

# Imaging Characteristics and Outcomes of A New Entity: Low-Grade Sinonasal Sarcoma with Neural and Myogenic Features

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## Abstract

**Objectives:** Low-grade sinonasal sarcoma with neural and myogenic features (LGSSNMF) is a new and rare malignancy that creates a diagnostic dilemma. Our goal is to describe the imaging characteristics and management of this new tumor classification.

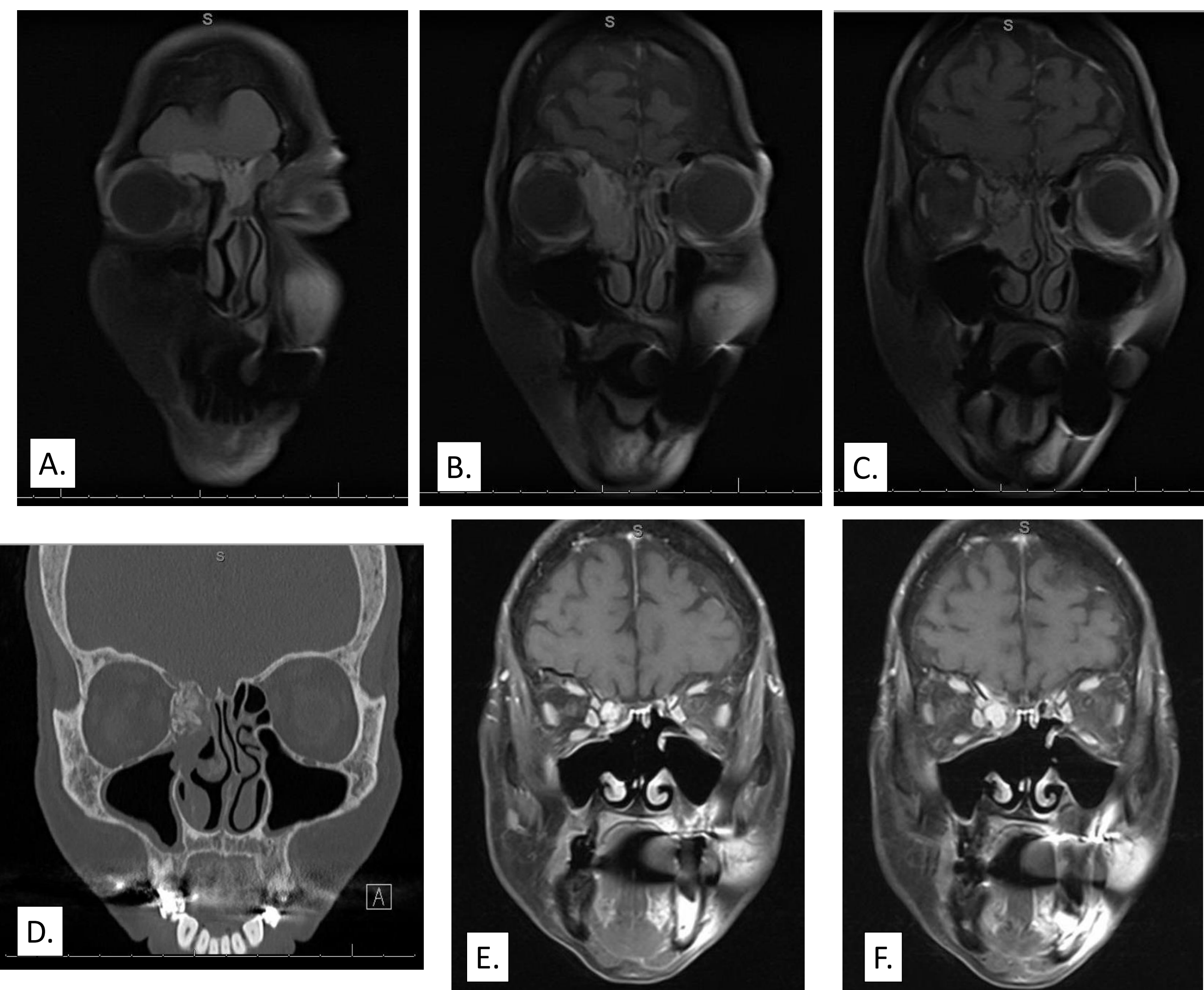
**Methods:** Case series of 3 patients with LGSSNMF.

**Results:** The 1<sup>st</sup> patient presented with diplopia and facial discomfort, the 2<sup>nd</sup> with a supraorbital mass, and the 3<sup>rd</sup> with nasal obstruction. MRI and CT imaging in each case showed an enhancing sinonasal mass with associated hyperostotic bone formation. Biopsy confirmed LGSSNMF. 2 patients underwent surgical resection, while the last patient is pending treatment. The 1<sup>st</sup> patient recurred after 17 months and required a repeat resection.

