Case Report / Olgu Sunumu

Hemangiopericytoma like tumor of the nasal cavity: a case report

Nazal kavitede hemanjiyoperisitom benzeri tümör: Olgu sunumu

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Sinonasal hemangiopericytoma originates from vascular pericytes. It may arise in any part of the body but it is an uncommon vascular tumor in the nose. It can show benign or malignant histologic features. Sinonasal hemangiopericytoma like tumor is important as it is recognized as a special variant of hemangiopericytoma. In this article, we present a 65-year-old male patient with hemangiopericytoma like tumor of the nasal cavity.

Key Words: Nasal obstruction; sinonasal hemangiopericytoma; vascular tumor.

Sinonazal hemanjiyoperisitom, vasküler perisit kaynaklıdır. Vücudun herhangi bir bölgesinde görülebilmekle birlikte, burunda nadir rastlanan bir vasküler tümördür. Benign veya malign histolojik özellikler gösterebilir. Hemanjiyoperisitomun özel bir türü olduğu için, sinonazal hemanjiyoperistom benzeri tümör de önemlidir. Bu makalede, nazal kavitede hemanjiyoperisitom benzeri tümör saptanan 65 yaşındaki bir erkek hasta sunuldu.

Anahtar Sözcükler: Nazal obstrüksiyon; sinonazal hemanjiyoperisitom; vasküler tümör.

Sinonasal hemangiopericytoma like (SHPCL) tumors are rare mesenchymal tumors which slowly enlarge at a local area.[1,2] Diagnostic differentiation between malignant and benign types of these tumors is rather difficult. Certain pathologic factors may correlate but there is no specific diagnostic test. It may be necessary to differentiate SHPCL tumors from other sarcomatous tumors.[3] Therefore clinical correlation as well as histological confirmation, electron microscopic examination and immunohistochemical staining is necessary to establish a diagnosis. It has a low incidence of metastasis and very little risk. Treatment consisting of wide local excision is usually curative.

CASE REPORT

A 65-year-old male patient complained of progressive right nasal obstruction and facial pain which had been present for one year. Systemic examination of the patient was normal and his medical history was unremarkable. He did not mention epistaxis among his complaints. Anterior rhinoscopic and rigid nasal endoscopic examination showed a large polypoid lesion in the right nasal cavity which did not bleed easily with manipulation. Endoscopic examination of the left nasal cavity was normal. Paranasal computed tomography (CT) demonstrated a soft tissue density filling the right nasal cavity. The mass was

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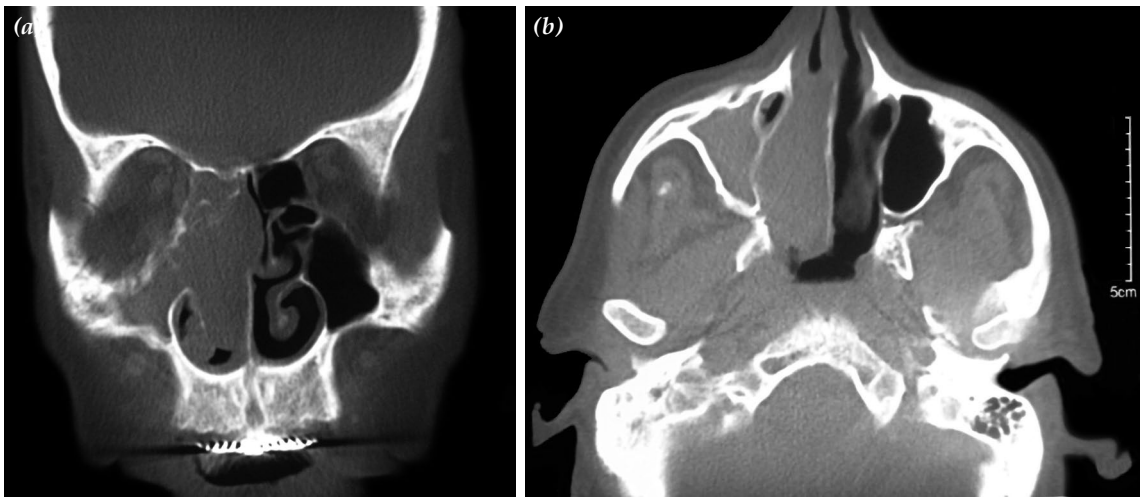

