Endoscopic Transnasal Resection of Sinonasal Hemangiopericytoma: A Report of Two Cases

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Abstract

Background: Hemangiopericytoma is an uncommon vascular tumor derived from the pericytes of Zimmermann, representing only a small proportion of head and neck tumors, and mostly occurring in the sinonasal tract.

Case report: We report two cases of hemangiopericytoma of the sinonasal tract in a 78-year-old woman and a 77-year-old man, both presenting with symptoms of frequent epistaxis. In Case 1, a purple tumor was identified originating from the posterior olfactory fissure in the left nasal cavity. To reduce bleeding from the tumor, the anterior and posterior ethmoidal arteries were coagulated via a transconjunctival incision approach prior to tumor removal. The tumor in Case 2 was arising from the posterior region of the right nasal septum. In Case 2, intensive bleeding was seen during biopsy. To reduce the bleeding and surgical time for the subsequent operation, preoperative angiography and embolization were performed. We removed the tumors en bloc in both cases by endoscopic operation.

Results: In these cases, postoperative histopathological and immunohistochemical examinations confirmed sinonasal hemangiopericytoma. Endoscopic and radiological follow-ups showed no recurrences within 5 years.

Conclusion: Sinonasal hemangiopericytomas were diagnosed histologically. Immunohistochemical analyses were helpful for tumor diagnosis. Herein we discuss the diagnosis and treatment of hemangiopericytoma.

Keywords
SINonasal hemangiopericytoma, Epistaxis, Immunohistochemical examination

Introduction

Hemangiopericytomas are unusual vascular tumors that were first described by Stout and Murray in 1942 [1]. These tumors originate from extravascular cells called pericytes, which presumably represent modified contractile smooth muscle cells found on the external surface of capillaries and post-capillary venules.

Approximately 15-30% of hemangiopericytomas occur in the head and neck. Of these, approximately 5% arise from the nasal cavity. Sinonasal hemangiopericytomas are believed to behave less aggressively than hemangiopericytomas that occur in other parts of the body.

Hemangiopericytomas are relatively radioresistant and show a major tendency to bleed profusely, so wide surgical resection under an open approach and preoperative embolization are the main management options for large sinonasal tumors. Endoscopic resection of sinonasal hemangiopericytoma is a novel method with special advantages. We report two cases of hemangiopericytoma of the sinonasal tract in a 78-year-old woman and a 77-year-old man, both of whom presented with symptoms of frequent epistaxis.

Case Descriptions

Case 1

A tumor in the left nasal cavity was detected in a 78-year-old woman on gastro fibrescopy at an internal medicine clinic. She was referred to our hospital because of frequent epistaxis after the examination. Nasal
hemangiopericytoma (Figure 3 and Figure 4). The patient had an uneventful recovery and no recurrence has been seen as of 5 years postoperatively.

Case 2

A 77-year-old man was referred to our hospital because of frequent epistaxis. Nasal endoscopy showed a reddish tumor arising from the posterior region of the right nasal septum (Figure 5a). CT revealed a well-marginated soft tissue mass in the posterior region of the olfactory cleft (Figure 5b). During biopsy, intensive bleeding was seen. Histological examination of the tumor identified hemangiopericytoma. To reduce bleeding during the subsequent operation, preoperative embolization of the tumor was carried out (Figure 6), and transnasal endoscopic surgery was performed with complete tumor resection the next day. The amount of bleeding at the time of tumor removal was also very small (volume of blood loss is 10 cc). As of 5 years postoperatively, the patient remains asymptomatic and has shown no signs of recurrence.

Discussion

Sinonasal hemangiopericytomas are low-grade, ma-
lignant vascular soft tissue tumors that can arise anywhere in the body, slowly invading into surrounding tissues. Generally, these tumors are considered to have malignant potential with late recurrence rates reportedly within the range of 8-53% [2-6] and metastasis occurring in 12-60% of cases [5,6]. Complete tumor removal is thus very important, because local recurrences tend to show metastasis.

**Figure 3:** Histochemical staining.
a) Excised tumor specimen; b) Spindle-shaped tumor cells with rounded nuclei are seen growing in sheets and whorls around vascular channels. Poor cellular atypia and no mitotic figures in tumor cells (HE, ×100).

**Figure 4:** Immunohistochemical staining.
Tumor cells appear strongly positive for factor XIIIa (a) and mildly positive for HHF35 (b).
The best visualization of the vessel supply for sinonasal hemangiopericytoma can be achieved by conventional digital angiography, which also helps to plan preoperative embolization [8].

