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CASE REPORT

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Multimodality management of sinonasal teratocarcinosarcoma in a 76-year-old Alaska Native female during the COVID-19 pandemic

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Abstract

Sinonasal teratocarcinosarcoma is a rare, highly aggressive tumor of the anterior skull base composed of malignant epithelial, mesenchymal, and neural tissue. Examination of cases in patients in minority populations is important in order to better understand the behavior of this neoplasm and outcomes of treatment in our nation's diverse population.

KEYWORDS

anterior skull base malignancy, cross-cultural medicine, sinonasal teratocarcinosarcoma

1 | INTRODUCTION

Sinonasal teratocarcinosarcoma (SNTCS) is a rare and highly aggressive tumor of the anterior skull base. Pathologically heterogeneous in nature, it is composed of epithelial, mesenchymal, and neural tissue. We present a case of SNTCS that diverges demographically from the majority of published reports and discuss the unique challenges of caring for this patient with a rare, highly aggressive sinonasal malignancy during the early days of the COVID-19 pandemic.

2 | CASE REPORT

A 76-year-old Alaska Native woman was referred to Otolaryngology with a history of worsening left nasal obstruction and recurrent epistaxis for several months. She complained of weight loss, decreased appetite, impairments in smell and taste, bifrontal headaches, and left facial pressure. Anterior rhinoscopy demonstrated a necrotic, bulging mass in the left nasal vestibule completely obstructing the nasal airway and precluding passage of an

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endoscope. Imaging including CT and MRI showed a heterogeneous 2 cm × 1.8 cm × 4.5 cm sinonasal mass with extension into the nasopharynx. Preoperative biopsy was consistent with a high-grade malignant neoplasm.

Our patient's care is notable in its timing: Her biopsy was performed in February 2020, shortly before the COVID-19 disease pandemic interrupted healthcare delivery worldwide. She was unable to receive care locally or regionally due to slowdowns in clinical operations in nearby centers. Given the highly aggressive nature of this rare malignancy, she opted to travel nearly 3000 miles by airplane in early April 2020 for definitive treatment. New imaging at that time (CT and MRI, Figure 1) demonstrated interval enlargement of the sinonasal mass (now 7 cm) with erosion of the left medial orbital wall and ethmoid roof. The mass was hypermetabolic on ^{18} FDG-PET, but there was no evidence of metastatic disease. Examination was notable for the absence of vision changes, cranial nerve abnormalities, or palpable neck lymphadenopathy.

After multidisciplinary tumor board recommendations, she underwent image-guided endoscopic resection of the tumor including posterior septectomy and resection of the olfactory bulb area, and bilateral neck dissection (levels I–III) (Figure 2). Preoperative embolization was not performed as this tumor was not vascular in nature on imaging, the patient's epistaxis was intermittent and not severe, and thus, concern for hemorrhage was low. Intraoperatively, the mass was necrotic but well circumscribed and did not invade the floor of the nose, the anterior septum, or the lateral nasal wall. We removed the mass with a microdebrider beginning anteriorly and following it posteriorly to its origin at the face of the sphenoid near the superior turbinate and involving the posterior septum. Margins were taken (superior turbinate, face of the sphenoid, periorbital posterior ethmoid cells, and the septum); all were negative except for the posterior superior septum and face of the sphenoid, its likely origin. We then performed a posterior septectomy and removed the entire anterior wall of the sphenoid, after which final margins were negative. Reconstruction utilized a superiorly based nasal septal flap pedicled on the anterior ethmoid artery because the traditional posterior septal artery-based flap was not possible due to tumor involvement. Pathology revealed epithelial elements composed of immature tubuloglandular structures, mesenchymal elements ranging from highly cellular round blue cells to short spindle cells, and a primitive neural component, consistent with SNTCS (Figure 3). All lymph nodes were negative for tumor. Her