Figure 1. *Paranasal sinus computed tomography image:* ***(a)*** *Coronal section.* ***(b)*** *Axial section. The right maxillary sinus and the frontal sinus showed no aeration and demonstrated a soft tissue density filling the right nasal cavity.*

expanding towards the lateral wall of the middle concha. The right maxillary sinus and the frontal sinus showed no aeration. There was no defect in the neighboring bony structures and the left paranasal sinuses were normal (Figure 1a, b). The pathological diagnosis of SHPCL tumor was made after a punch biopsy of the mass. Under general anesthesia, the mass was excised, the middle concha was partially resected, uncinectomy was performed and the maxillary antrum, frontal recess and sphenoid ostium were widened with functional endoscopic sinus surgery. We saw that the mass showed minimal invasion into the sphenoid sinus (Figure 2a). Purulent contents of the maxillary and frontal sinuses were discharged (Figure 2b). There was excessive bleeding during the surgery, which the surgeon had a hard time stopping. Postoperatively, Merocel® (Medtronic. Xomed, FL, USA) buffer was placed into the intranasal cavity and removed two days after the surgery (Figure 3). Additional bleeding was observed after removal of the buffer.

Histopathology

Histopathologically, sections showed a tumoral formation with hypercellular areas under the respiratory epithelium and hypocellular loose myxoid areas in between. The tumor generally had mesencymal cells that had indistinguishable cytoplasm, oval/round nuclei, small nucleoli with a pale chromatin and no significant atypia. There was generally a diffuse, and less

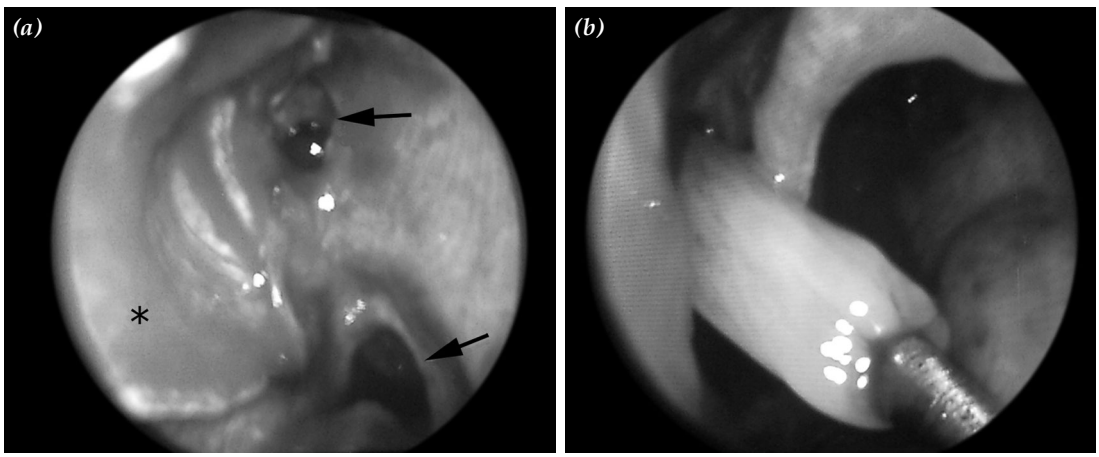

Figure 2. *Endoscopic appearance of nasal passage after extraction of the mass.* ***(a)*** ** Middle concha which was expanding towards the lateral nasal wall. Upper arrow: sphenoid sinus ostium. Lower arrow: choana and nasopharynx.* ***(b)*** *Purulent discharge in the maxillary sinus.*