**Conclusions:** The imaging and surgical management of LGSSNMF is described to further characterize this unique, locally-aggressive tumor.

## Introduction

- Sarcomas of the head and neck are rare entities accounting for approximately 3% of head and neck malignancies.
- Sinonasal sarcomas are even rarer and create a diagnostic and treatment dilemma, particularly due to their varied tumor histologies.
- Review of the SEER database showed 5-year survival was 47% for all sinonasal sarcomas, and increasing age, male sex, frontal and maxillary sinus subsites, and rhabdomyosarcoma and Kaposi sarcoma histologies were associated with a significant increase in mortality<sup>1</sup>.
- In 2012 a new tumor was described in this category: Low-grade sinonasal sarcoma with neural and myogenic features (LGSSNMF) with 28 cases review at the Mayo clinic<sup>2</sup> and 1 other case reported in 2015<sup>3</sup>.
- This is a locally aggressive tumor with a propensity for recurrence.
- All cases showed a characteristic histology: uniform spindle cell neoplasm with elongate nuclei, neural and myogenic differentiation, and expression of S-100 and most with actin positivity (96%).
- Most tumors involved the nasal cavity (54%) or the ethmoids (57%).
- There was a 3:1 female to male ratio and age ranged from 24 to 85 years old (mean – 52 years).
- Follow-up was available for 16/28 patients (57%), mean of 8.3 years, and 7/16 (44%) developed a recurrence and no patient has developed metastases or died of this disease.



**Figure 1.** Patient #1 A. – C. Coronal post-contrast fat-saturated T1 MRI pre-op images showing her right sinonasal mass centered at the frontal ethmoidal recess with minimal enhancement. D. Coronal CT pre-op image in bone window showing the mass with hyperostotic bone formation. E. and F. Coronal post-contrast fat-saturated T1 MRI images showing her recurrence adjacent to the skull base and medial to the right medial rectus muscle with avid homogeneous enhancement.

## Methods and Materials

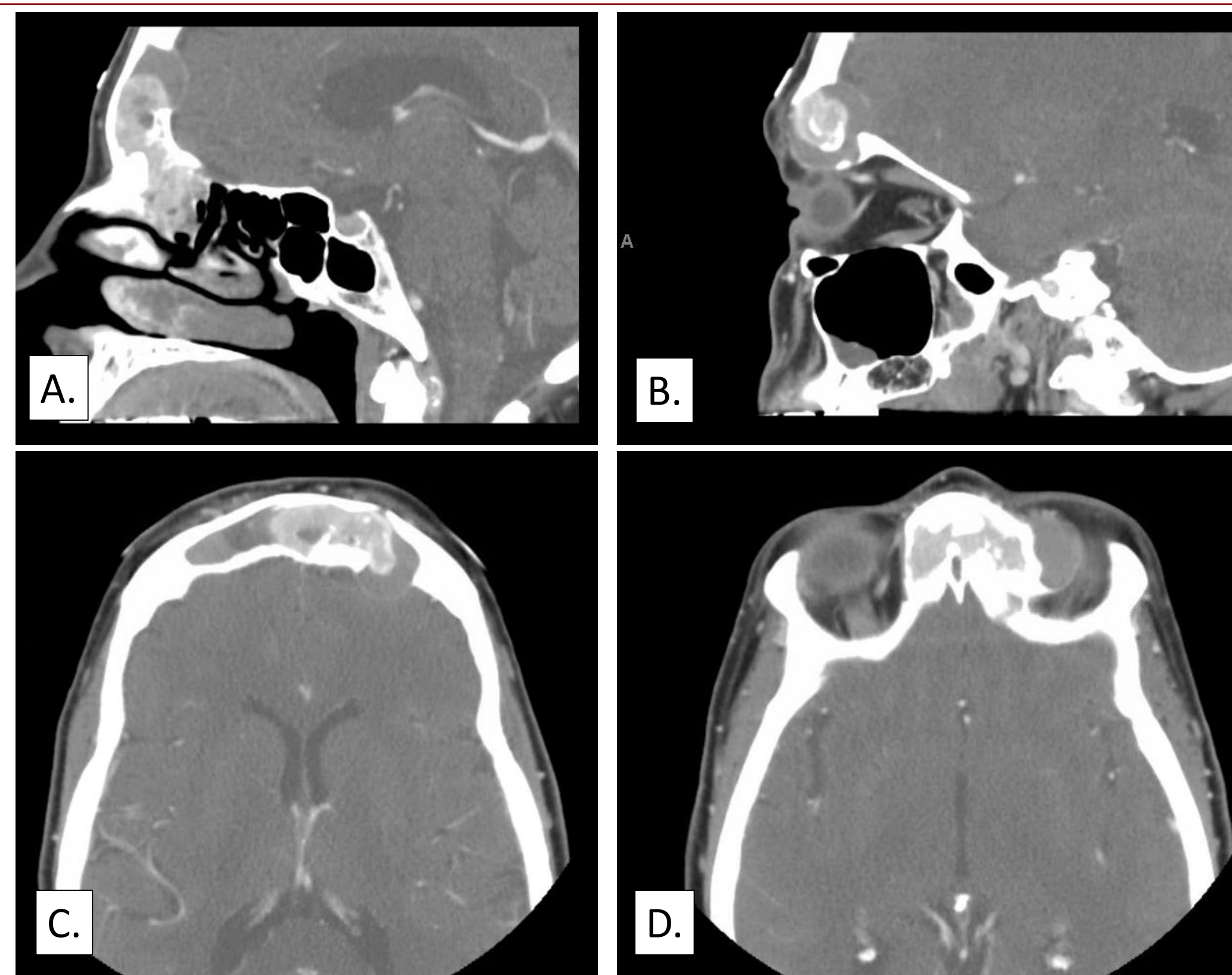
- Case series of 3 patients treated at the University of Utah with low-grade sinonasal sarcoma with neural and myogenic features.

