Two cases of sinonasal teratocarcinosarcoma: Confounders, treatment, and review of the literature

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Abstract

Introduction: Sinonasal teratocarcinosarcoma (SNTCS) is an extremely rare and aggressive tumor of the anterior skull base, with less than 100 cases reported in the literature, only five of which are pediatric. Due to its rarity, the long term prognosis of this tumor is difficult to assess. Standard treatments include multimodality treatment with significant morbidity. We present two cases of SNTCS. The first is a 70-year-old male resected via combined open and endoscopic approach. The second is a 16-year-old male, the first reported patient resected through an endoscopic-only approach.

Case 1: 70-year-old male presented with epistaxis and anosmia and was noted to have a sinonasal mass with right sided lypophadenopathy. He was presumed to have adenocarcinoma and was taken to the OR for combined open and endonasal approach and resection of tumor. Pathologic analysis confirmed SNTCS. Patient was treated with adjuvant chemoradiation. He would have recurrence in his neck, which was treated with surgical excision. He remains disease free 14 months out of surgery.

Case 2: 16-year-old Hispanic male who presented with nasal obstruction. A CT scan revealed a large sinonasal mass with bone remodeling and MRI confirmed a heterogeneously enhancing mass filling the posterior sinonasal region. Multiple biopsies were taken to obtain a diagnosis, which remained ambiguous. He was eventually taken for total endoscopic resection after outside pathology confirmed SNTCS. He remains disease free after chemotherapy.

Conclusion: SNTCS is a rare, invasive sinonasal tumor with five-year survival around 20%. Pathologic diagnosis is difficult given mixed cell lines on specimen. High clinical suspicion in the male patient, particularly when pathologists struggle with obtaining a diagnosis, is key. Metastatic work up is warranted, and aggressive resection with adjuvant chemoradiation is necessary.

Introduction

Combining the histopathological features of carcinomas and malignant teratomas, sinonasal teratocarcinosarcomas (SNTCS) are rare, aggressive tumors of the anterior skull base and sinonasal cavities. Patients are usually male and are diagnosed in their fifth decade of life.1,2 Treatment is usually multimodal involving surgical resection and radiotherapy, but may be difficult secondary to aggressive tumor spread, intracranial extension, and high rate of recurrence.

There have been less than 100 cases of SNTCS described in the literature, and of these, five have been reported as pediatric cases. We present two cases of SNTCS: one involving a 70-year-old male and one involving a 16-year-old male, the latter having the first endoscopic-only resection of his skull base tumor.

Case 1

A 70-year-old male with hypertension and hyperlipidemia as well as a several month history of epistaxis, anosmia, and vision changes in the right eye presented to our clinic after being seen at his local emergency department (ED) with massive epistaxis. Outside ED performed a computed tomography (CT) scan of the maxillofacial region with contrast that demonstrated a large, right-sided skull base mass with erosion of the cribiform with intracranial, orbital, and slight contralateral involvement (Fig. 1a). Subsequent CT neck demonstrated right-sided lymphadenopathy. Head and neck exam was normal save for prototypical right eye with restricted motion. Nasal endoscopy demonstrated obvious right-sided tumor of the entire superior sinonasal cavity with left-sided involvement superiorly through the septum into the ethmoid sinuses. Subsequent MRI confirmed intracranial extension with vanishing edema (Fig. 1b).

Given the concern for malignancy, the patient was taken to the operating room for nasal endoscopy with biopsies that returned as poorly differentiated adenocarcinoma with necrosis and sinonasal adenocarcinoma (colonic type) with neuroendocrine differentiation/component. Because of the uncertainty with diagnosis, labs were sent out to Mayo Clinic and returned as SNTCS. PET scan confirmed activity within the right neck.

Once deemed suitable for surgery, the patient underwent bilateral craniotomy with combined transnasal endonasal approach and resection of the tumor, right orbital exenteration, and radical neck dissection with sacrifice of cranial nerve XII secondary to tumor invasion. Frozen margins were negative for malignancy and patient’s skull base defect was reconstructed using a periarticular iliac, Alloderm, and sealant. His post-operative course was uneventful.

Surveillance scans 7 months after surgery demonstrated right level II lymph node concerning for malignancy, and he underwent resection of his neck dissection. The patient is now 14 months out of surgery, 13 months out of radiation, and 2 months out of chemotherapy with no evidence of disease.

Case 2

In this case series, we discuss two cases of SNTCS, one involving a 70-year-old male resected via combined external and endoscopic approach, and a second involving a 16-year-old male resected through endoscopic approach alone. The latter patient is the first report of endoscopic only approach with no external incision. As of writing this manuscript, both patients remain disease free with morbidity only related to metastatic disease.

To date, less than 100 cases have been published in the literature. Typically, these tumors present in the 4th to 5th decade of life and have a 7:1 male predilection.3 Five-year survival is 20%.4 The most common symptoms at presentation are nasal obstruction, epistaxis, and headache.5 Treatment usually entails surgical resection with radiotherapy. In a retrospective systematic review, patients receiving chemotherapy and radiation fared better, but the study is limited by poor power and limited follow up.1 Both of our patients underwent surgical resection with chemotherapy. Metastatic disease is common to the neck, and was seen in our first patient. A single institution review demonstrated metastatic nodal disease in 23% of patients.7 Metastatic disease to the lung, brain, and dura has also been reported.8,9

Our second patient was sixteen years old at the time of his diagnosis making him the fifth pediatric patient diagnosed with SNTCS.9 All of these patients were male, and reportedly treated with surgery in combination with radiation and/or chemotherapy. Given the aggressive nature of these tumors, and the difficulty in obtaining truly negative margins, adjuvant chemoradiation seems most appropriate despite the long-term effects of these therapies.

One of the difficulties we had in treating these patients was obtaining a suitable pathologic diagnosis despite obtaining adequate specimen. This is understandable given both benign and malignant epithelial, mesenchymal, and neuroepithelial components. SNTCS is thought to have a 50% initial identification rate with imaging. A previous case series demonstrated that adenocarcinoma was the primary epithelial component in all tumors in their study.7 This was similar to our study, as our first case was initially diagnosed as adenocarcinoma, and our second case also had components of adenoacarcinoma on final specimen. Stains were of little use, as they bound to the corresponding cell line within the tumor. The distinguishing factor of these tumors is that they lack germ cell elements.5

Conclusion: SNTCS is a rare, invasive sinonasal tumor with five-year survival around 20%. Pathologic diagnosis is difficult given mixed cell lines on specimen. High clinical suspicion in the male patient, particularly when pathologists struggle with obtaining a diagnosis, is key. Metastatic work up is warranted, and aggressive resection with adjuvant chemoradiation is necessary.

References