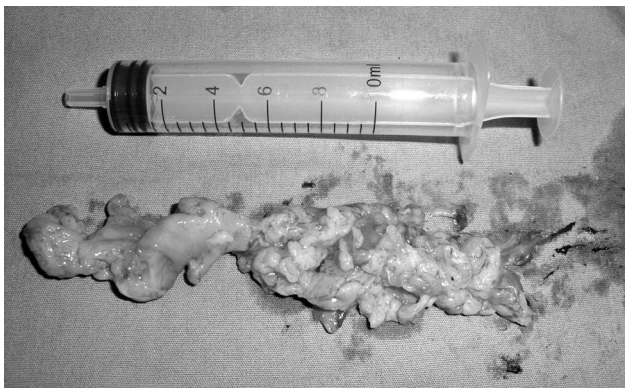

Figure 3. *Specimens tissue obtained by after the resection.*

frequently a short fascicular pattern. There was no necrosis or prominent mitotic activity. The ground showed abundant mast cells and a rich network of thin vessels some of which were in the form of dilated vessels with perivascular hyalinization (Figure 4a, b). Reticulin staining did not show an organoid structuring. Periodic Acid-Schiff (PAS) and mucicarmine stains did not demonstrate a mucinous accumulation. On immunohistochemical staining there was diffuse positivity of alpha-smooth muscle actin (SMA) and vimentin, and a Ki67 index maximum of 5%. The high-molecular-weight kininogen (HMWK), low molecular weight cytokeratin (LMWK), HMB-45, the S-100, desmin, epithelial membrane antigen (EMA), CD34, CD68, lysozyme, CD99 and CD56 stainings were negative. These findings mostly pointed to a diagnosis of HPCL tumor with most probably a low malignant potential. The postoperative course of the patient within the six months following surgery was good with no local recurrence or distant metastasis.

DISCUSSION

Hemangiopericytoma (HPC) is an uncommon vascular tumor which arises from pericytes surrounding the capillaries.[1] It was first described by Stout and Murray in 1942.[2] It can occur in various parts of the body but it is quite rare in the nasal cavity. This mesenchymal tumor is characterized by a prominent perivascular growth pattern.[4] Sinonasal hemangiopericytoma like tumor is seen in the sinonasal area and is characterized by a perivascular growth pattern and myoid differentiation. In general, it appears as a soft tissue mass and could show benign or malignant histologic features.[3] The characteristic clinical course, location, biologic behavior, and histologic pattern of intranasal HPCL tumors were described, as were the differential diagnostic considerations. The importance of recognizing this tumor, as a special variant of HPC because of its considerably more silent prognosis is emphasized.[5] Although certain pathologic factors may correlate there is no specific diagnostic test.

It may be necessary to differentiate SHPCL tumors from similar lesions such as solitary fibrous tumors of soft tissue and conventional HPC. Solitary fibrous tumor is mainly composed of spindle cells, and frequently exhibits a striking pericytic vascular pattern, mimicking SHPCL. In solitary fibrous tumors intercellular collagen and CD34 are positive and can be differentiated from SHPCL tumors by this property. Conventional HPC occurs in different areas of the body, and some have coined the term "hemangiopericytoma-like tumor" for lesions in the sinonasal region that demonstrate minimal cellular pleomorphism