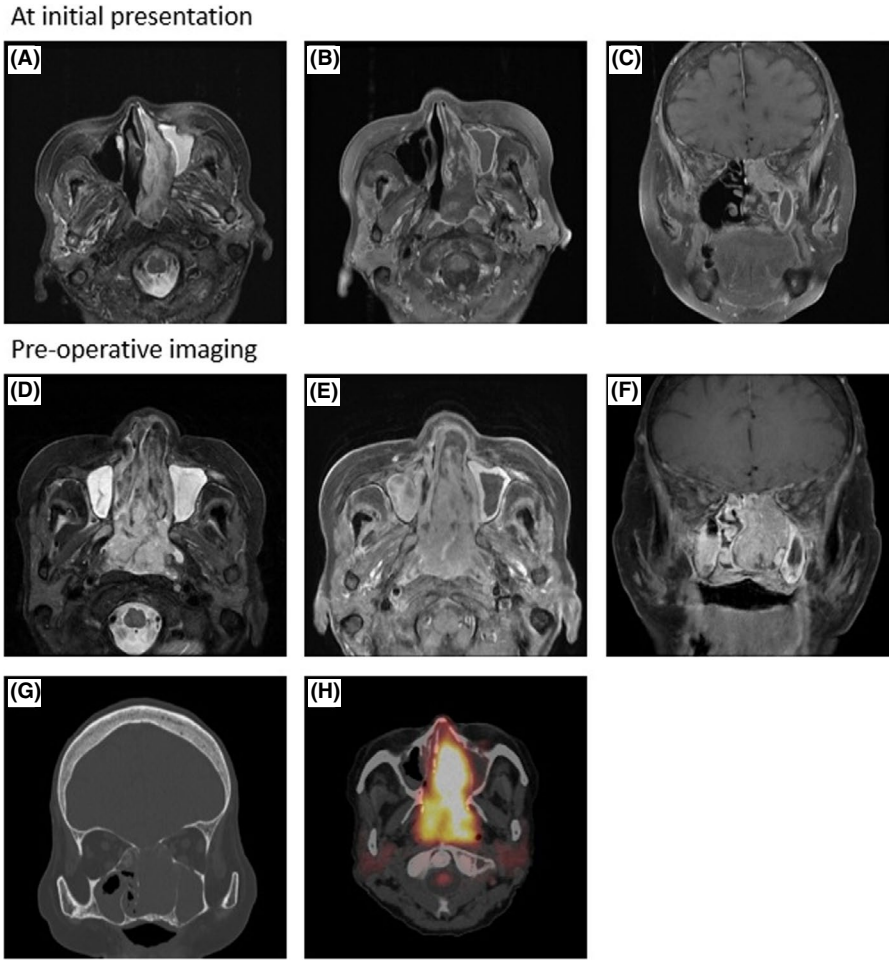

FIGURE 1 At initial presentation, axial fat-suppressed T2-weighted (A), axial fat-suppressed postcontrast T1-weighted (B), and coronal fat-suppressed postcontrast T1-weighted (C) MR images show a heterogeneous left sinonasal mass with extension into the nasopharynx and associated obstructed secretions in the left maxillary sinus. Two months later, axial fat-suppressed T2-weighted (D), axial fat-suppressed postcontrast T1-weighted (E), and coronal fat-suppressed postcontrast T1-weighted (F) MR images show interval enlargement of enhancing sinonasal tumor. Coronal CT image (G) shows erosions in the left medial orbital wall and ethmoid roof associated with the sinonasal mass. Axial^{18} FDG-PET/CT imaging (H) shows that the sinonasal mass is markedly hypermetabolic

recovery was uneventful, and she was discharged. She returned home 10 days later, where she completed adjuvant radiation to the primary site (6300 cGy in 35 fractions) in August 2020. She has since been disease free for 12 months (Figure 4).

3 | DISCUSSION

Sinonasal teratocarcinosarcoma is a rare and highly aggressive tumor, with 127 reported cases since 1966.[1] The largest published series from a single institution includes 10 patients treated over 35 years at a major cancer center.[2]

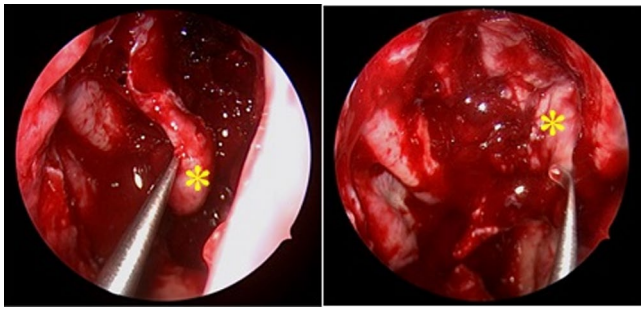

FIGURE 2 Intraoperative images of the reconstruction utilizing a superiorly based nasal septal flap pedicled on the anterior ethmoid artery. *Nasal septal flap

