CASE REPORT

Sinonasal Hemangiopericytoma: Case Report and Literature Review

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Abstract—Hemangiopericytoma (HPC) is a rare vascular tumor that can arise in any organ system, but occurs most frequently in skeletal muscle. We present a case of a primary sphenoid sinus HPC unusual for its spontaneous remission after biopsy. There have been approximately 55 cases of sinonasal HPC reported in the literature, representing less than 5% of all lesions. In general, HPC behaves aggressively, demonstrating greater than 50% local recurrence and 10% metastatic disease. Although extended surgical resection is traditionally considered the most effective therapy for all HPC, critical literature review does not support this method of treatment for HPC occurring in the paranasal sinuses and skull base. Long-term follow-up indicates that the majority of sinonasal HPC have a benign clinical course regardless of treatment. As a result, we do not recommend extended resection as the initial therapy for sinonasal HPC. Instead, a period of observation coupled with serial MRI or CT scans should be used to detect tumor progression. These indolent tumors should undergo extended resection only after progression has been confirmed. In addition to the case report, a synopsis of the reviewed literature and a summary of treatment recommendations are also presented. (*Skull Base Surgery*, 5(3):185–190, 1995)

Hemangiopericytoma (HPC) is an unusual vascular tumor derived from pericytes.^{1,2} These cells can be found surrounding all capillaries and are thought to be immature smooth muscle derived from mesenchyme. Pericytes add structural support to the capillaries while playing a role in capillary blood flow regulation. Approximately 1% of all vasoformative neoplasms are HPC, with 15 to 25% presenting in the head and neck.^{3,4} Of those occurring in the head and neck, approximately 50% involve the paranasal sinuses and nasal cavity. Other sites of predilection are the soft tissues of the scalp, face, and neck, parotid gland, orbit, and rarely the salivary glands and larynx. Since HPC is derived from capillary pericytes, its theoretical site of origin can be virtually anywhere in the body. Indeed, HPC has been reported in a wide variety of tissues outside the head and neck, most frequently occurring in skeletal muscle and the retroperitoneum.^{3,5} As will be discussed in this report, the site of lesion has a significant

bearing on clinical behavior and perhaps on histologic character as well.

CASE REPORT

A 58-year-old Caucasian woman with a past medical history significant for coronary artery disease, hypertension, gout, follicular thyroid carcinoma, cutaneous melanoma, degenerative joint disease, and "chronic sinusitis" presented to another institution in April 1992 complaining of worsening "sinus headache," epistaxis, and left periorbital and malar pressure. Head and neck examination was remarkable only for a left posterior "rubbery" nasal mass. There was no cervical adenopathy, and cranial nerves were intact. Computed tomography (CT) scan in April 1992 revealed a mass in the left sphenoid sinus and magnetic resonance imaging (MRI) in May 1992 indi-

Figure 1. Low-power micrograph of the hemangiopericytoma of the nasal cavity. Microscopically, the hemangiopericytoma is a circumscribed, vascular neoplasm composed of a proliferation of spindle cells about numerous thin-walled endothelial-lined vascular spaces. These spaces may vary from gaping, sinusoidal channels to capillaries. (Hematoxylin & eosin stain; original magnification $\times 40$.)

cated additional left posterior ethmoid involvement. A biopsy of the mass was suggestive of glomus tumor and the patient was referred to our institution. The area was again imaged with both CT and MRI, which did not show substantial interval progression. The patient was taken to the operating room August 1992 for endoscopic examination and rebiopsy, which resulted in a subtotal excision of the mass. The biopsy revealed an HPC (Figs. 1, 2). Post-operative CT and MRI scans demonstrate residual tumor in the left sphenoid sinus (Figs. 3, 4). As the patient's general physical condition was poor, we elected to defer surgery and follow the lesion with serial physical examinations and CT and MRI scans. With more than 2 years of follow-up, there is no evidence of recurrent tumor. Furthermore, the residual tumor that had been present

following the subtotal excision is no longer detectable on physical or radiographic examination (Fig. 5).

