Myxoma of the Vomer Bone

David Besachio[1*], Edward Quigley III[1], Richard Orlandi[2], Hugh Harnsberger[1], Richard Wiggins III[1]

1. Department of Radiology, University of Utah, Salt Lake City, USA

2. Department of Otorhinolaryngology, University of Utah, Salt Lake City, USA

* **Correspondence:** David Besachio, D.O., University of Utah Hospital, Department of Radiology, Neuroradiology Division, 30 N 1900 E, Salt Lake City, UT 84132, USA
(*David.Besachio@hsc.utah.edu*)

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ABSTRACT

Myxomas of bone in the head and neck are rare tumors. We present a 68 year old female with pain and epistaxis who was found to have the first reported case of a myxoma arising within the vomer bone. Some atypical magnetic resonance imaging features are described, however, myxoma imaging features are often non-specific and typically evoke a benign differential diagnosis. Surgical excision is the treatment of choice.

CASE REPORT

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A 68 year old non-smoking female without significant past medical history presented to an otolaryngologist following two episodes of epistaxis over several weeks associated with sinus pain. CT was initially obtained, followed by outpatient MRI. CT demonstrated a well-circumscribed, expansile, low density soft tissue mass centered in the posterior aspect of the vomer bone (Figure 1). There was expansile remodeling of the anteroinferior wall of the sphenoid sinus. Coarse internal calcification was noted in addition to erosion of the inferior margin of the vomer. Upon MR imaging, the mass demonstrated intermediate T1 signal intensity, heterogeneously hyperintense T2 signal intensity, with a hypointense internal ring which persisted following administration of gadolinium contrast while the remainder of the tumor enhanced (Figure 2).

The nasal cavity was examined with a 4 mm, 30 degree angled nasal telescope that demonstrated bilateral submucosal fullness of the posterior vomer without surface ulceration.

Differential diagnosis included schwannoma, hemangioma, giant cell granuloma and less likely carcinoma or low-grade chondrosarcoma [1]. Malignant entities such as lymphoma, chondrosarcoma, and squamous cell carcinoma were felt to be unlikely given the relative lack of aggressive imaging features.

Endoscopic excisional biopsy with clear tissue margins was performed with the tumor sent to pathology in three portions. Pathological analysis of the tissue specimen demonstrated a hypovascular sheet of spindle cells in a myxoid stroma compatible with a myxoma (Figure 3). Follow-up imaging and physical examination at 17 months demonstrated no evidence of recurrent tumor.

DISCUSSION

Myxomas of bone are an uncommon benign tumor of connective tissue that is seen almost exclusively in the maxilla and mandible when documented in the head and neck [2]. Their histological origin is uncertain. Head and neck myxomas of bone may sometimes be subdivided into true osteogenic myxomas or odontogenic myxomas, however, this distinction is rarely addressed in the clinical literature as their histological origin remains unclear [3,4,5]. These tumors are characterized by their generally benign, expansile appearance on radiography, however, locally aggressive imaging characteristics have been described as well as a propensity for local recurrence following excision [2,5]. Excision with adequate margins is considered the treatment of choice [2]. These tumors are reported to have a slight female predilection with a wide age range, having been reported from the first through seventh decades.

The CT appearance of myxomas of bone is often non-specific and will evoke a differential diagnosis primarily composed of benign entities such as hemangioma, schwannoma, and giant cell granuloma. A unilocular or multilocular lytic soft-tissue mass with or without internal calcification and a well-circumscribed margin are common. Internal "honey-comb" or "lace-like" bony internal septations, especially in the maxilla, have been described as a helpful feature when trying to separate these lesions from malignant entities [6]. Local cortical disruption has been described but is considered an atypical feature.

On MRI, imaging characteristics are reported as quite variable. Signal intensity on T1 weighted images ranges from homogenously hyperintense to hypointense. T2 weighted tumor hyperintensity is a more consistent finding. In this case, the myxoma was found to have a hypointense internal ring, a previously unreported feature in these tumors. These tumors will typically demonstrate mild-moderate enhancement following administration of contrast, however, imaging characteristics of a myxoma is expected to demonstrate overlap with entities such as a schwannoma, hemangioma, and granuloma. Dynamic contrast enhanced MRI has been reported to be of utility in differentiating these tumors from ameloblastoma when conventional imaging makes this distinction impossible in odontogenic lesions by demonstrating a more gradual, homogenous pattern of enhancement in myxomas [7]. A pattern of peripheral hypointense T2 signal that demonstrates post-contrast enhancement is described in chondromyxoid fibroma, however, the hypointense T2 components of this lesion remained relatively hypointense following contrast administration [8]. A more locally aggressive appearance, intrinsic hyperdensity on CT, or evidence of internal hemorrhage suggests a malignant neoplasm such as chondrosarcoma, lymphoma, and squamous cell carcinoma.

