Endoscopic Management of Sinonasal Hemangiopericytoma

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Abstract

Objective. Sinonasal hemangiopericytomas (SNHPCs) are rare perivascular tumors with low-grade malignant potential. Traditionally, these tumors have been treated with open approaches such as lateral rhinotomy, Caldwell-Luc, or transfacial approaches. Increased experience with endoscopic management of benign and malignant sinonasal tumors has led to a shift in management of SNHPC. The authors present their experience in the largest series of patients with SNHPC managed endoscopically.

Study Design and Setting. Case series at a tertiary care medical center.

Subjects and Method. A retrospective chart review of all patients undergoing endoscopic management of SNHPC at the University of Miami between 1999 and 2008 was conducted. All endoscopic resections were performed with curative intent.

Results. Twelve patients with the diagnosis of SNHPC were treated endoscopically. Mean age was 62.5 years (range, 51-83 years). There were 6 men and 6 women. The mean follow-up was 41 months (range, 15-91 months). Seven (58.3%) presented with nasal obstruction, whereas 4 (41.6%) had epistaxis as their initial presenting symptom. Preoperative angiography or embolization was not performed in any case. Mean estimated blood loss was 630 mL (range, 100-1500 mL). Six patients underwent endonasal endoscopic anterior skull base resection; 4 had complete endoscopic resection all with negative margins. None underwent postoperative adjuvant treatment. No recurrence or metastatic disease was observed in this patient population.

Conclusion. Endoscopic management of SNHPC is a feasible approach and did not compromise outcomes in this experience. In this series, familiarity with advance endoscopic sinus surgery was necessary to manage these patients. Postoperative adjuvant therapy was not necessary in this cohort.

Keywords

hemangiopericytoma, sinonasal tumors, epistaxis, sinonasal hemangiopericytoma, malignant sinonasal tumors, endoscopic sinus surgery, endoscopic skull base surgery, perivascular tumors, anterior skull base resection

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inonasal hemangiopericytomas (SNHPCs) are low-grade malignant vascular soft tissue tumors that can arise anywhere in the body. They make up approximately 2% to 3% of soft tissue sarcomas. Of that, an estimated 5% arise in the nasal cavity and paranasal sinuses. Males and females are equally affected and are usually seen in the third and fifth decades of life. In patients diagnosed with SNHPC, the most common presenting sign and symptom are epistaxis and nasal obstruction.1,2

As first described by Stout and Murray3 in 1942, SNHPCs are vascular tumors that arise from pericytes of Zimmermann. Pathologists describe a classic histologic staghorn pattern on low-power microscopic field. These patterns are vascular channels that are seen in almost all specimens.1,2

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Previously, treatment of SNHPCs relied on wide surgical excision through open craniofacial approaches. However, in the past decade, with the advent of endoscopic equipment and the evolution of specialized endoscopic skull base surgeons, a progression toward less invasive approaches has emerged. Several studies have shown that a solely endoscopic approach is feasible. This report is the largest series of a single institution’s experience with endonasal endoscopic management of SNHPC.

Materials and Methods

A retrospective chart review was performed on all patients with the histological diagnosis of SNHPC who were resected endoscopically from October 1999 to December 2008 at the University of Miami (Figure 1). Information regarding age, gender, presenting symptom, preoperative management, intraoperative complications, type of endoscopic surgery, intraoperative blood loss, and clinical follow-up was collected. All patients were evaluated with a computed tomography (CT) scan of the head, neck, and chest to rule out distal metastasis prior to curative surgical resection. No patients were found to have distal metastasis in our patient population.

The study population was limited to patients who underwent endoscopic tumor resection with curative intent. Preoperative biopsies were not performed as part of the workup because of the vascular appearance of the tumors. Endoscopic resection was performed after maxillary antrostomy, complete sphenethmoidectomy, frontal sinusotomy, or septectomy as necessary to gain complete visualization of the tumors’ periphery. Extended procedures, including modified Lothrop (ML) and extended sphenoidotomy (ES) involving bilateral wide sphenoidotomy with posterior septectomy and intersinus septum resection, were performed for patients who had tumors involving the cribriform plate, sphenethmoid recess, or frontal recess.

Tumor debulking was performed to identify the area where the tumor was pedicled (“epicenter” of the tumor). Wide endoscopic local resection of surrounding tissue was performed and the underlying bone was drilled or removed if involved. Frozen section analysis of soft tissue margins was performed to ensure the margins were clear of residual tumor. For tumors involving the cribriform plate or surrounding anterior skull base, a hemi or complete anterior skull base resection and reconstruction using acellular dermal allograft was performed. This reconstruction technique was described in detail by Germani et al in 2007. The protocol for this study was reviewed and approved by the institutional review board of the University of Miami, Miami, Florida.

