



Get Clarity On Generics

Cost-Effective CT & MRI Contrast Agents



FRESENIUS
KABI

WATCH VIDEO

AJNR

An Unusual Sinonasal Tumor: Soft Tissue Perineurioma

W.S. Fang, L.L. Emerson, J.P. Hunt and K.L. Salzman

AJNR Am J Neuroradiol 2009, 30 (2) 437-439

doi: <https://doi.org/10.3174/ajnr.A1284>

<http://www.ajnr.org/content/30/2/437>

This information is current as
of August 23, 2025.

CASE REPORT

W.S. Fang
L.L. Emerson
J.P. Hunt
K.L. Salzman

An Unusual Sinonasal Tumor: Soft Tissue Perineurioma

SUMMARY: Soft tissue perineuriomas are an unusual type of peripheral nerve sheath tumors distinct from schwannomas and neurofibromas, with interesting histologic findings. They are not well characterized on radiographic examination. We report this case of a patient with sinonasal perineurioma to help define the imaging and pathologic features of this rare head and neck tumor.

Soft tissue perineuriomas are slowly growing peripheral nerve sheath tumors. They are extraordinarily rare in the sinonasal cavity, with only 1 previous report to our knowledge.¹ They are difficult to distinguish from other benign lesions; however, their imaging findings are not well characterized. We report this case of a patient with sinonasal perineurioma to better define the imaging and pathologic features of this rare head and neck tumor.

Case Report

Clinical Findings

A 63-year-old woman experienced progressive left-sided nasal obstruction and rhinorrhea. She denied epistaxis, trauma, or allergic rhinitis. Symptoms persisted 2 years despite nasal steroids and antibiotics. Endoscopy revealed a mildly vascular mass in the posterior nasal cavity (Fig 1).

Radiologic Findings

A contrast-enhanced CT study showed a very well-defined, lobulated, heterogeneously enhancing soft tissue mass centered in the posterior nasal cavity, extending through the choanae into the nasopharynx. There was mild widening of the choanae without bony sclerosis or destruction. The mass abutted the sphenopalatine foramen but did not widen the vidian canal (Fig 2).

Surgical Findings

The lesion was excised on endoscopic examination. A firm stalk from the mass was noted, extending into the vidian canal. The mass was retracted inferiorly, drawing the vidian nerve out of the canal, before being resected with the stalk en bloc.

Pathologic Findings

Gross Examination. A 10.9-g, 4.0 × 3.0 × 2.3-cm lobulated, rubbery, pale pink-white mass attached to a small, tattered, 0.6-cm pedicle was submitted.

Histologic Features

The tumor was characterized by an unencapsulated but well-circumscribed proliferation of bland spindle cells with tapered nuclei and delicate bipolar cytoplasmic processes. The spindle cells formed a storiform (cartwheel) pattern of growth with short, interlacing, wavy

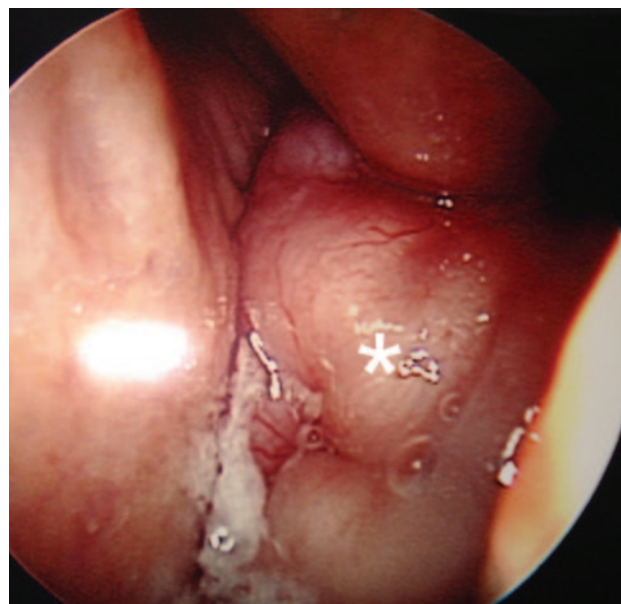


Fig 1. Nasal endoscopy. A lobulated mass with mild vascularity is seen left of the midline (asterisk). The middle turbinate is partially visible above the mass. The nasal septum is on the right.

fascicles in the background of a diffusely collagenous and focally myxoid stroma. Perivascular whorling was also noted focally.

Immunohistochemical Features

Multiple small foci of cells showed positive cytoplasmic staining with epithelial membrane antigen, characteristic of a perineurioma. No immunoreactivity was seen for S100 or CD57 proteins, typical for a schwannoma. Multiple other preparations were also negative.