The histological identification of a benign myxoma of bone relies on the presence of characteristic benign appearing spindle cells in a myxoid stroma with the absence of pleomorphic spindle cells arranged in a lobular pattern more characteristic of entities such as a chondromyxoid fibroma or extensive chondroblasts with myxoid liquefaction more commonly seen in chondrosarcomas [9]. The distinction between fibromyxoma and myxoma may be more difficult and likely relies on the relative degree of fibrous and myxoid stroma components [10].

TEACHING POINT

Myxomas arising within the vomer bone have generally benign CT and MR imaging findings that are in keeping with previously reported head and neck myxoma characteristics. These include expansile, often well-circumscribed margins, T2 hyperintensity, and heterogenous enhancement. Although rare, a myxoma of the vomer bone may be considered in the differential diagnosis of benign appearing nasal septal masses.

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FIGURES

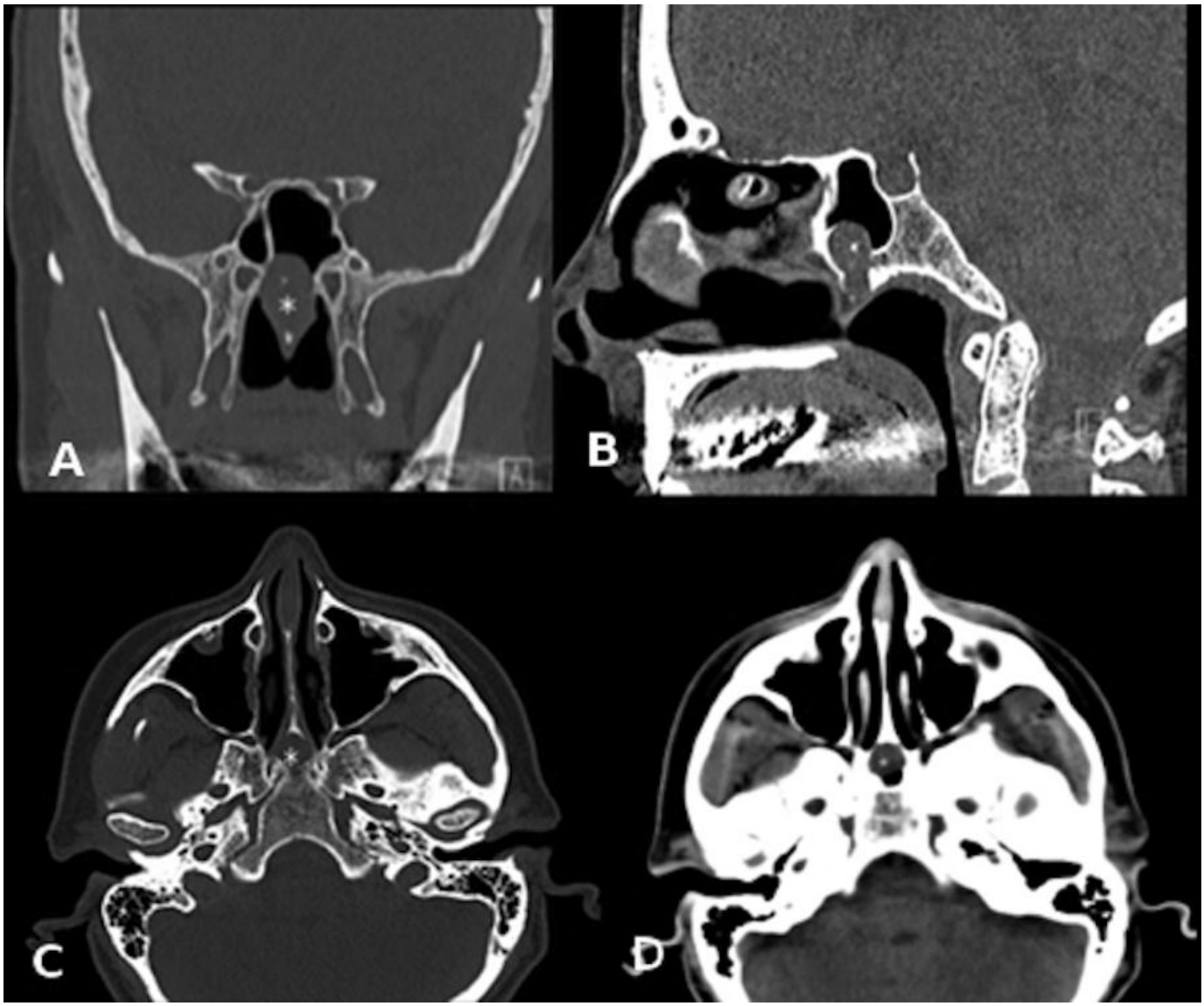

Figure 1: 68 year-old female who presented with nasal pain and epistaxis found to have a myxoma arising within the posterior nasal septum (*). Coronal (A), sagittal (B), and axial (C,D) non-contrast CT scan of the facial bones demonstrating an expansile soft tissue mass arising within the vomer bone with central calcification. (Protocol: 64 multi-detector row, Siemens, kV 120, mAs 300, 1.0 mm slice thickness, helical acquisition)