Results

The study population included 12 patients with the histological diagnosis of SNHPC from the University of Miami.
Endoscopic Tumor Resection Database. There were 6 men and 6 women. The patients’ age ranged from 51 to 83 years with an average of 62.6 years. Seven (58.3%) patients presented with nasal obstruction, whereas 5 (41.7%) patients had epistaxis as their initial presenting symptom. Six (50%) patients had tumors involving the cribriform plate and underwent endonasal endoscopic anterior skull base resection and reconstruction. All patients who underwent endonasal endoscopic anterior skull base resection had large dural defects with high-flow cerebrospinal fluid (CSF) leaks that were repaired with acellular dermal allograft. One (8%) patient needed a modified Lothrop procedure to clear tumor margins in the frontal recess. Three (25%) patients needed extended sphenoid sinusotomy to adequately resect the posterior margin, and the remaining 2 (17%) patients underwent wide endoscopic resection with negative margins. The average blood loss was 630 mL (range, 100-1500 mL) with none of the patients having undergone preoperative embolization. All the patients underwent complete resection of the tumor without the need for transfusion. All tumors were resected with at least a 3-mm margin around the lesion and 5 mm when feasible. The mean operative time was 175.5 minutes (range, 64-285 minutes). There were no minor or major complications such as need for reoperation, postoperative CSF leak, meningitis, blood transfusion, prolonged hospital stay, epistaxis, stroke, or death.

There was no local tumor recurrence or distant metastatic disease with a mean follow-up of 41 months (range, 15-91 months). None of the patients received adjuvant postoperative therapy. All patients were examined frequently in the first 3 postoperative months until the sinonasal cavity had completely healed. Regular postoperative endoscopic surveys were scheduled every 3 months for the first postoperative year, every 6 months for the second year, and annually thereafter.

Discussion

Hemangiopericytomas commonly arise in the musculoskeletal system, the skin, and the retroperitoneum. In the head and neck, these tumors most commonly involve the soft tissue of the scalp, face, or neck. Sinonasal hemangiopericytomas are exceedingly rare but usually involve the ethmoid and sphenoid sinuses. There is no known sex or racial predominance, and most cases occur in patients’ third to fifth decades. However, there have been reports of a slight increase in the incidence of SNHPC in the sixth and seventh decades. Because of the indolent nature of these tumors, patients with SNHPC present with nasal obstruction or bleeding. Compressive orbital findings of proptosis, epiphora, and diplopia are rare but can be found with increased tumor size or orbital extension.

Radiographically, SNHPCs appear as soft tissue masses with locally destructive behavior. On CT scan, they show significant enhancement with contrast administration (Figure 1) secondary to the numerous vascular channels present in these tumors. On magnetic resonance imaging (MRI), SNHPCs become hyperintense after gadolinium administration. They also often exhibit flow-void as a sign of increased vascularity. Traditionally, surgical treatment of SNHPCs has been similar to that of benign sinonasal tumors such as inverted papilloma using a lateral rhinotomy or midface degloving to perform wide local resection with negative margins. Postoperative adjuvant therapy has been used when tumor size and extension limited complete resection. However, SNHPCs are histologically malignant tumors with metastatic potential, and incomplete resection has led to reports of a recurrence rate of 9% to 50%.

Endoscopic surgery of malignant sinonasal tumors has gained acceptance as long-term results of tumor control and survival have become available showing excellent functional outcomes and better quality of life with comparable disease-free survival. However, most of these studies have within them a heterogenic tumor population with varied clinical courses, making it difficult to discern the effectiveness of endoscopic resection for a given tumor type.

This study reports the results of the largest series of patients with SNHPC treated completely with endoscopic surgery. It is important to note that 6 (50%) of the 12 patients had disease involving the cribriform plate, necessitating endoscopic skull base resection and reconstruction to obtain negative margins. These cases resulted in large skull base and dural defects with high-flow CSF leaks that were repaired with acellular dermal allograft. Although prior studies have reported high recurrence rates, there has been no local recurrence or distant metastatic disease in our study population after a mean follow-up of 41 months.

Important limitations of the endoscopic technique must also be factored into the decision-making process. Familiarity with advanced endoscopic surgical techniques is necessary to manage patients with SNHPCs to achieve complete tumor resection. A graduated and incremental approach to complex endoscopic cases may improve outcomes and decrease surgical morbidity. Furthermore, these results, although promising, should be evaluated in the context of a retrospective study with its inherent limitations, a small sample size, and the lack of a control group.

Conclusion

Sinonasal hemangiopericytomas can be successfully managed endoscopically. The indolent nature of these malignant tumors lends itself to endoscopic resection and surveillance. Safe and complete resection can be performed by an experienced endoscopic surgeon with minimal morbidity. Adjuvant therapy was not necessary in our group where complete resection was achieved with negative margins. Long-term scheduled endoscopic surveillance is necessary to diagnose and manage recurrence.

Author Contributions

Belachew Tessema, conception and design, acquisition of data, analysis and interpretation of data, drafting the article, approval of the version to be published; Jean Anderson Eloy, conception and design, analysis and interpretation of data, revising the article.
critically for important intellectual content, approval of the version to be published; **Adam J. Folbe**, interpretation of data, revising the article for important intellectual content, approval of the version to be published; **Amy S. Anstead**, analysis and interpretation of data, revising the article for important intellectual content, approval of the version to be published; **Neena M. Mirani**, acquisition of data, analysis and interpretation of data, revising the article for important intellectual content, approval of the version to be published; **Deya N. Joudy**, acquisition of data, revising the article for important intellectual content, approval of the version to be published; **Jose W. Ruiz**, interpretation of data, revising the article for important intellectual content, approval of the version to be published; **Roy R. Casiano**, substantial contributions to conception and design, analysis and interpretation of data, revising the article for important intellectual content, approval of the version to be published.

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**References**

Corrigendum


The sixth author’s name was submitted as Deya N. Joudy. The correct spelling is Deya N. Jourdy.