

Clinical Challenge | PATHOLOGY

Nasal Obstruction and Epistaxis

Kaete A. Archer, MD; Parul Goyal, MD; Anthony J. Mortelliti, MD

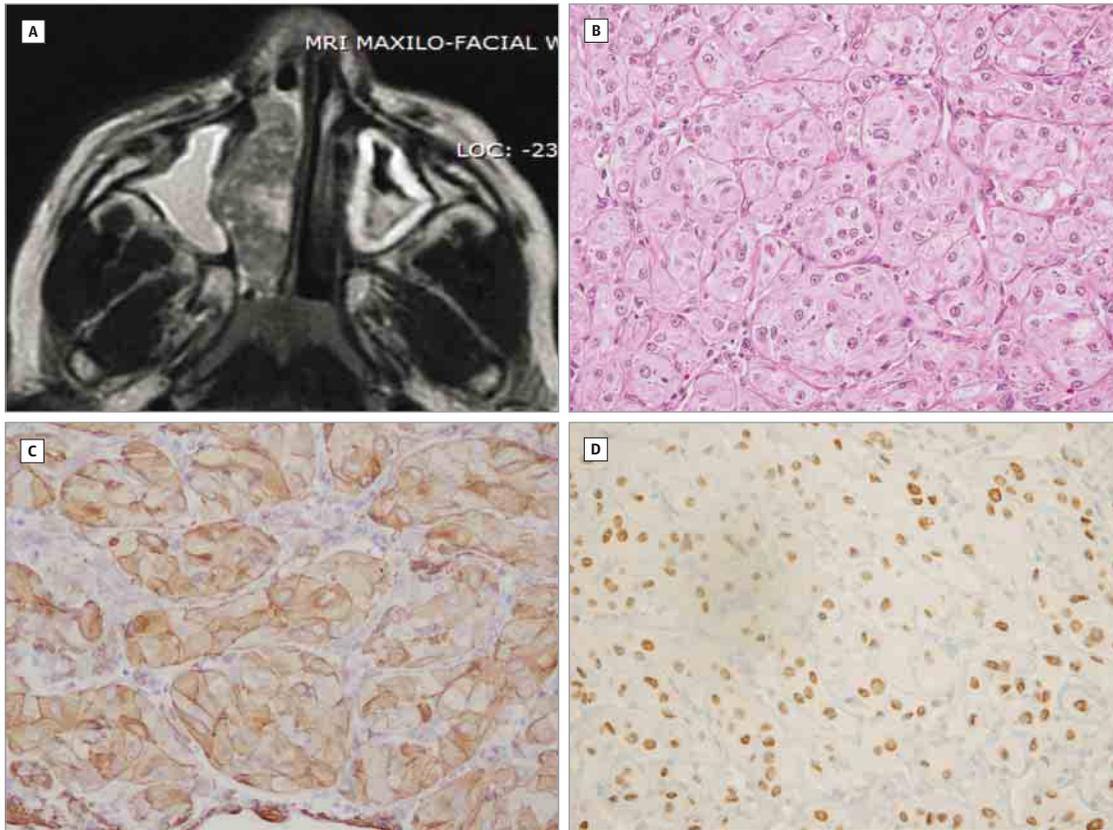


Figure. A, T2-weighted magnetic resonance imaging (MRI) scan of a right nasal cavity mass extending to the posterior choana with heterogeneous intermediate signal intensity. B and C, Histopathologic images. B, A sheetlike proliferation of epithelioid and polygonal cells with pale eosinophilic granular cytoplasm and relatively uniform vesicular nuclei (hematoxylin-eosin, original magnification $\times 40$). C, The tumor cells stained positive for smooth muscle actin (original magnification $\times 20$). D, The tumor cells stained positive for transcription factor E3 (original magnification $\times 20$).