Fluorescent In Situ Hybridization

The dual color breakapart probe LSI EWSR1 (Vysis, Downers Grove, Ill) located at region 22q12.1–22q12.3 was used to detect partial or complete loss of chromosome 22. There were 58% of the tumor cells that showed monosomy for chromosome 22 (Fig 3).

The histologic and immunohistochemical findings support the diagnosis of soft tissue perineurioma. The finding of a partial or complete deletion of chromosome 22 by fluorescent in situ hybridization confirms the neoplastic nature of the tumor and lends further support.

Discussion

Perineuriomas are composed of perineurial cells, normally found as a thin layer around myelinated and unmyelinated peripheral nerves. These cells function as a perifascicular diffusion barrier. Perineuriomas are classified as soft tissue (ex-

Received July 7, 2008; accepted July 14.

From the Departments of Neuroradiology (W.S.F., K.L.S.), Pathology (L.L.E.), Otolaryngology (J.P.H.), and Radiology (K.L.S.), University of Utah, Salt Lake City, Utah.

Please address correspondence to Wayne S. Fang, University of Utah, Department of Neuroradiology, 50 North Medical Dr, Salt Lake City, UT 84132; e-mail: wayne.fang@hsc.utah.edu

DOI 10.3174/ajnr.A1284

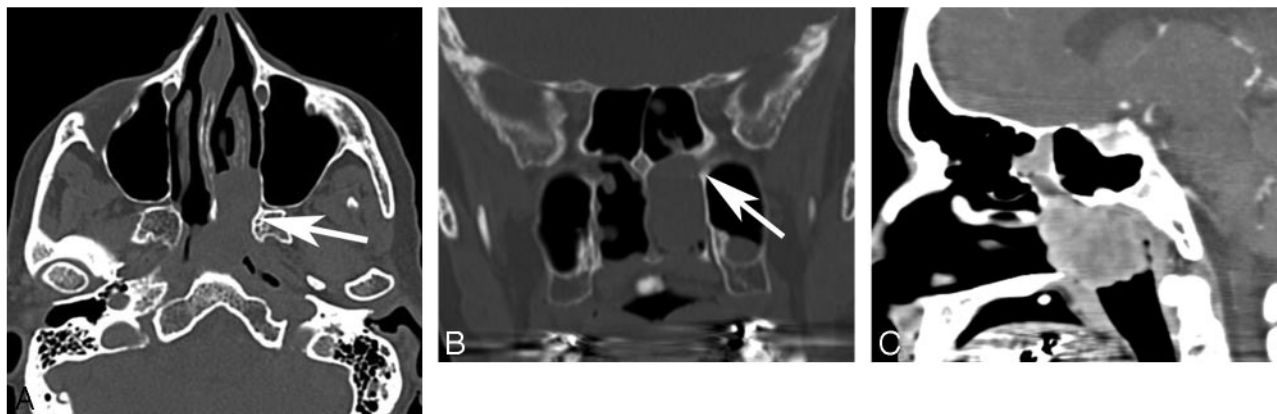


Fig 2. Sinonasal CT scan. *A*, A mass in posterior nasal cavity and nasopharynx widens left choanae (arrow). *B*, Coronal reconstruction shows mass abuts the sphenopalatine foramen (arrow). *C*, Postcontrast sagittal image shows heterogeneous enhancement of the mass and extension into nasopharynx.

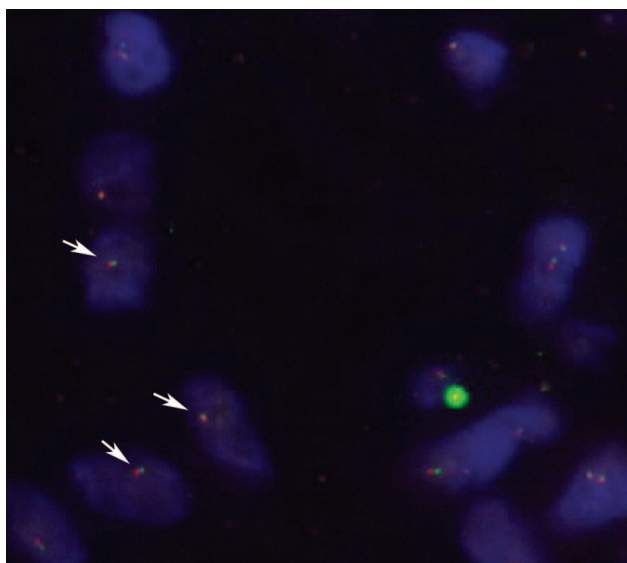


Fig 3. Fluorescent in situ hybridization. A significant number of cells show only 1 fusion signal intensity (paired green-red signal intensity [arrows]) for the EWSR1 probe rather than the normal 2 fusion signals, indicating monosomy of chromosome 22 in these cells.