HISTOLOGIC APPEARANCE

Under light microscopy, HPC is characterized by vascular channels ranging from compressed fingerlike projections to capillaries and sinusoids. The vascular channels may be unrecognizable before reticulin staining, which highlights the vascular nature of this lesion and demonstrates the tumor cells outside the vascular sheath. Around these channels, pericytes proliferate in varying degrees of differentiation. Because of the varying maturation of these cells, they can be difficult to distinguish from endo-

Figure 2. Higher power photomicrograph of Figure 1. The hemangiopericytoma is characterized by numerous capillary spaces surrounded by an accumulation of spindle cells. The tumor cells possess an oval or elongated nucleus with an indistinct cytoplasm. The organization of these cells is very haphazard, but the cellular confirmation is uniform. (Hematoxylin & eosin stain; original magnification $\times 200$.)

Figure 3. Axial computed tomography scan demonstrating soft tissue mass in left posterior sphenoid sinus (arrow). There is no evidence of bony erosion.

Figure 4. Axial magnetic resonance scan, T₁ weighted image with gadolinium contrast. Three months following subtotal excision, an enhancing left posterior sphenoid sinus mass is evident (arrow).

Figure 5. Axial magnetic resonance scan, T₁ weighted image with gadolinium contrast. Two years following subtotal excision of hemangio-pericytoma, there is no evidence of residual or recurrent tumor.

thelial cells, fibroblasts, and histiocytes. Marked variation in histologic appearance allows HPC to be misdiagnosed as a glomus tumor, angiosarcoma, fibrous histiocytoma, lymphoma, hemangioendothelioma, and schwannoma.^{6,7} Electron microscopy (EM) and immunohistochemical staining facilitate the histologic diagnosis of HPC as the pericytes in these tumors usually retain features sufficient to make them recognizable.^{8,9} Immunohistochemical staining demonstrates a strong staining pattern to vimentin, with approximately 20% of cases weakly positive to actin, and 10% positive to S-100¹⁰⁻¹²

With regard to histopathologic staging, HPC is generally divided into three groups: low, intermediate and high grade. Staging features include tumor cellularity, mitotic figures, nuclear atypia, and the presence of hemorrhage and necrosis.^{5,13}

- **Low grade:** These tumors have spindle-shaped pericytes with little crowding of vascular channels. There are few or no mitotic figures.
- **Intermediate:** These are more cellular tumors with plump pericytes and increased compression and crowding of vascular spaces. Mitotic figures are still not prominent.
- **High grade:** Characteristics include increase compression or obliteration of vascular spaces, increased anaplasia, and increased mitotic figures.

This classification provides reasonable predictive value when applied to all anatomic sites of tumor occurrence. This is particularly true with sinonasal disease in which histologic appearance and clinical course are consistently benign.^{3,12}

DISCUSSION

The biologic aggressiveness of HPC is well documented in the literature, as is the more benign course of those lesions occurring in the nasal cavity and paranasal sinuses.^{7,9,14,15} Differences in behavior between HPC of the sinonasal tract and other somatic sites is significant, and has led some investigators to consider these as two separate tumor types.⁷ In a review of 23 cases of sinonasal HPC, Compagno and Hyams⁷ found a surprising and uncharacteristic histologic homogeneity among the group. Along with benign histologic features, they reported very low rates of recurrence, metastasis, and mortality when compared with lesions from other sites. This series was conservatively reported as HPC-like tumors, possibly representing a distinct entity. Subsequent studies using EM and immunohistochemical techniques have confirmed that these tumors are, in fact, low-grade HPC and not a distinct pathologic entity.^{9,12} In another review of sinonasal HPC, Hughes and Bard¹⁴ reported a series of 15 cases over a 39-year period. They, too, note benign behavior and confirm the true pericyte origin of these tumors through light microscopy and immunohistochemical staining techniques. Interestingly, eight of these cases were previously reported by Gorenstein and Facer¹⁶ in 1978, thus adding 15 years of follow-up to the original report. From the Hughes and Bard data, three patients died from 1 to 12 years following the originally published series; however, no deaths were due to HPC or its sequelae. Combining the series, the longest follow-up to death after surgery was 25 years in a patient who had been

symptomatic for more than 37 years. Again, death was not related to HPC.

