200).*

duct obstruction two years ago. During physical examination of the patient, a bloody fragile mass that completely filled the right nasal cavity was detected. An intracranial connection was not determined, and hematologic tests were normal. Erosion of the inferior nasal concha and air-fluid level in the right ethmoid sinuses by a mass in the right nasal cavity was demonstrated using paranasal sinus computed tomography (CT) (Figure 1). After conducting a punch biopsy of the mass, a histopathologic diagnosis of ganglioglioma was made. Because of the limited extent of the lesion on the CT scan and diagnostic nasal endoscopy, we planned an endoscopic approach for excision under controlled hypotensive anesthesia. Preoperatively, a mass was detected in the anterior $^{2}/_{3}$ of the left inferior turbinate. The posterior end of the inferior turbinate and choana were normal, and the middle meatus and osteomeatal complex were bilaterally normal. The entire mass was excised by an endoscopic-assisted dissection with bipolar cautery. The postoperative period was uneventful.

Based on a macroscopic examination, the resected material had a diameter of 4x2x2 cm, an irregular outer surface, and smooth bloody-colored outlines. Tissue fragments were fixed in 10% buffered formalin and processed according to protocol. Sections (4 μm thick) were stained with hematoxylin and eosin (H-E). Additionally, glial fibrillary acidic protein (GFAP) (Dako, Glostrup, Denmark), epithelial membrane antigen (EMA), CD34 and CD68 antigens, vimentin, cytokeratin, neuron specific enolase (NSE), S-100, and

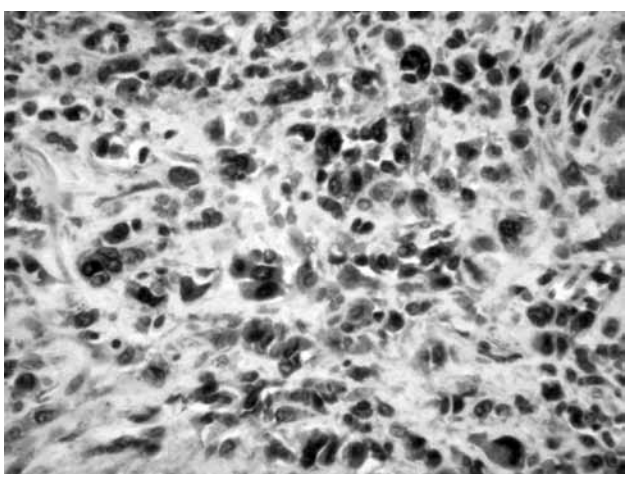

Figure 3. *Neuron specific enolase positivity and ganglial cells in specimen (H-E x 400).*

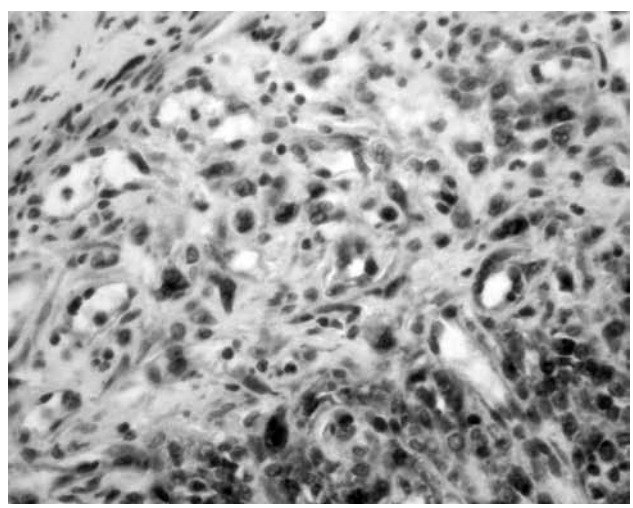

Figure 4. *S-100 positivity in glial tissue (H-E x 400).*

Ki-67 were applied. There was tumoral infiltration under the squamous and prismatic epithelium (Figure 2) as well as glial and neuronal proliferation. Some of the cells were pleomorphic, bizzarely shaped, and closely arranged in some areas. Mitosis and tumorigenic necrosis was not observed. Immunohistochemically, GFAP, S-100, and NSE were positive, whereas the Ki-67 index was negative (Figure 3, 4). A pathologic diagnosis of ganglioglioma was made based on the morphologic and immunologic findings.

DISCUSSION

Extracranial glial tissue is very rare. First described by Reid in 1952 and still in use today, the term 'nasal glioma' (or nasal glial heterotopia) is a misnomer since it does not refer to an actual tumor. Based on the 300 cases mentioned in literature, the heterotopic tissue is treated by total excision with a 4-10% chance of recurrence.[2]

Tumors in the nasal cavity have been observed in both the young and elderly.[1,4] Based on histopathologic examination of the gliomas and light microscopy of the astrocytic groups and fibrovascular tissues, neurons are not observable in 90% of the gliomas. This may be due to insufficent oxygen or a possible defect in neuronal differentiation of the embryonal neuroectoderm.[5] An intracranial connection can exist in 15% of the nasal gliomas, so this probability should not be underestimated during radiological evaluations.[1]

In contrast to most gliomas, gangliogliomas contain both astrocytic and neuronal components.[5] Courville first described a ganglioglioma in 1930, and Rubinstein and Herman[6] described its classic ultrastructural features. The pathological spectrum of a ganglioglioma can be diverse. At one end of the spectrum, a 'gangliocytoma' comprises mainly neoplastic ganglion cells; at the other end of the spectrum, it generally comprises neoplastic glial cells.[7] Such differences suggest that these tumors have hamartomatous structures and are histologically benign.[8] Dense vesicles that are histologically detected and tyrosine hydroxylase that exists in the neural component are indicative of ectopic neural crest cells. Also, glial tissues are predominantly aggressive in appearance.[9] Histopathological appearances of glioneural tumors and gangliogliomas are designated by the World Health Organization and are now beginning to be placed into a different tumor category.[10] Observed in the brain, especially in the temporal lobe, gangliogliomas can be found in the frontal lobe, third layer of the subventricular zone, pineal gland, hypothalamus, and optic chiasma as well as the brainstem and spinal cord.[8] Gangliogliomas are more commonly observed in children than adults. For these slowly developing tumors that are rarely malign,[11] surgical resection is the method of treatment.

Gangliogliomas have been identified based on heterotopia. In our case, identification of the ganglioglioma was based on nasal glial heterotopia and has never been reported before. Even though in 2008 Niedzielska et al.[5] reported a ganglioglioma located in the mid-frontonasal zone with an intracranial connection, our case is totally intranasal in origin and has no intracranial connection. It is the first case with such properties.

The ganglioglioma must be surgically treated by total excision,[8] but the surgeon must estimate the probability of intracranial connection. In our case, the entire mass was excised by endoscopic-assisted dissection, and no intracranial connection was observed. Recurrence in the patient was not postoperatively observed in month 12 of the follow-up.

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