Prognosis is poor, with a 2-year survival rate of 55% and a recurrence rate of 38%.[1] The majority of cases involve men (83%), and age at diagnosis ranges from 10 to 82 years old (mean 50). Information on race and ethnicity is not consistently available, but the largest case series included 8 white and 2 Hispanic patients.[2] Given the rarity of this diagnosis, these demographic characteristics may not be representative. Disparities in head and neck cancer outcomes among Alaska Natives may exist. Thus, it is important to share the experience of our patient, a 76-year-old Alaska Native woman.

Patients commonly present with epistaxis (76%), nasal obstruction (62%), and frontal headaches (20%).[3] Tumors are oftentimes large (average 5.3 cm) and are frequently located in the nasal cavity (72%), ethmoid sinus (53%), and maxillary sinus (31%). A significant percentage of patients have intracranial extension, cribriform plate, and anterior cranial fossa involvement (21%). Diagnosis of SNTCS can be challenging given histopathological heterogeneity, and reports of initial misdiagnosis exceed 50%.[3] Diagnosis of SNTCS requires identification of at least one malignant epithelial component and two malignant mesenchymal components, such as fibroblasts, cartilage, bone, or smooth muscle.[3] Additionally, 50–75% of tumors contain "fetal appearing" clear cell squamous epithelium that can aid in diagnosis. *SALL4*, normally expressed in germ cell tumors, has emerged as a potential useful

