An unusual mass of nasal cavity: recurrent glomangiopericytoma

Nazal kavitede nadir bir kitle: Reküren glomanjoperisitom

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ABSTRACT
Hemangiopericytomas are rare tumors of sinonasal region. In this article, we described a case of recurrent mass in right nasal cavity which presented with nasal obstruction and intermittent epistaxis. Patient had a surgical history on the same nasal region without any histopathological document. Abnormal bleeding was observed during surgery and mass was partially removed for histopathological investigation. The result was glomangiopericytoma of the sinonasal region. Although extended surgery was recommended to the patient including preoperative and perioperative measures for bleeding problem, patient preferred to follow-up rather than removal. The clinical progress and review of glomangiopericytoma have also been discussed.

Keywords: Epistaxis; recurrent glomangiopericytoma; sinonasal tumor.

ÖZ

Anahtar Sözcükler: Epistaksis; reküren glomanjoperisitom; sinonasal tümör.

Hemangiopericytoma originate from pericytes of vascular structures and are very rare vascular tumors.1,2 Head and neck hemangiopericytoma are a group of tumors that develop in the head and neck region.3 Although they have similar properties with other hemangiopericytoma they are accepted as having a better prognosis. They differ especially in histopathological features as they have more myogenic differentiation like a glomus tumor. They are also called glomangiopericytoma or sinonasal type hemangiopericytoma and account for less than 0.5% of all sinonasal tumors.4 The etiology is not clear but trauma, hypertension, pregnancy and use of corticosteroids are accepted as causes.

We present a case of regrowth of glomangiopericytoma in the right nasal cavity.
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It is noteworthy because they are very rare tumors in the sinonasal cavity, histopathological diagnosis may be difficult due to similar lesions and improper surgical intervention may result in repetition of operations as in our case, which may have multiple complications.

CASE REPORT

A 32-year-old man was admitted to our unit with right nasal obstruction, difficulty in breathing and intermittent epistaxis. He had a history of surgical intervention that took place three years ago in another country for a mass lesion of the right paranasal sinuses. He did not have any documentation of the surgical procedure or the pathological result. The patient only knew that he had a benign tumor in his right nasal cavity and also had hemorrhage during surgery. Due to the hemorrhage, the mass was not removed totally. The patient was not referred for further treatment after the surgical intervention. We were not able to obtain the patients’ documents because of the unavailability of his surgeons and the hospital management.

The patient’s complaints had been better in the early postoperative period after surgery, but reappeared last year. His main complaint was nasal obstruction, difficulty in breathing especially in the right nasal cavity as well as intermittent epistaxis. He had no other disease and did not use any drugs. He only had nasal decongestant abuse 10 years ago in his history.

On examination, there was a polypoid tissue originating from the lateral wall of the right nasal cavity, which extended into the nasal cavity through the middle turbinate by penetrating it in the middle. There was no significant structure (such as a vascular network) noted. There were no other pathological findings in the otorhinolaryngologic examination.

A computed tomographic scan of the paranasal sinuses showed a mass located in the right nasal cavity and maxillary sinus. There was no significant appearance in the mass such as septa or enhancement. Other nasal structures appeared normal.

Endoscopic sinus surgery including mass excision was planned. Nasal examination identified a mass lesion originating from the right maxillary posteromedial region which penetrated the middle turbinate and extended into the right nasal cavity. During exposure of the mass lesion borders, an abnormal profuse hemorrhage from the mass ensued.

After multiple biopsies, hemorrhage control was achieved. Total removal of the mass was not carried out, because of the unexpected situation and insufficient preoperative preparation (lack of erythrocyte suspension and informed consent).

Figure 1. Preoperative paranasal sinus scanning coronal section images.
The specimens were analyzed with a panel of immunohistochemical markers [smooth muscle actin (SMA), desmin, vimentin, beta-catenin, and CD31]. The tumor was composed of closely packed cells, forming short fascicles exhibiting a palisaded pattern, interspersed with many vascular channels. Vascular channels were in the form of capillary-sized to large sized staghorn configuration. The overlying respiratory and squamous epithelium remained intact. The tumor cells were diffusely positive for vimentin and beta-catenin; on the other hand, there was focal staining for SMA. There was no reaction in the neoplastic cells with CD31.

Nasal packages were removed after two days and no complication was observed. The patient was discharged after two days. After the possible complications were explained, the patient decided to wait a little more and further surgery will be planned.