A preteen boy presented with a 1-year history of right-sided nasal obstruction and a 4-day history of intermittent right-sided epistaxis following blunt trauma to the nose. The epistaxis occurred 3 to 4 times a day and resolved with pressure. He did not have facial pain, facial paresthesia, or visual changes. There was no family or personal history of bleeding disorders. Nasal endoscopy revealed a large, well-vascularized, polypoid mass filling the right anterior nasal cavity. A computed tomographic scan showed a right nasal cavity mass (4.5×1.7 cm) extending to the posterior choana with opacification and bony remodeling of the right maxillary sinus. The mass had heterogeneous intermediate signal intensity on T2-weighted magnetic resonance imaging (Figure, A). The patient was taken to the operating room for biopsy and possible excision of the nasal mass. As the lesion was biopsied, there was brisk bleeding. However, the lesion was found to have a narrow pedicle of attachment and was resected in its entirety. The mass was based on the superior aspect of the nasal septum and cribriform plate. Hematoxylin-eosin stain showed a sheet-like proliferation of epithelioid and polygonal cells with pale eosinophilic granular cytoplasm and relatively uniform vesicular nuclei (Figure, B). The cells were arranged in nests. Branching, staghorn-like blood vessels were scattered throughout the tumor. The tumor cells stained positive for smooth muscle actin (Figure, C), muscle specific actin, Bcl-2, *INI-1*, and transcription factor E3 (Figure, D).

WHAT IS YOUR DIAGNOSIS?

- A. Alveolar rhabdomyosarcoma
- B. Nasal alveolar soft part sarcoma
- C. Malignant melanoma
- D. Juvenile nasopharyngeal angiofibroma

Diagnosis

B. Nasal alveolar soft part sarcoma

Discussion

These findings were consistent with alveolar soft part sarcoma (ASPS), an extremely rare malignant tumor representing 0.4% to 1.0% of all soft-tissue sarcomas.¹ These tumors usually present in the lower extremities of adolescents or young adults.¹ Approximately 25% of ASPS tumors present in the head and neck, with a predilection for the tongue and orbit.^{1,2} Alveolar soft part sarcoma of the nasal and paranasal region is even more rare, with only 4 cases reported in the English literature.^{1,3-5} They are more common in women than men by a 2:1 ratio.¹ The etiology of ASPS is unknown.⁶

This tumor was first described by Christopherson et al⁶ based on a unique histological appearance and clinical behavior. Alveolar soft part sarcoma is usually a slow-growing and well-vascularized mass with no clinical features to suggest malignant disease.¹ When ASPS presents in the nasal cavity, nasal obstruction may be the only symptom. The patient in this case had nonspecific nasal obstruction for 1 year but denied facial pain or clinical symptoms consistent with neural or orbital invasion. Magnetic resonance imaging was negative for intracranial extension.

Grossly, ASPS tends to be friable and hemorrhagic with a yellow-red surface.⁶ The light microscopic characteristics are distinctive and nearly pathognomonic. Alveolar soft part sarcoma cells are large, oval to polyhedral, usually with distinct cell boundaries.⁶ Typically, cells are arranged in nests with central sloughing creating a pseudo-alveolar shape.⁷ These tumor cell nests are separated by vascular channels and fibrous septa.² Of note, head and neck ASPA in children will more commonly have a solid pattern of growth without nesting.⁷ The nuclei are round with vesicular chromatin and single prominent eccentric nucleoli.^{6,8} The cytoplasm is finely granular with occasional vacuoles,

lightly eosinophilic on hematoxylin-eosin stain, and contains rhomboid-shaped, periodic acid-Schiff-positive crystalline structures.⁸

Alveolar soft part sarcoma has uncertain histogenesis. This tumor stains negatively for myoglobin.⁶ Only a subset of ASPS cases express desmin as well as neuron-specific enolase.² Nuclear expression of transcription factor E3 (TFE3) is a sensitive and specific marker for ASPS that represents the fusion of the novel alveolar soft part sarcoma gene (*ASPL*) on chromosome 17q25 to the *TFE3* gene on chromosome Xp11.⁹

Histologically, the differential diagnosis includes paraganglioma, adrenal cortical carcinoma, hepatocellular carcinoma, alveolar rhabdomyosarcoma, malignant melanoma, granular cell tumor, and metastatic renal cell carcinoma.⁸ The pseudoalveolar arrangement of cells is similar to that found in paragangliomas.⁶ Vascular invasion and, more rarely, mitotic features seen in ASPS may also be seen in melanoma. Clear-cell changes in ASPS may closely resemble those in metastatic renal cell carcinoma.⁸ This differential diagnosis was narrowed based on age, physical examination, immunohistochemical staining, and radiographic workup.