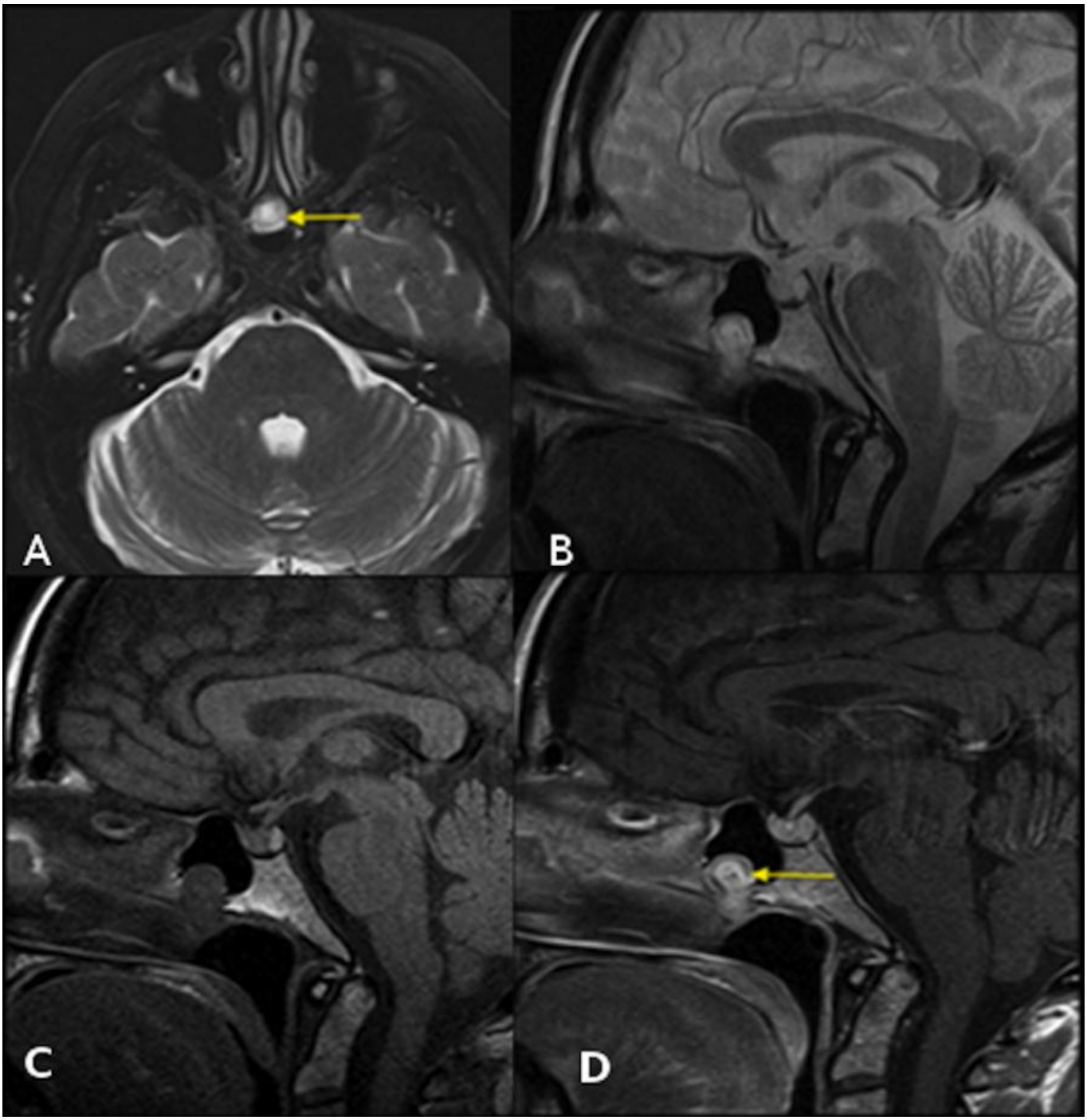

Figure 2: 68 year-old female who presented with nasal pain and epistaxis found to have a myxoma arising within the posterior nasal septum. Axial T2 weighted fat saturated image (A) and sagittal (B) T2 weighted images through the nasal cavity demonstrating a heterogenously hyperintense vomer mass with a peripheral hypointense signal internal ring (yellow arrow) (Protocol: TR 4000 ms, TE 109, thickness 3 mm and TR 4000 ms, TE 53 ms, thickness 3 mm, respectively). Pre- (C) and post-contrast (D) sagittal T1 weighted images demonstrating a heterogenously enhancing mass arising within the vomer bone with hypoenhancing internal ring (yellow arrow) (Protocol: TR 700 ms, TE 14 ms, thickness 3 mm. Gadobenate dimeglumine, 0.2 mL/kg)