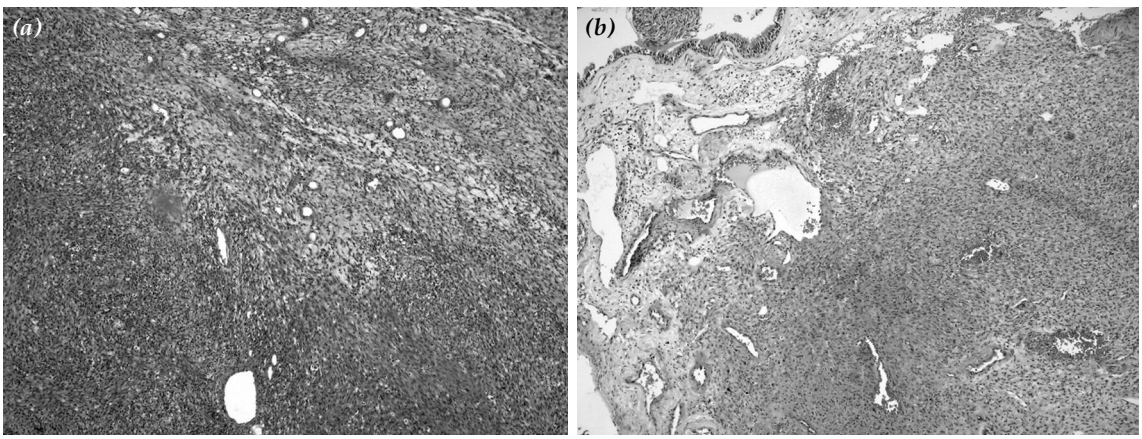

Figure 4. ***(a)*** *On the surface under the airway epithelium spindle cell and vascular proliferation (H-E x 100).* ***(b)*** *Composed of spindle cells in tumor development (H-E x 100).*

and minimal mitotic activity. It is not high-grade malignancies and rare local recurrence or distant metastasis.[1] Some authors group this tumor with soft tissue HPC's. On the other hand, other authors suggest that SHPCL tumor is biologically similar to or the same as the glomus tumor.[6,7]

Tse and Chan[3] propose that SHPCL tumors are biologically a variant of the glomus tumor since these two tumor types have similarities in their biological behavior, cytological profiles and immunophenotypes. Both the intranasal glomus tumor and SHPCL tumor are characterized by a perivascular growth pattern and myoid differentiation, having a close relation to the 'perivascular myomas', which was recently designated.[3]

Sinonasal hemangiopericytoma like tumor is rarely identified under the age of 30. It frequently affects adult patients in the sixth and seventh decade of life. It has no sex tendency. The most common symptoms are nasal obstruction and epistaxis.[6,8] It is slow growing, which can be the reason for its association with advancing age. In our case, the main complaint of the patient was progressive right nasal obstruction and facial pain which had been present for one year. He had no epistaxis. But there was excessive bleeding during the surgery. Therefore we think preoperative embolization may have provided a better surgical result.

Most patients may complain of nasal stuffiness or epistaxis. Clinically, this condition is most often interpreted as nasal polyps and hemangiomas. Sinonasal hemangiopericytoma like tumor generally shows a lower malignant potential than conventional soft tissue HPC. The choice of treatment of these tumors is surgical removal. Wide local excision is usually curative but the tumor may show a local recurrence if it is not totally excised. The recurrence rate is approximately 7% to 40% and metastasis almost never reported.[9]

Surgical extirpation with lateral rhinotomy or endonasal endoscopic approach is the most commonly used method. But infratemporal, anterior and middle cranial fossa approaches have been used for skull base and cribriform plate HPC.[10-12] Other treatment modalities, for example chemotherapy and radiation therapy have been used very rarely.[13]

In a series of five patients with SNHPL tumor, all cases were operated with wide local excision. In only one case was there active mitosis, and postoperative radiotherapy was applied to this patient. No recurrence was seen during 13 to 120 months of follow-up.[14]

We preferred total mass exision with endoscopic endonasal procedure under general anesthesia. The postoperative course of the patient was good with no local recurrence during the six months after the surgery.

In patients with complaints of nasal obstruction and nasal bleeding, especially in the advanced age group, intranasal rare tumors should be kept in mind. Detailed endoscopic, pathologic and radiological examinations should be performed for diagnosis and appropriate treatments should be planned.

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