True Hemangiopericytoma of the Nasal Cavity

Immunohistochemical and Electron Microscopic Study of 2 Cases and a Review of the Literature on Sinonasal Hemangiopericytomas

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- Two cases of nasal tumors with pericytic myoid differentiation are reported. The tumors occurred in a 77-year-old woman and a 60-year-old man as polypoid lesions covered by normal mucosa. Histologically, the tumors were composed of uniform short spindle or stellate cells with indistinct cell borders arranged in narrow and short fascicles. Numerous blood vessels of various sizes were common in both cases. The tumor cells of both cases stained intensely with anti-vimentin and anti-actin antibodies, but not with anti-desmin, CD34, or anti-high-molecular-weight caldesmon antibodies. Ultrastructural examination revealed well-developed actin thin filaments with dense bodies, subplasmalemmal plaques, intercellular junctions, and irregular discontinuous basement membranes. These histopathologic features suggest true pericytic differentiation of the tumors (true hemangiopericytoma), unlike soft tissue-type hemangiopericytoma. Generally, sinonasal hemangiopericytomas are subdivided into soft tissue-type hemangiopericytomas and true hemangiopericytomas identical to the cases presented here. Soft tissue-type hemangiopericytomas are frequently highly aggressive, whereas true hemangiopericytomas show localized benign behavior. Sinonasal true hemangiopericytomas should be strictly differentiated from soft tissue-type hemangiopericytomas.

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Since first reported by Stout and Murray, hemangiopericytomas have been accepted as soft tissue tumors with distinct clinicopathologic features. They are characterized as benign or malignant, round to spindle cell tumors with numerous "staghorn" branching vascular channels. Another characteristic is the difficulty encountered in predicting their clinical behavior. The cellular differentiation of hemangiopericytomas has been questioned recently, because they commonly lack the immunohistochemical and electron microscopic differentiation properties of pericytes. Indeed, the hemangiopericytoma-like pattern is a nonspecific feature observed in various soft tissue tumors. Thus, hemangiopericytoma has become a nonspecific diagnosis rather than a specific concept indicating cellular differentiation.

Sinosonal hemangiopericytomas, which were also described by Stout, are thought to be distinct from hemangiopericytomas of the soft tissues because of their excellent prognosis and uniform cellular features. Compagno et al described this entity as "hemangiopericytoma-like tumor" as early as 1976, and Fletcher recently pointed out that sinonasal hemangiopericytomas are tumors with true pericytic myoid differentiation distinct from soft tissue hemangiopericytomas.

In this article, we present 2 cases of nasal hemangiopericytoma with conspicuous pericytic myoid differentiation, which seem to be intimately related to perivascular myoma. We also review cases reported previously as sinonasal hemangiopericytoma, hemangiopericytoma-like tumor, and glomus tumor and attempt to classify these cases into distinct clinicopathologic entities.

MATERIALS AND METHODS

Tissue specimens from 2 patients were fixed in formalin, embedded in paraffin, and stained with hematoxylin-eosin and silver impregnation stain. The streptavidin-biotin-peroxidase complex method (SABC kit, Dako, Kyoto, Japan) was used for immunohistochemistry. The primary antibodies and their final dilutions were anti-desmin (D33, Dako, 1:50), anti–α-smooth muscle actin (Dako, 1:200), anti-muscle actin (HHF-35, Enzo Diagnostics, New York, NY, 1:8000), CD34 (NU-4A1, Nichirei, Tokyo, Japan, 1:100), and anti-high-molecular-weight caldesmon (Dako, 1:50). Microwave pretreatment was performed for desmin and high-molecular-weight caldesmon analyses. The nuclei were counterstained with hematoxylin. Fresh tissue from case 1 was fixed in 2% glutaraldehyde and then embedded in epoxy resin. Ultrathin sections were stained with uranyl acetate and lead citrate and were observed using a JEM 1200EX transmission electron microscope (JEOL Ltd, Tokyo, Japan).

REPORT OF CASES

Case 1

A 78-year-old woman was admitted to the hospital complaining of right nasal obstruction and hemorrhage. Rhinoscopic examination disclosed a slightly red polypoid tumor covered by mucosa. The tumor grew slowly, and 3 months after her first admission a polypectomy was performed. The tumor arose from the right nasal septum and was extirpated together with neighboring normal mucosa. The tumor was approximately 25 mm in...
its maximum length. This was a recent case, and as of 4 months’ follow-up there had been no recurrence.

Case 2

A 60-year-old man was admitted to the hospital with a complaint of nasal hemorrhage. A right nasal polyp arising from the septum was observed under the rhinoscope. The polyp was excised piece by piece. The tumor recurred 1 year later and was re-excised with neighboring normal mucosa. The histology of the recurrent tumor was unchanged from that of the primary tumor. The patient’s postoperative course has been uneventful for 8 years.

