Neoplasm with perivascular epithelioid cell differentiation (PEComa) of the nasal septum

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CASE REPORT

PEComa of the nasal septum

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Neoplasms showing perivascular epithelioid cell differentiation (PEComas) constitute a group of tumors composed of distinctive perivascular cells that coexpress melanocytic and muscle markers.[1] This family of tumors includes angiomyolipoma, clear cell sugar tumor of the lung, lymphangiomatosis, clear cell myomelanocytic tumor of the falciform ligament/ligamentum teres, and other unusual clear cell tumors arising at a variety of visceral or soft tissue sites.[1-4] In recent years, there has been an increase in the number of reported cases of PEComas, and it now appears that these tumors may arise at any anatomic location. We report the second case of PEComa occurring in the nasal cavity.[2]

A 50-year-old woman visited our hospital complaining of epistaxis, rhinorrhea, and unilateral nasal obstruction. Physical examination revealed a well-demarcated exophytic lesion attached to the anterior portion of the nasal septum by a relatively broad base. The patient's clinical history was otherwise unremarkable, and there was no family history of tuberous sclerosis. The lesion involved only the mucosa and could be excised surgically without septal perforation. Recovery was uneventful, and six years after excision, the patient is alive and well without evidence of local recurrence or metastatic disease.

Microscopic examination revealed a well-circumscribed but not encapsulated subepithelial lesion composed of fascicles and nests of plump epithelioid and spindled cells separated by a rich network of thin-walled capillary-sized vessels (Fig 1). Tumor cells presented clear to granular lightly eosinophilic cytoplasm with small, centrally placed, round to ovoid nuclei. There were rare mitoses and no necrosis. The overlying respiratory epithelium did not show any alteration.

The immunohistochemical stains showed a diffuse vimentin and α-smooth muscle actin positivity, whereas desmin and HMB-45 were focally positive (Fig 2). Epithelial membrane antigen, cytokeratins, CD10, CD31, CD34, S-100 protein, melanoma-associated antigen recognized by T cells (MART-1), estrogen receptor, and progesterone receptor immunostains were negative. Furthermore, about three percent of the neoplastic cells were in cell cycle, as estimated with the Ki-67 marker.

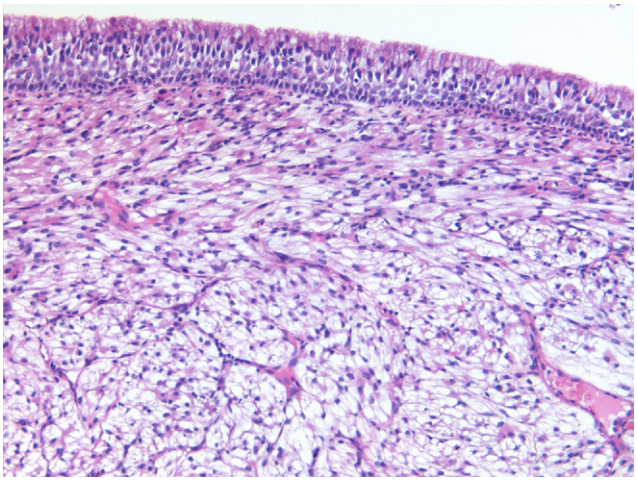

Figure 1 The tumor consisted of a uniform population of spindle and epithelioid cells mostly showing a clear cytoplasm.

Ultrastructurally, neoplastic cells contained numerous dense granules, whereas bundles of microfilaments were recognized in the peripheral cytoplasm of few neoplastic elements. The institutional review board of our hospital approved this study.

DISCUSSION

PEComas have been described only rarely in the head and neck region, and they involved the nasal cavity,[2] the soft tissues,[3] and the oral mucosa.[5] The differential diagnosis of nasal PEComa includes several entities. Malignant melanoma can be distinguished from PEComa by the expression of S-100 protein and the absence of actin immunoreactivity, even if rare spindled melanomas may show limited actin expression and S-100–negative melanomas have been described. In such cases, perivascular accentuation of tumor cells and strong and diffuse melanocytic marker positivity are additional important features. Epithelioid PEComa may be confused with metastatic clear cell renal carcinoma. The

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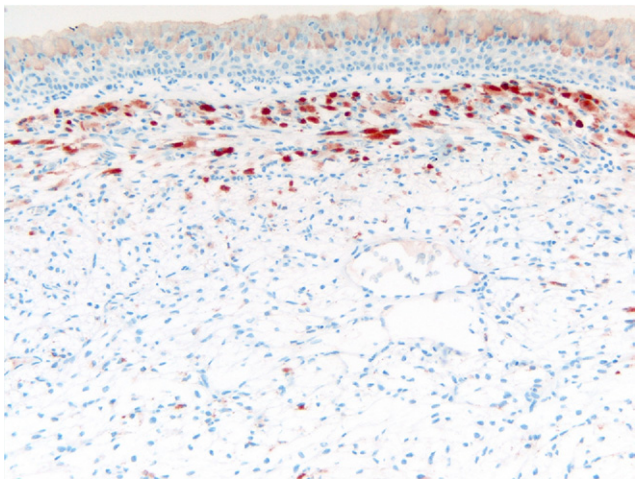

Figure 2 Focal immunostaining for HMB45 is present.

expression of melanocytic markers and the lack of reactivity for cytokeratins and CD10 are decisive for the diagnosis of the former. Paraganglioma is characterized by a more organoid growth pattern and positivity for protein S-100, chromogranin A, and synaptophysin. Morphologic evaluation is the cardinal criterion in distinguishing PEComas from true smooth muscle tumors because the role of immunohistochemistry in such differential diagnosis is less straightforward. PEComas show clear to lightly eosinophilic cytoplasm and round to oval nuclei with small nucleoli instead of the diffuse cytoplasmic eosinophilia and the "cigar-shaped" nuclei of the smooth muscle tumors that typically lack the delicate capillary network seen in PEComas. In sinonasal hemangiopericytoma, neoplastic cells show amphophilic to eosinophilic cytoplasm and are organized in short fascicles, often around vascular channels ranging from capillary size to large sinusoidal spaces, thus mimicking the histologic picture of PEComa. Both tumors are positive for myogenic markers, but sinonasal hemangiopericytoma does not express melanocytic markers.

Clinically, the vast majority of PEComas follow a benign course, although a subset of these tumors behaves in a malignant fashion. Malignant PEComas can be very aggressive, with clinical behavior similar to a high-grade sarcoma. In a recently published study, criteria for malignancy were suggested,[3] but, until more cases with long clinical follow-up are evaluated in a systematic fashion, firm criteria for malignancy remain uncertain. Our patient is alive and well without evidence of local recurrence or metastatic disease six years after the surgical excision of the lesion, indicating a benign clinical course.

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John Panelos, acquisition of histopathological data, writing of the manuscript, final approval of the version to be published; **Oreste Gallo,** acquisition of clinical data, review of the manuscript, final approval of the version to be published; **Iacopo Scala,** acquisition of clinical data, review of the manuscript, final approval of the version to be published; **Alessandro Franchi,** acquisition of histopathological data, writing of the manuscript, final approval of the version to be published.

DISCLOSURES

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