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Recurrent Sinonasal Hemangiopericytoma: A Rare Case Report

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Abstract

Hemangiopericytoma (HPC) is a rare vascular tumor arising from capillary pericytes. This tumor represents 3% - 5% of all soft tissue sarcomas and 1% of all vascular tumors. Only 15% - 30% of cases occur in the head and neck region, among which sinonasal hemangiopericytoma (HPC) is accounted for around 5% of cases. Hemangiopericytoma (HPC) has a tendency for recurrence. Here, a case of recurrent nasal hemangiopericytoma (HPC) after 12 years of treatment is reported.

Keywords

Hemangiopericytoma, Sinonasal Hemangiopericytoma, Endoscopic Approach, Staghorn Pattern

1. Introduction

Hemangiopericytoma (HPC) is a rare vascular tumor arising from capillary pericytes [1]-[8]. Hemangiopericytoma (HPC) was first described by Stout and Murray in 1942 [1]-[6]. It affects mainly middle-age adults but can affect all age groups with an equal sex distribution [1]-[7]. These tumors represent 3% - 5% of all soft tissue sarcomas and 1% of all vascular tumors [1] [2] [3] [4]. It tends to occur in all body parts [1] [2] [3] [4] [6] [8] but only 15% - 30% of cases occur in the head and neck area [1]-[8]. Sinonasal hemangiopericytoma (HPC) is accounted for around 5% of cases [1] [2] [3]. Symptoms of sinonasal HPC are nasal bleeding and obstruction. Local swelling, headache and visual symptoms suggest orbital, intracranial and cranial nerve involvement [1] [2] [3] [5] [7] [9]. Because these lesions tend to recur despite treatment [1] [2] [3] [4] [6] [7] [8] [9], any new symptoms in a patient diagnosed previously with sinonasal HPC should be taken into consideration [6].

2. Case Report

A 66-year-old male who is a known case of nasal hemangiopericytoma that was excised 12 years ago, and who missed regular follow-ups, presented with unilateral nasal bleeding for a couple of weeks. The bleeding was on the right side which was the previously operated side. Nasal examination showed a tan-colored swelling in the middle part of nasal septum (Figure 1). Paranasal sinuses CT scan was ordered. It showed a small well-defined mass in the right side of nasal septum around 1 × 1 cm, not extending into other regions of nasal cavity or paranasal sinuses (Figure 2). The lesion was completely excised by endoscopic surgery. Pathological examination of the lesion showed spindle cells surrounding dilated vessels suggestive of hemangiopericytoma. The patient was followed up for 1 year after the surgery and no recurrence was found. Given the nature of his tumor and its tendency to recur, he was advised for a lifelong follow-up.

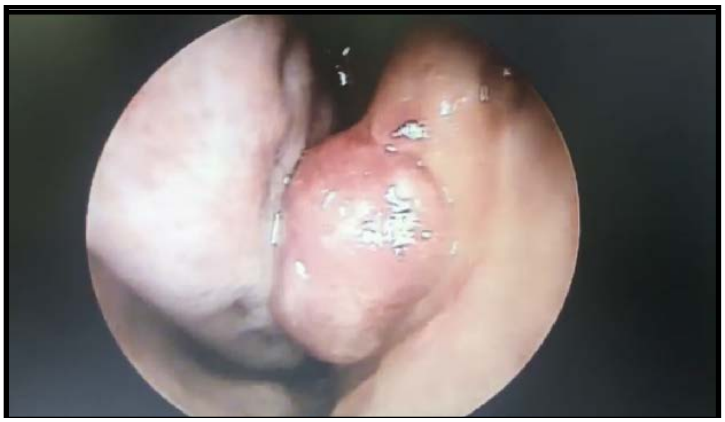

Figure 1. Endoscopic view of the of the right nasal cavity of the patient showing the lesion arising from nasal septum.

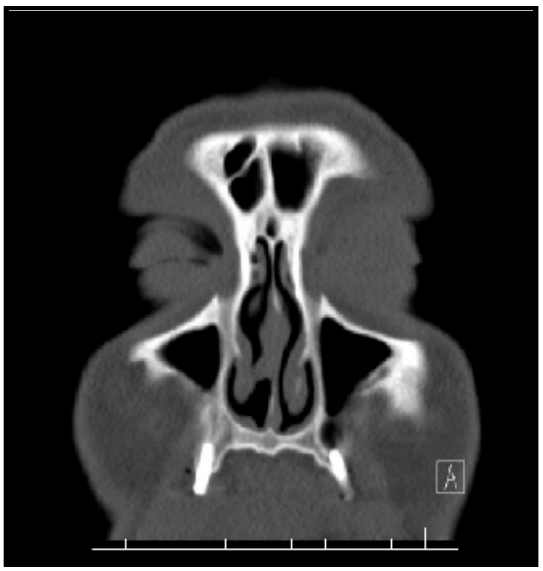

Figure 2. Coronal cut of paranasal sinuses showing the lesion arising from the right nasal septum.