Despite the slow growth of the tumor, overall prognosis is poor, with a high tendency for early metastatic spread. In a study by Cho et al,⁸ 48% of patients with ASPS presented with metastatic disease. Lieberman et al¹⁰ reported a 19% incidence of local recurrence over 27 years. The overall survival rates reported are 77% at 2 years, 60% at 5 years, 38% at 10 years, and 15% at 20 years.¹⁰ No specific survival data exist in the literature on ASPS in the sinonasal region.

Radical surgical excision with tumor-free margins is the preferred treatment for ASPS.⁸ Radiation and/or chemotherapy is used as adjuvant treatment. Nasal and paranasal sinus tumors can be difficult to excise with tumor-free margins, and therefore, postoperative radiation is common. In this patient, we performed complete surgical resection of the tumor. He recovered well and is currently receiving adjuvant chemotherapy.

ARTICLE INFORMATION

Author Affiliations: SUNY Upstate Medical University, Syracuse, New York.

Corresponding Author: Anthony J. Mortelliti, MD, Department of Otolaryngology and Communication Sciences, SUNY Upstate Medical University, 750 E Adams St, Syracuse, NY 13210 (mortella@upstate.edu).

Published Online: March 5, 2015.
doi:10.1001/jamaoto.2015.118.

Conflict of Interest Disclosures: None reported.

Additional Contributions: Chris Curtiss, MD, Department of Pathology, SUNY Upstate Medical University, prepared the pathology images. He was not compensated for his contribution.

REFERENCES

1. Yigitbasi OG, Guney E, Kontas O, Somdas MA, Patiroglu T. Alveolar soft part sarcoma: report of a case occurring in the sinonasal region. *Int J Pediatr Otorhinolaryngol*. 2004;68(10):1333-1337.

2. Mukai M, Torikata C, Iri H, et al. Histogenesis of alveolar soft part sarcoma: an immunohistochemical and biochemical study. *Am J Surg Pathol*. 1986;10(3):212-218.

3. Barbareschi M, Ferrero S, Ottaviani F. Alveolar soft part sarcoma of the nasal cavity. *Pathologica*. 1988;80(1067):363-370.

4. Chatterji P, Purohit GN, Ramdev IN, Soni NK. Alveolar soft part sarcoma of the nasal cavity and paranasal sinuses. *J Laryngol Otol*. 1977;91(11):1003-1008.

5. Dezano P, Lifschitz-Mercer B, Czernobilsky B, Rosai J. Alveolar soft-part sarcoma of paranasal sinuses. *Int J Surg Pathol*. 2010;18(1):66-67.

6. Christopherson WM, Foote FW Jr, Stewart FW. Alveolar soft-part sarcomas: structurally characteristic tumors of uncertain histogenesis. *Cancer*. 1952;5(1):100-111.

7. Bocklage TJ, Quinn R, Schmit B, Verschraegen C, eds. *Bone and Soft Tissue Tumours*. London, England: JP Medical Ltd; 2014.

8. Cho YJ, Kim JY. Alveolar soft part sarcoma: clinical presentation, treatment and outcome in a series of 19 patients. *Clin Orthop Surg*. 2014;6(1):80-86.

9. Argani P, Lal P, Hutchinson B, Lui MY, Reuter VE, Ladanyi M. Aberrant nuclear immunoreactivity for TFE3 in neoplasms with *TFE3* gene fusions: a sensitive and specific immunohistochemical assay. *Am J Surg Pathol*. 2003;27(6):750-761.

10. Lieberman PH, Brennan MF, Kimmel M, Erlandson RA, Garin-Chesa P, Flehinger BY. Alveolar soft-part sarcoma: a clinico-pathologic study of half a century. *Cancer*. 1989;63(1):1-13.