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Soft tissue tumors of the sinonasal tract

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ABSTRACT

Primary soft tissue tumors arising in the sinonasal tract are rare. While many mesenchymal neoplasms have been reported in the nasal cavity, sinuses, and nasopharynx, few are distinctive to this anatomic region. Some tumor types are relatively more common in this area, such as schwannoma and rhabdomyosarcoma. Nasopharyngeal angiofibroma and sinonasal hemangiopericytoma are unique entities of the sinonasal tract, as well as the recently characterized biphenotypic sinonasal sarcoma. This review discusses the clinical, morphologic, and immunohistochemical features and currently known molecular data of the more frequently encountered soft tissue tumors of the sinonasal tract.

Introduction

Primary soft tissue tumors of the sinonasal tract are rare. Sarcomas account for approximately 1–3% of all head and neck malignancies; overall up to 15% and 35% of sarcomas arise in the head and neck region in adults and children, respectively.[1,2] In the sinonasal tract specifically, epithelial neoplasms far outnumber mesenchymal tumors, and in this location most sarcomas need to be distinguished from the more likely possibilities of spindle cell or sarcomatoid squamous cell carcinoma and spindle cell or desmoplastic melanoma. Given that many head and neck malignancies undergo radiation treatment, post-irradiation sarcomas may also be considered in the appropriate clinical context. The clinical presentation of tumors in the sinonasal tract is typically non-specific and patients often suffer mass-related symptoms secondary to obstruction or locoregional extension/invasion, such as pain, congestion, epistaxis, rhinorrhea, or cranial nerve abnormalities. Due to the anatomic complexity and constraints of the region, benign tumors may appear clinically or radiologically worrisome and cause significant morbidity due to local effects or required composite resections.

Soft tissue tumors comprise numerous and diverse tumor types, and owing to their rarity and significant morphologic overlap, they often present diagnostic challenges. Molecular advances in soft tissue pathology over the past two decades have led to the development of numerous diagnostic tools, such as immunohistochemical markers and fluorescent in situ hybridization (FISH) probes. Nearly all tumor types have been reported in the sinonasal tract, but some entities are more common or are unique to this anatomic region. Herein we review the clinicopathologic features of the most frequently encountered primary sinonasal soft tissue tumors, with attention to pertinent diagnostic immunohistochemical, molecular features, and differential diagnoses.

Lobular capillary hemangioma

Lobular capillary hemangioma (LCH), previously known as pyogenic granuloma, may rarely arise in the sinonasal tract.

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LCH occurs most commonly during the fourth and fifth decades and affects men and women equally; tumors are most common in the lip and oral cavity. In the nasal cavity, LCH typically arises in either the anterior portion of the nasal septum or on the turbinates, and grossly appears as red or tan lobulated polypoid masses and measure up to 1.5 cm.[3] LCH appears as a multilobulated capillary proliferation in the submucosa with frequent ulceration of the overlying mucosa and are often associated granulation tissue and acute and chronic inflammation. The lobules are comprised of a proliferation of variably sized capillaries; a larger "feeder" vessel may be present (Fig. 1A). Long-standing lesions show maturation with stromal hyalinization and gaping vessels. While the more cellular foci may resemble solid growth and exhibit mitotic activity, careful examination reveals that the lobules are comprised of packed vessels with intact pericytes (Fig. 1B). Immunohistochemical stains can confirm the capillary architecture within the lobules, with CD31, CD34, and ERG positivity in endothelial cells and SMA positivity in pericytes. LCH is benign and associated with excellent prognosis, although higher recurrence rates (42%) have been reported for sinonasal tumors.[4]

The typical lobular architecture and circumscription of LCH allows distinction from most other vascular tumors, most notably cavernous hemangioma (which is characterized by variably sized but much larger dilated vessels) and angiosarcoma. Primary angiosarcoma in the sinonasal tract is rare and is characterized by endothelial atypia, infiltrative growth, absence of pericytes around vasoformative spaces, and frequent foci of solid growth.[5] Late-stage LCH may occasionally resemble nasopharyngeal angiofibroma but lacks a cellular stromal component and does not have a smooth muscle component in the vessel walls.