### Patient #1

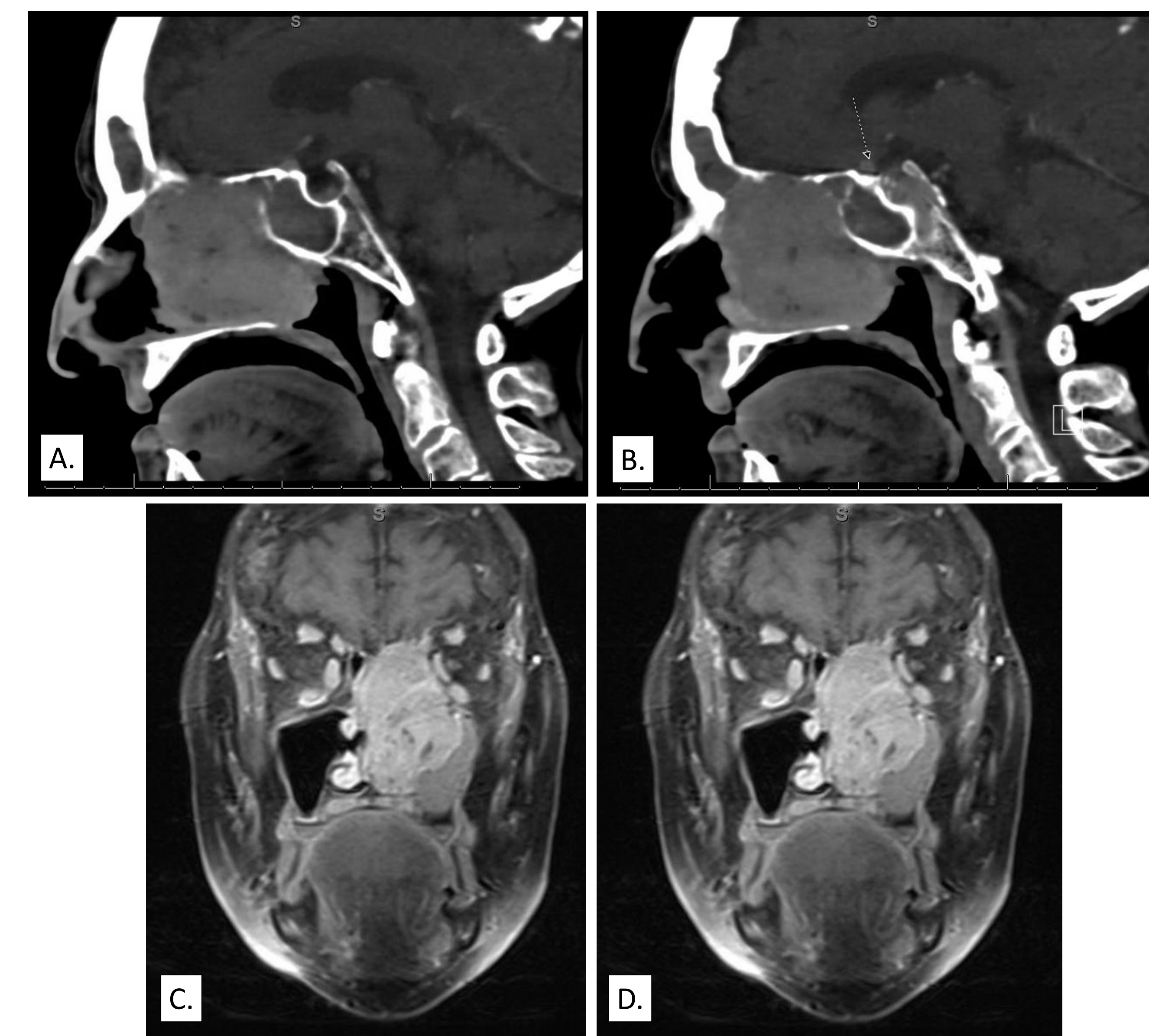
- 67 year old female with diplopia and facial discomfort.
- CT and MRI imaging identified a sinonasal mass in the right frontal recess with extension into the ethmoids, intimately associated with the lamina papyracea with hyperostotic bone formation (**Figure 1**).
- She underwent an endoscopic biopsy that confirmed a spindle cell neoplasm and she underwent an endoscopic anterior skull base resection with removal of the lamina papyracea.
- Surgical pathology showed a mucosal lesion comprised of bland spindle cells without mitosis, necrosis, or pleomorphism, but the neoplastic proliferation was infiltrative into bone.
- It stained focally positive for S100, smooth muscle actin, and desmin and it was negative for CD34, Sox-10, EMA, and cytokeratin AE1/AE3. It was evaluated for SS18 (SYT) gene translocation at location 18q11, which was negative, and thus, ruled out a synovial cell sarcoma.
- Final diagnosis was a LGSSNMF.
- She recovered well, without morbidity, but recurred 17 months later and underwent an endoscopic repeat resection with removal of the adjacent anterior skull base and dura due to involvement demonstrated on frozen section. Her intra-operative CSF leak was repaired, and again she recovered well, without long-term morbidity.