Even though the final diagnosis is obtained from histopathological examination, several authors have suggested [9-12] avoiding biopsies if the tumor size suggests a risk of severe bleeding [2]. However, misdiagnosis can lead to an incorrect surgical approach, with incomplete excision, which may favor recurrences or metastases. Preoperative tissue biopsy is thus warranted whenever possible, while preparing for bleeding. In cases where biopsy is not possible before surgery, frozen section
analysis should be performed intraoperatively to determine the suitable range for tumor resection.

Morphologically, hemangiopericytoma is very similar to a wide range of soft tissue tumors. Differential diagnoses include solitary fibrous tumor, myopericytoma, glomus tumor, perivascular epithelioid cell neoplasm, extra-gastrointestinal stromal tumor, and synovial sarcoma [13]. Histochemical and immunohistochemical techniques are essential, mainly to exclude morphologically similar soft tissue tumors. No specific immunohistochemical markers for sinonasal-type hemangiopericytoma have been identified to date.

However, the tumor cells in hemangiopericytoma are typically positive for vimentin (98% of specimens) and smooth muscle act in (92%) [14]. Positive staining for cytokeratin excludes a diagnosis of hemangiopericytoma. Intimate relationships between hemangiopericytoma and solitary fibrous tumor have been reported. Differentiating between these tumors is important, because most solitary fibrous tumors of the soft tissues are benign. CD34 is an extremely useful marker for this purpose [15,16].

Preoperative angiography is very useful for identifying whether the tumor has a large blood supply and judging the necessity of preoperative embolization of the tumor. In Case 1, preoperative embolization was not performed because the base of the tumor was limited and the feeder vessel to the tumor was thought to be a branch of the internal jugular artery. However, preoperative embolization is needed in cases of very large lesions or those showing intracranial lesions to reduce blood loss intraoperatively and tumor size. However, depending on the facility, angiography and embolization may be difficult. Angiography itself also carries a risk of complications such as cerebrovascular accident [17,18]. In cases where angiography cannot be enforced, the amount of bleeding may be able to be reduced by cauterizing blood vessels that seem to be feeding the tumor during surgery. If the anterior or posterior ethmoidal arteries are considered to be the feeding vessel, coagulation of the vessel can be performed under the canopy of the ethmoid sinus or within the orbit. For posterior tumors, coagulation of the sphenopalatine artery at the sphenopalatine foramen may reduce bleeding from the tumor to some extent.

Due to its latent potential for malignancy, wide local excision is still considered the gold-standard treatment for sinonasal hemangiopericytoma [19]. For tumors in paranasal sinuses, an external procedure is most frequently chosen to achieve total tumor excision. In particular, a craniofacial approach is necessary when the cribiform plate or base of the skull is breached, but tumor located purely intranasally or strictly within the ethmoid or sphenoid sinus can be removed via an endonasal approach. We were able to remove both tumors en bloc in endoscopic operations leaving behind bone of the cribiform plate.

No complications such as leakage of cerebrospinal fluid (CSF) were seen in either case. However, endoscopic skull base surgery must be performed after tumor resection at the time of CSF leaks during the operation. Most recurrences are probably the result of inadequate surgical excision. Because recurrence can develop years after treatment, lifelong follow-up is necessary [2-4].

Conclusion

We encountered two cases of sinonasal hemangiopericytoma. We removed the tumors en bloc in endoscopic operations. Prior to tumor resection, the anterior and posterior ethmoidal arteries were cut via a transconjunctival incision in Case 1, and preoperative angiography and embolization were performed in Case 2 to reduce both surgical time and blood loss.

Tumor excision must be complete due to the latent potential for malignant behavior of these tumors. This may be achieved using an endonasal endoscopic approach. If this approach is not possible, an external approach is needed and can potentially be combined with endonasal endoscopic control. No recurrences have been found so far in either of our patients, but recurrence can develop years after treatment, so lifelong follow-up will be continued.

Conflict of Interest

The authors declare no potential conflicts of interest.

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Authors’ Contributions

Dr. Arimoto (Area of interest: Sleep medicine), Dr. You and Dr. Kawade (Area of interest: Rhinology) consulted on treatment methods as part of the same nasal sinus group, and performed the surgery together. Dr. Kishimoto (Area of interest: Audiology), Dr. Uchida (Area of interest: Audiology), Dr. Ogawa, and Dr. Hujimoto (Area of interest: Speech and language) were asked to read and critique the paper and identify points for improvement. All authors have read and approved submission of the manuscript.

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