Recurrence rates for sinonasal HPC vary in the literature from 7 to 50% (Table 1). Average time to recurrence is 6 to 7 years with more than 30 years reported for a case of ocular HPC.¹⁷ High recurrence rates have been traditionally accepted in the literature and are usually correlated with incomplete tumor removal. When multiple series are compared, it becomes clear that a substantial number of recurrent lesions are confined to a minority of patients. When the sinonasal tract is excluded, there are several reports of very aggressive HPC of the head and neck.^{18–20} Birzalgis et al²¹ reported on three cases of HPC involving the skull base, all with considerable local destruction. One of the cases was treated with primary radiation therapy (45 Gy) with some palliation of symptoms for nearly 4 years. The tumor was removed via an infratemporal fossa approach, with a small fragment of known disease remaining on the intrapetrous portion of the carotid artery. Over the next 2 years, the tumor relentlessly spread to intracranial and extracranial sites, but at death there were no known metastases. The other two cases in this report by Birzalgis et al are more recent and do not have long-term follow-up.

Treatment recommendations for sinonasal HPC center around wide local excision, the specifics of which are dependent on tumor size and location. Lateral rhinotomy has been the most frequently used surgical approach, although endoscopic techniques may be appropriate in selected cases.^{16,22} Specific tumor histologic findings are probably not a significant factor in dictating therapy for sinonasal HPC. Isolated reports of chemotherapy and radiation therapy trials have met with limited success, although brachytherapy has shown early promise at local control.^{6,23,24} Innovative surgical approaches have made an unresectable tumor the exception. When considering an aggressive resection, it should be remembered that very few affected patients actually die from sinonasal HPC. Because of reported recurrences that are decades removed from primary treatment, lifelong follow-up is mandatory.

The reason for the low-grade behavior of sinonasal

HPC is unknown. At least part of the explanation may be that nasal obstruction and epistaxis lead to earlier diagnosis and treatment. In addition, there tends to be more polypoid growth in sinonasal HPC instead of frank tissue invasion, as occurs with other somatic tumor sites. With several mature series of sinonasal HPC now reported, it is clear that with conservative treatment very few patients will actually die of their disease. Based on this, we recommend conservative therapy for patients with sinonasal HPC. This conservative approach should consist of debulking the tumor so that sinonasal obstruction is relieved and careful follow-up with serial endoscopic examinations and imaging studies. We recommend aggressive resection only if the tumor shows a propensity for growth and local invasion. In contrast to HPC occurring in somatic sites, there appears to be little or no risk of metastatic spread of sinonasal HPC. Head and neck HPC in locations outside of the sinonasal tract have demonstrated a more malignant character and merit a more aggressive treatment plan.

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Table 1. Recurrence Rates of Sinonasal Hemangiopericytoma

Series	Case No.*	Total Recovered	Percent
Compagno and Hyams ⁷	23	2	8
Chowla ²⁵	23	11	47
Backwinkel and Diddams ²⁶	21	12	57
Gudrun ⁶	15	5	13
Hughes and Bard ¹⁴	15	2	13
El-Naggar and Batsakis ¹⁵	14	2	14
Gorenstein and Facer ¹⁶	10	2	20
Eichhorn and Dickersin ¹²	9	4	44
Hesham ²²	2	0	0

*Number more than 100% of actual cases, as some cases reported in multiple series.

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