3. Discussion

Hemangiopericytoma (HPC) is a rare vascular tumor arising from capillary pericytes [1]-[8]. These pericytes are modified smooth muscle cells surrounding capillaries and post-capillary venules [1] [2] [5] [9]. It represents 3% - 5% of all soft tissue sarcomas and 1% of all vascular tumors [1] [2] [3] [4]. HPC affects mainly middle-age adults but can arise in all age groups with an equal sex distribution [1]-[7]. The etiology remains largely unknown but trauma, previous steroid treatment, pregnancy and hypertension, are factors thought to be associated with the development of HPC by some [1] [2] [4] [5] [7]. Sinonasal HPC is extremely rare and it is accounted for around 5% of cases [1] [2] [3]. It tends to occur twice as common in the nasal cavity as in the paranasal sinuses [1] [2]. Sinonasal HPC behaves less aggressively than HPC arising from other body parts [1] [2] [5], however its behavior is unpredictable and it has both benign and malignant variants [3]-[7] [9]. It has a tendency for local recurrence of around 25% [3]. In some studies the local recurrence ranges from 8% to 53% [1] [5]. Local recurrence can be related to inadequate surgical resection particularly if occurring within one year of surgical resection [1] [2] [5] [8] [9]. Recurrence may arise decades later [1] [2] [3] [7] [9], which warrant a long-term follow-up [1] [2] [3] [7] [9]. Cheng *et al*. recommended working up for recurrence, any patient with a history of treated HPC that develops new symptoms regardless of the length of disease free survival [6].

Metastasis is less common [1] [3] [6] [7] [8] and maybe proceeded by multiple recurrences [1] [9]. The commonest metastasis is to the lungs [4].

Clinically, symptoms are mainly of nasal bleeding and obstruction. Local swelling, headache and visual symptoms suggest orbital, intracranial and cranial nerve involvement [1] [2] [3] [5] [7] [9]. On examination sinonasal HPC appears as a unilateral nasal mass, which could be firm, rubbery or soft with or without redness and can be as small as 1cm or as large as 20 cm [2] [3] [7].

Radiological imaging of paranasal sinuses can show the lesion. It appears as a unilateral soft tissue density that enhances with intravenous contrast on CT scan [1] [5] [7]. MRI would show an isointense lesion on T1and an iso- or hypo-intense lesion on T2 that enhances with gadolinium [1] [5].

Histopathology is the main stay diagnostic method [1] [3] [4] [5] [7] with the aid of immunohistochemical staining. Histologically, these tumors show tightly packed spindle-shaped cells with little stroma, surrounding dilated vascular channels, forming a characteristic staghorn pattern [1] [2] [3] [4] [6] [7] [8] [9]. Immunohistochemical staining are usually required to assist in the diagnosis [1] [3] [4] [5] [6] [8] [9] particularly reticulin stain [1] [2] [3] [4].

Sinonasal HPC is largely treated by complete surgical resection with a wide margin [1] [3] [5] [7] [9]. Negative margins are usually difficult to achieve in the sinonasal area especially for large tumors [1] [8]. Small-sized sinonasal HPC can be removed efficiently by endoscopic approach [1]. Endoscopic resection is currently the best approach, as endoscopy would avoid external scaring and localize

the tumor more precisely. It would preserve the physiology of the nasal mucosa better than external approach, with the advantage of less blood loss and less chance of damaging the nasolacrimal system [5]. Large tumors tend to require open surgery and may require preoperative angiographic embolization to decrease the chance of excessive intraoperative bleeding [2] [5] [7] [9]. For unclear margins and recurrent tumors, radiotherapy is advocated by some [2] [8] [9], though sinonasal HPC is generally considered radioresistant [2] [5] [7] [8]. Chemotherapy is usually used for metastatic HPCs [2] [5] [9]. The role of radiotherapy and chemotherapy alone or combined as initial treatment is not yet clear [1] [2] [8]. Because of the tendency of HPCs to recur years after the initial treatment, a lifelong follow up is a must [1] [2] [3] [5] [6] [7].

4. Conclusion

Though sinonasal HPC is a rare condition, it should be always kept in the differential diagnosis of a sinonasal mass presenting with bleeding or nasal obstruction especially in a patient who is known to have HPC treated previously. Sinonasal HPC is a lesion with a high tendency for recurrence; hence it requires a lifelong follow up.

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Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

Informed Consent

Verbal consent was obtained from the patient for publication of this manuscript and accompanying images.

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