Sinonazal Hemanjioperisitoma

Sinonasal Hemangiopericytoma

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ABSTRACT

Hemangiopericytoma is a rare mesenchymal tumor comprising about 1% of all vascular tumors. Fifteen to thirty percent of all hemangiopericytomas are encountered in the head and neck region, and 15% of those originate from the sinonasal area. They are believed to originate from the pericytes, the extravascular cells surrounding the blood vessels, and 5-10% of them show distant metastases. A sinonasal hemangiopericytoma was excised from a 51-year-old patient complaining of nasal obstruction and nose bleeding by medial maxillectomy. Twenty one months later, a recurrent mass at the level of the anterior sphenoidal wall was endoscopically excised. No recurrence or relapse was noticed during the next 22 months of the follow up. Sinonasal hemangiopericytoma is a rare vascular tumor which is treated surgically and should be followed up due to its potential of late recurrences and metastases.

Keywords

Hemangiopericytoma; paranasal sinus neoplasms

ÖZET

Hemanjioperisitoma tüm vasküler tümörlerin yaklaşık %1'ini oluşturan nadir bir mezenkimal tümördür. Hemanjioperisitomaların %15-30'u baş-boyun bölgesinde görülürken, bunların %15'i sinonazal bölgeden köken alır. Burun tıkanıklığı ve burun kanaması şikayeti ile polikliniğimize başvuran 51 yaşındaki bayan hastaya medial maksillektomi yapılarak sinonazal hemanjioperisitoma eksizyonu yapıldı. Takiplerinde ameliyat sonrası 21. ayında sfenoid sinüs ön duvarı hizasında lokal nüks saptanan hastanın kitlesi endoskopik olarak çıkarıldı. Hastanın daha sonraki 22 aylık takibinde nüks veya rekürrens saptanmadı. Sinonazal hemanjioperisitomalar; cerrahi olarak tedavi edilebilen, geç dönemde rekürrens ve metastaz potansiyeli nedeniyle uzun dönem takibin gerekli olduğu nadir vasküler tümörlerdir.

Anahtar Sözcükler

Hemanjioperisitoma; paranazal sinüs neoplazmları

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INTRODUCTION

Hemangiopericytoma (HP) is a rare mesenchymal tumor and was initially described by Staut and Murray in 1942.[1] HPs are believed to originate from the extravascular pericytes which are modified smooth muscle cells capable of contraction.[2,3] HPs comprise 3-5% of all soft tissue sarcomas and 1% of all vascular tumors.[4]

Trauma history and long-term steroid use were accused in the etiology.[5] They may be encountered in all age groups with no sex predilection, but mostly in the 5th and 6th decades of life.[6] The great majority of them are located specifically in the lower extremities and 15-30% are in the head and neck region. Of those in the head and neck region 15% are located in the sinonasal region, and 5-10% of those show evidence of distant metastases.[4,6,7-9] Marianovsky et al.[10] performed a literature research and reported 135 cases of sinonasal region HPs. The present study describes a rare sinonasal HP case.

CASE REPORT

A 51-year-old female presented to the outpatient ENT clinic of our hospital with the complaints of nasal obstruction and nose bleeding for two months. The physical examination revealed a bleeding polypoid mass completely filling the right nasal passage. The paranasal computed tomography imaging revealed a mass occupying the whole right nasal passage (Figure 1). A biopsy was obtained and the histopathological diagnosis was reported as "hemangiopericytoma". The patient underwent a medial maxillectomy by a lateral rhinotomy approach, and the histopathological examination of the specimen was reported to as "low-grade hemangiopericytoma". The control follow up visit 21 months later revealed a local recurrence in the anterior sphenoid sinus wall. The limited lesion was endoscopically excised. The patient is followed up for 22 months and no recurrence or relapse is noted (Figure 2).

DISCUSSION

HPs can be encountered anywhere in the body where capillaries are present, and symptoms are relevant to location. They can be encountered in both nasal cavities and in both sexes with no predilection.[3] The tumor typically grow slowly through the years and symptoms appear rather late. The most frequent symptoms of sinonasal HPs are nasal obstruction and nose bleeding.[3] Similarly, our patient presented with the complaints of nasal obstruction and nose bleeding.

