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Asymptomatic Septal Mass

Richard L. Arden, MD; Zachary H. Griggs, DO; Elizabeth A. Wey, MD

A woman in her 40s presented with a 2-month history of a slowly enlarging, asymptomatic anterior septal mass. She denied a history of localized trauma, substance abuse, or prior endonasal surgery. Examination revealed a nontender, 8-mm, pink-red, soft, fleshy growth just posterior to the membranous septum near the nostril apex. Histopathological analysis demonstrated a well-circumscribed submucosal lesion comprised of bland, round, syncytial cells, arranged in a concentric fashion around variably sized slit-like vascular spaces, a minority of which were staghorn in appearance (Figure, A-C). No significant perivascular hyalinization was identified. The overlying nasal mucosa and surrounding submucosal glands were unremarkable. Immunohistochemical stains demonstrate the cellular population to be strongly positive for smooth-muscle actin (SMA), muscle-specific actin, and CD34 (Figure, D), but negative for B-catenin and epithelial membrane antigen.

**Diagnosis**

D. Myopericytoma

**Discussion**

The new World Health Organization (WHO) classification of soft tissue tumors (previously reported in 2002) was updated in 2013 and now acknowledges that myofibroma and/or myofibromatosis represents morphological points along the spectrum of myopericytic neoplasms. Myopericytoma (MPC) is a mesenchymal tumor derived from the perivascular myoid cell, sharing features with smooth-muscle and glomus cells. Previously encompassed under the broad category of hemangiopericytomas, these perivascular myomas can...
be categorized based on the predominant histologic pattern: myofibromatosis, glomangiopericytoma, and myopericytoma. Ultimately, diagnosis is based on these characteristic morphological features and supportive immunohistochemistry, recognizing that some overlap exists with the exception of glomangiopericytoma, which is histologically distinct and occurs in the sinonasal region.

Myopericytoma occurs mainly in childhood to mid-adult years with a male predominance and predilection for dermal and subcutaneous locations in the lower extremities. Of the roughly 15.0% that occur in the head and neck, only 2.5% are represented in the nose and paranasal sinuses, accounting for less than 1.0% of sinonasal tumors overall. This typically benign, slowly growing neoplasm, often presents as a solitary painless mass that can cause epistaxis and nasal obstruction. On examination, the lesion appears nodular to polypoid, soft and fleshy, friable, circumscribed, possesses a gray-pink to beefy red color, and can be mistaken for an inflammatory polyp.

Myopericytoma are well circumscribed (usually <2 cm) but unencapsulated neoplasms with bland and relatively monomorphic oval to spindle-shaped cells that are arranged in a concentric or whorling perivascular pattern. They differ from glomus cells by their larger size, more abundant cytoplasm, and lack of a clearly demarcated cell border. Immunostaining is characteristically strong for vimentin and SMA expression but limited to small vessels for CD34. Pericytes are SMA positive. Staining is characteristically negative for EMA, cytokeratins, and usually desmin. By contrast, the rare sinonasal solitary fibrous tumor exhibits a patternless proliferation of spindle cells admixed with variable amounts of ropy keloidal collagen, increased perivascular and stromal collagen, haphazardly arranged cells, and thin-walled vascular spaces. Strong diffuse immunoreactivity for CD34 and CD99 is seen, with variably positive to absent expression for SMA.3,4,6 Unlike MPC, perivascular concentric growth with accentuated perivenuous myoid proliferation is not a prominent feature of myofibroma. Diagnosis of myofibroma is typically considered for inoperable tumors or metastases. Radiation therapy has been reserved for unresectable and recurrent tumors. Chemotherapy has demonstrated limited efficacy and considered for inoperable tumors or metastases.

ARTICLE INFORMATION

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