Nasopharyngeal angiofibroma

Nasopharyngeal angiofibroma (also referred to as juvenile angiofibroma) is a benign tumor unique to the sinonasal tract and comprises less than 1% of head and neck tumors.[6,7] It is considered to occur nearly exclusively in adolescent males during the second decade of life. It has been long considered that these tumors are hormonally driven based on their predilection for adolescent males and expression of androgen receptor.[8] Most nasopharyngeal angiofibromas arise near the sphenopalatine foramen; despite being benign with no metastatic potential, tumors may occasionally show locally aggressive growth into the nasal cavity or oropharynx, or invasion into craniofacial bones. Up to 24% of cases may recur, usually in the setting of incomplete resection or widely invasive tumors. Tumors typically have radiographic features suggestive of a vascular lesion, and a presumptive diagnosis is often made in the clinical setting of an adolescent male patient. Biopsy is often avoided given the propensity of the tumor for bleeding, and patients typically directly undergo surgical resection.

Tumors grossly appear as tan-white sessile or lobulated masses. Microscopically, nasopharyngeal angiofibroma is unencapsulated with a variably sized, irregularly shaped vessels set in a hyalinized and collagenous stroma. The vessels are thin-walled, and the walls show a variable smooth muscle component that is occasionally absent or discontinuous (Fig. 2A). The stromal component is overall moderately cellular, with haphazardly arranged bland spindle and stellate stromal cells having round-to-ovoid nuclei, small indistinct nucleoli, and palely eosinophilic cytoplasm (Fig. 2B). Occasional tumors may have multinucleated stromal cells, mild reactive atypia, stromal predominance, and myxoid stromal change. Extensive stromal fibrosis is often seen in long-standing tumors. Tumors lack significant cytologic atypia, high mitotic activity, and atypical mitotic figures.

The morphologic features are distinctive, and the immunohistochemical profile is non-specific. In the vascular component, endothelial cells are positive for CD31, CD34, and ERG, and SMA highlights the variable mural smooth muscle component. Androgen receptor is positive in both the endothelial and stromal component. The stromal component is positive for SMA, often showing perivascular accentuation. β-Catenin overexpression in stromal cells and *CTNNB1* mutations have been reported,[9] prompting some authors to suggest that nasopharyngeal angiofibroma may be related to familial adenomatous polyposis[10–12]; however, a definitive relationship remains uncertain.

The differential diagnosis of nasopharyngeal angiofibroma includes LCH and sinonasal hemangiopericytoma. LCH with

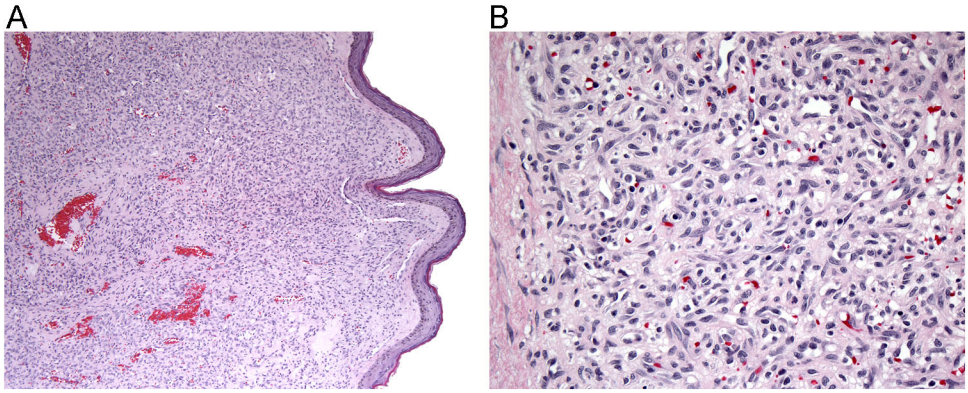

Fig. 1 – Lobular capillary hemangioma is a well-circumscribed submucosal tumor that has a lobular configuration of closely packed, variably sized vessels (A) that are lined by benign-appearing endothelial cells with pericytes around many of the smaller vessels (B).