### Patient #2

- 62 year old female who presented with a slowly progressive left supraorbital swelling.
- CT and MRI imaging shows a T1 enhancing, T2 hypointense mass in the left frontal sinus with hyperostotic bone formation that had eroded through the adjacent bone of the posterior table, contacting the dura, and pushing the frontal lobe posteriorly. It also extended inferiorly into the ethmoids, eroded the lamina papyracea, and contacted the left medial rectus. A post-obstructive frontal sinus mucocele had eroded the superior orbital roof (**Figure 2**).
- She underwent an endoscopic biopsy that showed LGSSNMF, and she underwent a bicoronal approach to the anterior skull base for resection.
- Final pathology showed a tumor focally eroding the bone comprised of monotonous spindle cells without cytologic atypia that were positive for S100 and SMA and negative for desmin, MYF4, and the SYT gene translocation.
- The final diagnosis was LGSSNMF, and she also recovered well, without morbidity.



**Figure 2.** Patient #2 A. and B. Sagittal and C. and D. Axial post-contrast CT pre-op images showing the frontal sinus mass with hyperostotic bone formation and erosion through the posterior table with intracranial extension.



**Figure 3.** Patient #3 A. and B. Sagittal and C. and D. Coronal post-contrast fat-saturated T1 MRI images showing her left sinonasal mass centered in the ethmoid cells with avid homogeneous enhancement and extension through the skull base.

### Patient #3

- 79 year old female who presented with nasal obstruction and left facial pressure.
- CT and MRI imaging shows a T1 enhancing, T2 hypointense mass with hyperostotic bone formation of the left ethmoid sinuses that has eroded through the adjacent lamina papyracea and anterior skull base and contacted the medial rectus and the dura (**Figure 3**).
- She underwent an endoscopic biopsy that showed LGSSNMF.
- It stained positive for S100, SMA, and desmin and it was negative for CD34, cytokeratin AE1/AE3, and muscle specific actin. SYT gene translocation was negative.
- She was found to have severe aortic stenosis in her work-up for surgery and recently underwent a transcatheter aortic valve replacement.
- She is recovering and still pending treatment.

## Discussion

- Low-grade sinonasal sarcoma with myogenic and neural elements (LGSSNMF) is a very rare tumor that is locally aggressive has a propensity for recurrence with 29 prior cases reported in the literature<sup>2,3</sup>.
- We report 3 more cases and review the imaging characteristics of these tumors which has not been previously discussed.
- All our cases showed the characteristic histology with uniform spindle cell neoplasm with elongate nuclei, neural and myogenic differentiation, expression of S-100 and SMA, and negative SYT gene translocation.
- All tumors enhanced on MRI, had hyperostotic bone formation on CT scan, had eroded through the skull base with extension up to the dura, but not through it, and had eroded through the lamina papyracea.
- These 3 cases continue to expand on the common clinical features: occur in the elder (Avg. age – 69 years), female predominant (all 3), infiltrative, particularly into bony structures (all 3), involve the ethmoids (all 3), frontal sinus (2/3), and can recur (1/2; mean follow-up of 1.5 years), but no patient has developed metastases or died of this disease.

## Conclusions

- Low grade sinonasal sarcomas with myogenic neural elements are a new, rare entity and their diagnosis, imaging, and treatment is reviewed.

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