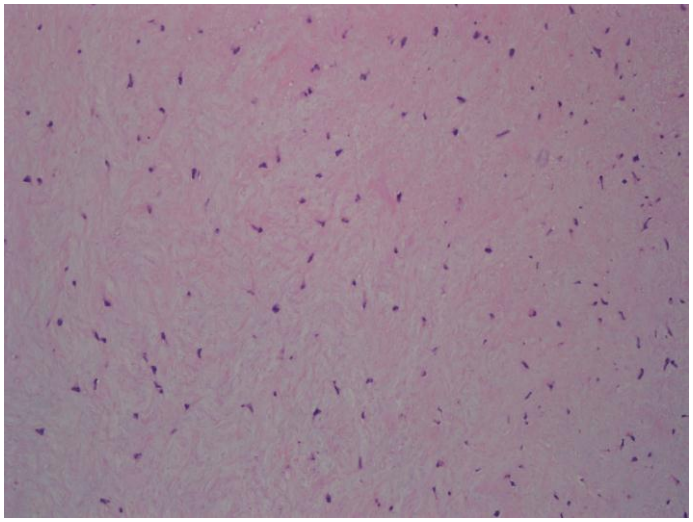

Figure 3 (right): 68 year-old female with a myxoma arising within the posterior nasal septum. High power (40x) photomicrograph of surgical specimen using Hematoxylin & Eosin stain demonstrating an avascular sheet of spindle cells in a myxoid stroma.

Etiology	Benign neoplasm of primitive mesenchymal origin
Incidence	Rare head and neck mass most often felt to have an odontogenic origin
Gender ratio	Slight female predominance
Age Predilection	Most common in 2nd-3rd decade of life
Risk Factors	None
Treatment	Wide local excision
Prognosis	Good; Local recurrence possible if surgical margins inadequate
Findings on Imaging	Variable; Expansile, well-circumscribed mass, typically arising from bone in the head and neck with multiple internal septations. Cortical thinning and disruption may be seen. On MRI the tumors are variable with regard to T1 weighted imaging appearance with T2 hyperintensity and variable patterns of post-contrast enhancement.

Table 1: Summary table for nasal septal myxoma

Entity	CT	MRI T1	MRI T2	Enhancement
Nasal Septum Vomer Myxoma	•Well-circumscribed soft tissue mass isodense to muscle •+/- calcification	•Isointense to hypointense	•Heterogenous hyperintensity •Low signal internal ring	•Heterogenous enhancement
Nasal Septal Lymphoma	•Bulky, lobular soft tissue mass •May be hyperdense to muscle •Bone erosion common	•Homogenous intermediate signal	•Low to intermediate signal	•Typically solid, homogenous enhancement
Nasal Septal Hemangioma	•Isodense to hypodense soft tissue mass with bone erosion	•Low to intermediate signal •Large lesion may demonstrate flow voids	•Hyperintense •Some lesions may show peripheral hypointensity •Large lesions may demonstrate flow voids	•Avid
Nasal Septal Squamous Cell Carcinoma	•Soft tissue mass with adjacent bone destruction common.	•Isointense to muscle •May see T1 hyperintensity with internal hemorrhage	•Low signal often seen	•Heterogenous solid enhancement
Nasal Septal Giant Cell Granuloma	•Heterogenous soft tissue mass •Sometimes with aggressive margins •Occasional intralesional hemorrhage or cysts	•Heterogenous	•Heterogenous	•Markedly variable enhancement
Nasal Septal Chondrosarcoma	•Hypodense soft tissue mass •Osteolysis and bone destruction •50% show chondroid matrix	•Low to intermediate signal	•Heterogenous hyperintense signal	•Heterogenous enhancement
Nasal Septal Schwannoma	•Soft tissue mass with benign margins	•Isointense to muscle	•Slight T2 hyperintensity; some heterogeneity seen	•Solid, mild enhancement •Central portion may show relative hypo-enhancement

Table 2: Differential diagnosis table for nasal septal mass

ABBREVIATIONS

CT = Computed tomography
MRI = Magnetic Resonance Imaging

KEYWORDS

Vomer; Myxoma; Nasal Septum