traneural) or intraneural. Whereas intraneural tumors are concentric fusiform lesions found around major peripheral nerves, soft tissue perineuriomas are found in the subcutaneous tissues, most commonly in the extremities and trunk. In a case series of 81 soft tissue perineuriomas, 7 were head and neck lesions.²

On radiographic examination, soft tissue perineuriomas are well-circumscribed masses, more often found in the superficial soft tissues (70%) than in deep tissues.² In our case, the mass showed heterogeneous enhancement and mild bony remodeling with widening of the choanae. Usually, they are not directly associated with a peripheral nerve, though this feature has been described in small lesions³ as was seen in the case of our patient.

Considerations for slow-growing sinonasal masses should include an inverted papilloma, antrochoanal polyp, solitary fibrous tumor, schwannoma, and neurofibroma. A few distinguishing features exist between these lesions and the mass in our patient. Inverted papillomas usually originate in the middle meatus causing an ostiomeatal obstructive pattern. Locally aggressive ones can invade paranasal

tissues, orbits, and the central nervous system. Antrochoanal polyps usually display thin peripheral enhancement and coexisting evidence of recurrent sinusitis. Solitary fibrous tumors of the head and neck most commonly occur in the oral cavity. Ganly et al⁴ reported that the most distinctive feature for these tumors was intense contrast enhancement on CT and MR imaging examinations. Schwannomas may demonstrate intratumoral cysts or a whorled contrast enhancement pattern, whereas neurofibromas may appear as homogeneously low attenuation on noncontrast CT examination.

In cytogenetic studies, perineuriomas show frequent monosomy or deletion of chromosome 22. This characteristic is shared with approximately 70% of sporadic meningiomas and in a small portion of schwannomas.¹ Perineurial cells also share similarities to arachnoid cap cells. Because meningiomas arise from arachnoid cap cells, perineuriomas have been considered by some to be a peripheral relative to meningiomas.³

It is interesting to note that the neurofibromatosis 2 gene is located at 22q11–q13.1, and abnormalities in this region are common in perineuriomas; however, perineuriomas have not been found to be associated with neurofibromatosis.⁵

Surgical resection with clean margins is considered curative. A subset of perineuriomas has been defined as atypical on histologic examination, but this designation has not been shown to change prognosis. Malignant transformation has not been reported. There is an entity known as a malignant peripheral nerve sheath tumor with perineurial differentiation. These are very rare tumors with perineurial immunohistochemical features and malignant cell morphologic changes. From a clinical standpoint, they behave in a malignant fashion but are less aggressive than schwannian malignant peripheral nerve sheath tumors.³

Conclusions

We describe a case of soft tissue perineurioma found in the posterior nasal cavity and nasopharynx that is associated with the left vidian nerve. The imaging features are consistent with a benign, slowly growing soft tissue mass. Imaging is most useful for defining the extent of the lesion. CT provides accurate assessment of the mass and surrounding bony changes. The diagnosis can be established by positive epithelial mem-

brane antigen and negative S100 staining by immunohistochemistry and by the partial or complete deletion of chromosome 22 by fluorescent in situ hybridization.

Acknowledgments

We thank Lester Layfield, MD, and Carlynn Willmore for technical support and Associated Regional and University Pathologists Laboratories for support with immunoassay/fluorescent in situ hybridization.

References

1. Giannini C, Scheithauer B, Jenkins R, et al. **Soft-tissue perineurioma: evidence for an abnormality of chromosome 22, criteria for diagnosis, and review of the literature.** *Am J Surg Pathol* 1997;21:164
2. Hornick J, Fletcher C. **Soft tissue perineurioma clinicopathologic analysis of 81 cases including those with atypical histologic features.** *Am J Surg Pathol* 2005;29:845
3. Macarenco R, Ellinger F, Oliveira A. **Perineurioma: a distinctive and underrecognized peripheral nerve sheath neoplasm.** *Arch Pathol Lab Med* 2007;131:625
4. Ganly I, Patel S, Stambuk H, et al. **Solitary fibrous tumors of the head and neck.** *Arch Otolaryngol Head Neck Surg* 2006;132:517
5. Pitchford C, Schwartz H, Atkinson J, et al. **Soft tissue perineurioma in a patient with neurofibromatosis type 2: a tumor not previously associated with the NF2 syndrome.** *Am J Surg Pathol* 2006;30:1624