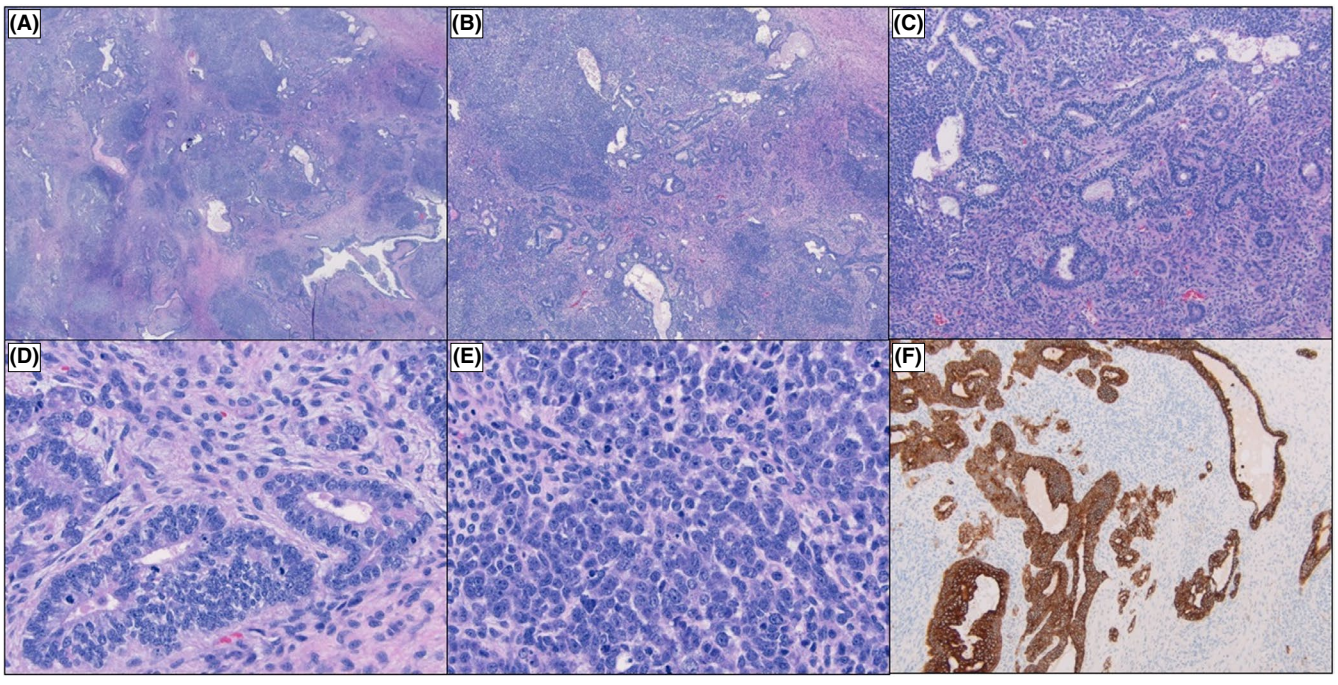

FIGURE 3 Histologic slides of hematoxylin and eosin stains showing (A–C) highly cellular neoplasm with two distinct morphologies: solid sheets of blue cells (mesenchymal area) and tubuloglandular structures (epithelial area). Higher power (×400) mitotically active tubular epithelial component (D) and sheet-like blue cell component (E). (F) Higher power Keratin stain (AE1/AE3, ×100) highlighting the epithelial component. Not shown: small neural component consisting of immature neuroblasts

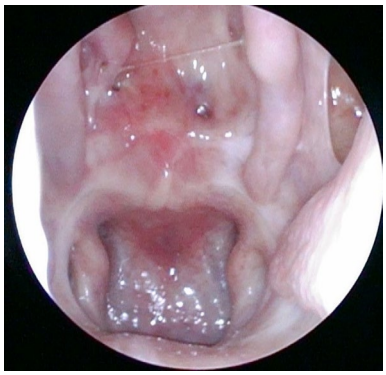

FIGURE 4 Postoperative endoscopic examination of nasal cavity at follow-up 10 months after completion of radiation

marker in confirming the diagnosis.[4] The molecular underpinnings of SNTCS are not fully elucidated; however, some studies demonstrate loss of BRG1 expression (up to 82%), which corresponds to biallelic somatic inactivation of the *SMARCA4* gene, which is involved in chromatin remodeling.[5]

Multimodal therapy is the most common treatment paradigm, with the majority of patients receiving surgery and adjuvant radiation (62%) or surgery and adjuvant chemoradiation (28%).[1] Surgery with adjuvant chemoradiation appears to demonstrate survival benefit compared to surgery with adjuvant radiation, which has improved survival compared to surgical resection alone. For patients with unresectable tumors, treatment with neoadjuvant chemotherapy followed by surgical resection has been performed.

The timing of our patient's diagnosis is notable for overlap with the onset of the COVID-19 disease pandemic, when many hospitals nationwide postponed surgeries and airlines canceled flights. Concerns included increased perioperative risk in infected patients and risk of transmission to the surgical team, particularly during aerosol generating procedures. Indeed, surgical de-escalation of neck management and reconstruction in head and neck cancer occurred during the early period of the pandemic. Despite these challenges and the significant distance that our patient travelled for treatment, she was able to receive timely management. Given our understanding of how this neoplasm behaves, it is possible that expeditious treatment despite the challenging external factors might impact her long-term outcome. Such burdens may affect future patients.

4 | CONCLUSIONS

Sinonasal teratocarcinosarcoma is a rare, highly aggressive tumor of the anterior skull base composed of malignant epithelial, mesenchymal, and neural tissue. Patients oftentimes present with epistaxis and nasal obstruction. Diagnosis can be challenging given histopathologic heterogeneity. Multimodality treatment is favored. Given the rarity of this diagnosis, examination of cases in patients in minority populations is important in order to better understand the behavior of this neoplasm and outcomes of treatment in our nation's diverse population.

ACKNOWLEDGMENT

None.

CONFLICT OF INTEREST

None.

AUTHOR CONTRIBUTIONS

DRA, RRG, JMP, and BLC were involved in the draft writing. DTG was involved in imaging review and preparation of article images. NAC was involved in histopathologic review and preparation of article images. All authors critically revised the report, commented on drafts of the manuscript, and approved the final report.

ETHICAL APPROVAL

This case report is exempt from the University of Chicago IRB review.

CONSENT

Written consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

DATA AVAILABILITY STATEMENT

Data available on request due to privacy/ethical restrictions.

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