# Distinctive Head and Neck Bone and Soft Tissue Neoplasms

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**KEYWORDS**

• Chondrosarcoma • Chordoma • Osteosarcoma • Biphenotypic sinonasal sarcoma • Angiofibroma • Glomangiopericytoma • Rhabdomyosarcoma

**Key Points**

- Pathologic features of the recently described biphenotypic sinonasal sarcoma (BSNS).
- Association of angiofibroma (AF) and familial adenomatous polyposis (FAP).
- Differentiate between skull base chondroid chordoma and chondrosarcoma.
- Unique features of gnathic osteosarcomas.
- Rhabdomyosarcoma (RMS) of the head and neck.
- Diagnosis of glomangiopericytoma (GPC).

## ABSTRACT

Benign and malignant primary bone and soft tissue lesions of the head and neck are rare. The uncommon nature of these tumors, combined with the complex anatomy of the head and neck, pose diagnostic challenges to pathologists. This article describes the pertinent clinical, radiographic, and pathologic features of selected bone and soft tissue tumors involving the head and neck region, including angiofibroma, glomangiopericytoma, rhabdomyosarcoma, biphenotypic sinonasal sarcoma, chordoma, chondrosarcoma, and osteosarcoma. Emphasis is placed on key diagnostic pitfalls, differential diagnosis, and the importance of correlating clinical and radiographic information, particularly for tumors involving bone.

## ANGIOFIBROMA

### OVERVIEW

AF, also known as juvenile nasopharyngeal AF, is an uncommon benign vascular neoplasm of the head and neck, making up 0.05% of head and neck tumors and occurring almost exclusively in adolescent boys (9–19 years of age).[1] AF originates from a fibrovascular nidus in the posterolateral nasal cavity near the sphenopalatine foramen or pterygoid canal.[2] The blood supply of AFs typically arises from the ipsilateral internal maxillary artery, but any branch from the internal or external carotid artery is a possible feeder vessel.[1]

Symptoms include unilateral nasal obstruction; epistaxis, which can be massive; nasal discharge; and otitis media. Although pathologically benign, larger tumors may behave aggressively, bulging into the soft palate and extending into the maxillary and sphenoid sinuses, orbit, and medial cranial fossa, causing facial deformities, headaches, and proptosis.[1] Approximately 10% to 20% of AFs extend intracranially.[3,4]

Routine radiographs and CT and MRI scans reveal a soft tissue density in the nasopharynx. Angiography allows identification of the feeder vessel, which is crucial for selecting the appropriate surgical approach and reveals characteristic irregular, tortuous vessels and the tumor blush of

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AF.[5] One of the more commonly adopted preoperative staging systems to determine extent of surgery is proposed by Radkowski and colleagues[6] (**Table 1**).

The pathogenesis of AF is unknown, and developmental, hormonal, and genetic causes have been proposed. The role of testosterone in tumor development and growth is suggested by its gender predilection, incidence during puberty, and possible regression after maturation.[7] Patients with FAP, an autosomal dominant disorder characterized by a germline mutation of the *APC* gene, are 25 times more likely to develop AFs.[8,9] Approximately 20% of patients with FAP do not have a family history and the diagnosis of AF may precede the diagnosis of FAP.[10]

## PATHOLOGIC FEATURES

AFs vary in size depending on the extent of disease. The mean size is approximately 4 cm.[5] They have a polypoid shape with a rounded or bosselated contour. Cut sections demonstrate porous and focally hemorrhagic tissue.

Microscopically, AFs are unencapsulated lesions covered by nasopharyngeal mucosa that may be focally ulcerated. At low power, a prominent hemangiopericytoma (HPC)-like vascular pattern is appreciated (**Fig. 1**). The blood vessels have an irregular smooth muscle layer that may be focally attenuated or absent, and the endothelial layer is typically thin and lacks atypia. The dense fibrous stroma contains scattered plump, epithelioid, or stellate fibroblasts and coarse and fine collagen fibers (see **Fig. 1**). Up to mild nuclear atypia may be occasionally present, but mitotic activity is low. Stromal mast cells are frequently identified.

In long-standing lesions, focal myxoid areas may be seen and the center of the tumor tends to be more collagenous with diminished vascularity. Embolization material in vessels and adjacent reactive changes are identified in resected specimens after embolization. Other degenerative features include multinucleated stromal cells.

*Table 1*
**Radkowski classification of angiofibromas**

| Stage | Extent |
|---|---|
| Ia | Limited to the nose and nasopharyngeal area |
| Ib | Extension into 1 or more sinuses |
| IIa | Minimal extension into pterygopalatine fossa |
| IIb | Occupation of the pterygopalatine fossa without orbital erosion |
| IIc | Infratemporal fossa extension without cheek or pterygoid plate involvement |
| IIIa | Erosion of the skull base (middle cranial fossa or pterygoids) |
| IIIb | Erosion of the skull base with intracranial extension with or without cavernous sinus involvement |

*From* Radkowski D, McGill T, Healy GB, et al. Angiofibroma. Changes in staging and treatment. Arch Otolaryngol Head Neck Surg 1996;122(2):122–9.

### *Key Pathologic Features*

- Unencapsulated, hypocellular submucosal lesion involving the nasopharynx
- Prominent HPC-like vascular pattern
- Blood vessels with irregular smooth muscle coats
- Dense fibrous stroma with scattered plump epithelioid or stellate fibroblasts and coarse and fine collagen fibers
- At most mild atypia but low mitotic activity
- Stromal mast cell frequently identified
- In long-standing lesions, the center of the tumor tends to be more collagenous with diminished vasculature.
- Other degenerative changes include focal myxoid change and multinucleated stromal cells.
- Embolization material in blood vessels

## IMMUNOHISTOCHEMICAL AND MOLECULAR FEATURES

The stromal cells of AFs are positive for vimentin and negative for desmin and smooth muscle actin (SMA) (**Fig. 2**).[5] SMA may highlight occasional myofibroblasts, which are likely regressive in nature.[11] CD117 may be positive in both the stromal cells and the vascular endothelial cells.[12] AFs also frequently demonstrate nuclear β-catenin staining (see **Fig. 2**).[13]

Studies of sex hormone receptors have shown inconsistent results.[14,15] In general, AF demonstrates nuclear staining for AR and infrequently for estrogen receptor (ER) (see **Fig. 2**). ER-β, however, which is found in normal mesenchymal tissues, is often positive in AF, unlike ER-α.[15] CD34, CD31, FLI-1, and factor VIII–related antigen (FVIIIRAg) are positive in the vascular component