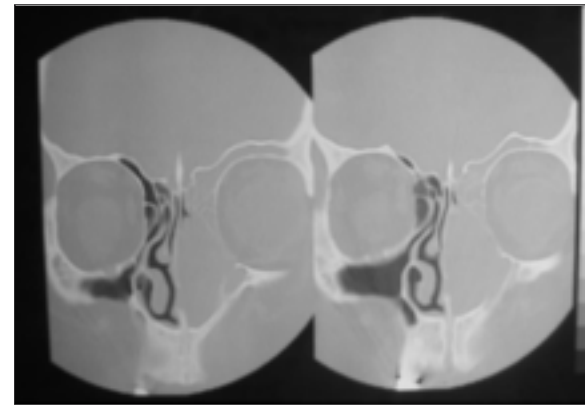

Figure 1. Computed tomography image showing soft tissue density in the right nasal cavity and sinuses.

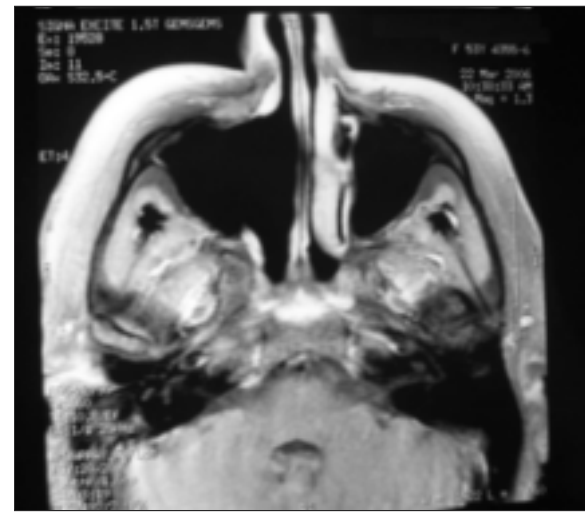

Figure 2. The magnetic resonance imaging of the right nasal cavity showing the postoperative cavity.

Histopathologically, HPs are non necrotic tumors which include staghorn-like vascular structures with uniform or spindle shaped cells showing rarely mitotic activity. Histologically, tumors such as haemangioendothelioma, glomus tumor, vascular leiomyoma, Kaposi's sarcoma, vascular meningioma, sclerosing haemangioma, post therapeutic malignant melanoma, neurilemmoma, paraganglioma, and vascular portions of fibrosarcomas should be considered in the differential diagnosis.[11,12] Thus, in addition to the silver reticulin dye, other immunohistochemical stains should be used in the diagnostic work up.[1,2,13] HPs are classified as benign, borderline, low-grade, and malignant depending on the mitotic activity observed, but this differentiation

is not correlated with the local recurrence and metastasis rates.[13] The tumor in our case was a low-grade HP with low level of mitotic activity.

The generally accepted therapy of HPs is total removal of the tumor with a wide surgical excision.[2,3,7,10,13] When the tumoral mass is large, embolization may be helpful prior to surgery.[10] In limited HPs endoscopic approach may be sufficient, while in cases where the tumor is big and its limits are indeterminable external approach is the procedure of choice.[10,13] In cases of cranial base and cribriform plate involvements a craniofacial approach may be required.[2] Local recurrences are usually due to insufficient surgical resection and are reported to be 8-53% in the literature.[5,8] The patients should be followed up lifelong because of very late local recurrences encountered.[2,7,9] The metastasis frequencies of the sinonasal regional HPs are quite low being 5-10% when compared to HPs of other regions.[5,10] Most frequent sites of metastasis are lungs and bones. Our patient underwent a lateral rhinotomy and the mass occupying the whole nasal cavity was totally excised. The recurrent limited lesion noticed after 21 months in the anterior sphenoidal wall was endoscopically resected and no recurrence was noticed in the follow up. The endoscopic surgical approach in local recurrences may prove to be appropriate because of the benign nature of the tumor course, but a close follow up would be beneficial, since distant metastases chances are suspected to be higher following local recurrences.[14] There is no significant study concerning radiotherapy as a therapeutic modality in HP cases, except a few case reports. Radiotherapy can be used as complimentary therapy in cases of incomplete surgical resection.[11,15] It may be used as a primary therapeutic modality in cases assumed as inoperable due to distant metastases, in cases where the tumor is too large and diffuse to be resected or in cases where an extremity amputation is not accepted.[11,15] Though chemotherapy is occasionally used in the treatment of HP, no precise technique is described.[16] Because the mass was totally removed in our case, no radiotherapy and/or chemotherapy was considered.

In conclusion, HPs are vascular tumors rarely encountered in the sinonasal region. Their appropriate treatment modality is wide surgical resection. The patients should be followed up for lifelong due to local recurrences and metastases though the sinonasal HPs are of more benign character. Patients with local recurrences following surgical removal should be monitored more closely because of the greater risk of metastases and local recurrences.

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