**DISCUSSION**

Hemangiopericytomas originate from pericytes, which means that they can originate in every part of the body where capillaries are present.\(^2\)\(^3\) 7.5 to 25% of all hemangiopericytoma are found in the head and neck region, especially in the nasal cavity and paranasal sinuses.\(^4\)

They have a significant pericytic vascular pattern with thin-walled, branching vessels often with a staghorn configuration.\(^5\) Hemangiopericytoma-like lesions may also show similar histopathological patterns.

Most of the lesions found in the head and neck region differ from classical hemangiopericytoma histopathologically,
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as they have a more prominent myogenic appearance and are called hemangiopericytoma-like lesions. Glomangiopericytoma or sinonasal-type-hemangiopericytoma is used instead of hemangiopericytoma-like lesions. This definition also describes the tumor thought to derive from perivascular modified smooth-muscle cells.[3] Glomangiopericytoma generally shows strong immunoreactivity for muscle-specific markers, such as SMA and muscle specific actin in contrast to classical hemangiopericytoma.[6] Our case has similar histopathological findings as well.

Besides having histopathological differences, glomangiopericytomas also have differences in clinical appearance. Glomangiopericytomas generally tend to have more indolent patterns when compared to hemangiopericytoma. Histopathological and clinical differences make glomangiopericytomas a different entity.[7]

Glomangiopericytomas are rare tumors and account for 0.5% of all sinonasal tumors. They have a very slight female preponderance and a peak incidence in the sixth and seventh decade of life.[8] They are variable in size and firm, beefy red or fleshy, soft and hemorrhagic in appearance.[8] Main symptoms are nasal obstruction and epistaxis as in our case. Patients may be also suffer from non-specific findings such as a mass, polyp, difficulty in breathing, sinusitis and headache.

Trauma, hypertension and long-term steroid use are said to be possible etiologic factors, but in our case we noticed abnormal nasal decongestant use instead of the reported etiologic factors.[9]

Glomangiopericytomas generally originate in the nasal cavity and may extend to the paranasal sinuses, although isolated paranasal sinus involvements have been reported.[7] After examination, imaging techniques must be carried out for differential diagnosis of mass lesions of the nasal cavity. Computed tomography scanning is always carried out and magnetic resonance imaging scanning may be necessary in some situations.[10]

The differential diagnosis of hemorrhagic sinonasal mass includes conventional hemangiopericytoma and all soft tissue tumors with a similar pattern like solitary glomangioma, myopericytoma, angioleiomyoma and glomus tumors.[11]

Histological examination is important for diagnosis. Modified perivascular glomus like myoid cells are the proposed cells of origin. Hematoxylin-eosin staining shows a well delineated subepithelial but nonencapsulated cellular tumor, characterized by diffuse growth of closely packed spindle cells growing in a variety of patterns.[8,12] These cells form short fascicles and sometimes exhibit storiform, whorled or palisaded patterns, interspersed with numerous thin-walled, branching staghorn vessels. The neoplastic cells are uniform and oval to spindle shaped. On differential diagnosis, glomangiopericytomas do not stain for demsin, S100, CD34, but do for SMA and vimentin.[2,5,7]

The gold standard treatment of glomangiopericytomas is total surgical removal of the mass with tumor-free margins.[13] An endoscopic approach is generally preferred.
instead of much more invasive approaches. Insufficient removal may lead to recurrences and a 16.8% recurrence rate has been reported. Because of the tendency for recurrence, close follow-up is usually recommended.

Postoperative radiation therapy is recommended in some cases. Like nasopharyngeal angiofibromas, preoperative embolization of afferent vessels may reduce the blood supply and may be useful during surgery.

The prognosis of glomangiopericytomas is good. Glomangiopericytomas are categorized as a borderline low malignancy tumor by the World Health Organization classification. There were only a few cases in which metastases were reported.

Malignant glomangiopericytomas are uncommon and display nuclear pleomorphism, high mitotic activity and necrosis.

Conclusion

Glomangiopericytomas are very rare tumors of the sinonasal cavity. Although several cases have been reported in literature, recurrent cases in the paranasal region have not been reported. The main complaints of glomangiopericytomas are nasal obstruction and epistaxis. It must be kept in mind in the differential diagnosis of hemorrhagic lesions of nasal cavity. Incomplete surgery may result in recurrences or regrowth of lesions.

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