PATHOLOGIC FINDINGS

Microscopic Findings

Both tumors exhibited polypoid growth covered by normal respiratory epithelium (Figure 1). The tumors were composed of uniform, rather eosinophilic short spindle or small stellate cells with indistinct cytoplasmic borders (Figures 2 and 3). The round to oval, uniform nuclei did not show atypia. The spindle cells tended to be arranged in short narrow fascicles. In case 1, concentric cellular configurations were occasionally seen (Figure 1). The tumors were rich in thin or dilated venule-like vessels, but staghorn-like vessels were rarely seen. There was sparse infiltration of a few lymphocytes and mast cells. Mitoses were rare and were seen only in case 1. Although silver impregnation stain disclosed numerous reticulum fibers between the tumor cells, they were thin and delicate and did not surround the individual cells clearly.

Immunohistochemistry

Both tumors were diffusely positive for vimentin. α-Smooth muscle actin and muscle actin (HHF-35) were also diffusely immunolabeled in both tumors, except for the concentric cellular area seen in case 1 (Figure 4). Neither tumor was positive for desmin, CD34, or high-molecular-weight caldesmon.

Electron Microscopy

Ultrastructurally, there were 2 cell types: spindle-shaped cells and stellate cells. Cells surrounding the blood vessels were characteristic elliptical cytoplasm with a small eccentric nucleus.
Recent, Granter et al described distinctive tumors showing perivascular proliferations of small spindle and round cells with myoid differentiation as “perivascular myomas.” They subdivided the tumors into myofibromatosis, hemangiopericytoma, and myopericytoma types, but these forms are interrelated and frequently indistinguishable. Perivascular myomas exhibit a broad histologic spectrum, from hemangiopericytoma-like to glomus tumor-like to epithelioid glomus tumor-like. However, it has become clear that hemangiopericytoma-like features are encountered in various soft tissue tumors and that soft tissue hemangiopericytomas generally lack pericytic differentiation, both ultrastructurally and immunohistochemically. Thus, hemangiopericytomas now tend to be recognized as heterogeneous tumors with various cellular differentiation.

Although hemangiopericytomas arising from sites other than soft tissues are rare, more than 60 cases of sinonasal hemangiopericytoma have been described in the literature since the first report by Stout. Because sinonasal hemangiopericytomas are frequently composed of more uniform, smaller cells and show excellent prognosis compared with the soft tissue types, it has been suggested that they may be distinct from soft tissue hemangiopericytoma. Compagno et al used the designation hemangiopericytoma-like tumor as early as 1976. In contrast, based on ultrastructural and immunohistochemical findings, Eichhorn et al suggested that nasal hemangiopericytoma does not differ from similar tumors in other locations. However, they observed focal smooth muscle actin immunoreactivity in nasal tumors, suggesting myoid differentiation, which is exceptional in soft tissue hemangiopericytomas. Recently, Fletcher concluded that sinonasal hemangiopericytoma is the only tumor showing convincing pericytic differentiation among the hemangiopericytomas that arise in adulthood.

We reviewed the cases of sinonasal hemangiopericytomas reported previously in the literature and found at least 3 histologic subtypes. The first subtype consists of tumors that are identical to soft tissue hemangiopericytomas composed of rather plump spindle cells with various degrees of nuclear atypia. At least 6 cases reported by Stout, Murashima, Lenczyk et al, Gürdin, and Tsuneyoshi et al, and some of the 11 cases reported by Eichhorn et al appear to be of this type (Table). Myoid differentiation was not confirmed in this type as well as in the soft tissue-type hemangiopericytomas. As expected, the tumors in these cases were frequently aggressive, and fatal cases were not uncommon. Very recently, intimate relationships between hemangiopericytoma and solitary fibrous tumor have been reported. It is important to differentiate these tumors, because most solitary fibrous tumors of the soft tissue are benign. CD34 is an extremely useful marker for this purpose.

The second subtype includes true hemangiopericytoma tumors, similar or almost identical to the present cases, which are composed of uniform spindle or stellate cells and which exhibit various degrees of myoid differentiation mimicking normal pericytes. Previously reported cases that seem to be of the true hemangiopericytoma type, based on the histologic descriptions and microphotographs, are summarized in the Table. Compagno et al analyzed the largest series, which included 23 cases. Although immunohistochemical and electron microscopic
examinations were not included in their study, we believe most of these cases are of the true hemangiopericytoma type, based on their distinctive uniform histologic features. We think that at least 1 (case 8) of the 11 cases described by Eichhorn et al also is of true hemangiopericytoma type, because of its histologic features and actin immunopositivity. Although some of 10 cases reported by Gorenstein et al are of true hemangiopericytoma, if not identical with respect to degree of myoid differentiation as judged by electron microscopy, are likely to belong to one cellular spectrum. In their biological behavior, these tumors seem to be almost benign, like true hemangiopericytoma. Recently, glomus tumors of the soft tissue were demonstrated to constantly express high-molecular-weight caldesmon, which does not exist in normal pericytes. It may be useful to analyze the differences between glomus tumors and pericytic tumors in the future.

In summary, the immunohistochemical and ultrastructural features of 2 cases of nasal hemangiopericytoma are described. They are identical to tumors formerly reported as hemangiopericytoma-like tumors and are thought to be true hemangiopericytomas. So-called sinonasal hemangiopericytomas reported in the past include different histologic types, and sinonasal true hemangiopericytoma should be differentiated from soft tissue–type hemangiopericytoma because of its far better prognosis.

References