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Case Report

Management of a pediatric patient with aggressive low-grade sarcoma of the maxillary sinus: Case report and literature review

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ABSTRACT

Pediatric sinonasal tumors are characterized by histological diversity, nonspecific clinical presentations, and variable recurrence and metastatic potential. We describe a rare low-grade, undifferentiated sarcoma with bland/benign histopathology but an aggressive course in a 10-year-old boy with nasal obstruction, rhinorrhea, and left eye proptosis. Imaging revealed a large sinonasal soft tissue mass completely obstructing the left nasal cavity, without intraorbital involvement or distant metastasis. The mass stained positive for S100, vimentin, glial fibrillary acidic protein, and Ki-67 and negative for desmin and EGFR (epidermal growth factor receptor). The patient underwent neo-adjuvant chemoradiation therapy, and definitive surgical resection via a left lateral rhinotomy/maxillectomy approach. Post-treatment results were excellent, and the patient has remained disease free at one-year follow-up based on clinical, radiographic, and direct endoscopic visualization examinations with minimal morbidity or functional disability.

1. Introduction

Head and neck malignancies constitute 5% of all pediatric primary cancers; of these, sinonasal sarcomas represent 16.2% [1]. Common presenting symptoms are epistaxis, headache, sinusitis, nasal congestion, and nasal obstruction. Rare but more ominous signs include proptosis, diplopia, and cranial nerve dysfunction, suggesting more aggressive or advanced-stage tumors [2].

Based on the pediatric SEER (surveillance, epidemiology, and end results) database published by the National Cancer Institute, between 1988 and 2005 only a small number of children ($n = 63$) were identified as having a sinonasal malignancy. Rhabdomyosarcoma was the most common type ($n = 27$, 43%) and the most common sinonasal location was the maxillary sinus ($n = 38$, 68%) [2,3]. Unspecified or undifferentiated sinonasal sarcomas are extremely rare among pediatric sinonasal sarcomas [4].

Because of the rarity and diversity of these mesenchymal/connective tissue sarcomas, a patient with an unspecified or undifferentiated sinonasal sarcoma presents a diagnostic and management challenge [1]. Various studies, biomarkers, and genetic characterization have been developed in the past four decades. Differentiating histologic features such as fibroid, myxoid, stellate, and spindle cell combined with the immunohistochemistry (IHC) markers such as S100, vimentin, glial fibrillary acidic protein (GFAP), Ki-67, and desmin, and EGFR are often used to aid clinicians in determining the origin or type of cell-line that these soft tissue tumors derived from and determine a treatment regimen appropriate for targeted chemo- or radiation therapy. Since there is lack of standard characterization or IHC profiles for these undifferentiated soft tissue malignancies, the pathologist plays a primary role in establishing the correct diagnosis to differentiate these clinically aggressive soft tissue malignancy in the sinonasal tract [5]. Table 1 is the summary generated from Googe et al. [6].

Here we present a case study and examine the current evidence regarding management of low-grade undifferentiated/uncharacterizable pediatric sinonasal sarcoma. We compare the options available for managing this tumor to maximize patient survival and minimize morbidity. The Drexel University College of Medicine institutional review board granted a waiver for this study. The patient's family consented to the presentation of this case and informed consent was obtained.

2. Case presentation

A 10-year-old boy presented to our otolaryngology clinic with a 1-month history of left-sided nasal obstruction and orbital proptosis. The patient reported headaches, diplopia, rhinorrhea,

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Table 1
Summary of possible interpretations of various immunohistochemistry (IHC) antibody stains [6].

Immunochemical antibody stain	Possible source
S-100	Melanomatous, epithelial, histiocytoma, clear cell sarcoma, or neural crest cells origin
GFAP (glial fibrillary acidic protein)	Glial cell origin
Vimentin	Mensenchymal origin
Ki67	Cytokeratinous, epithelial, melanomatous origin
EGRF (epidermal growth factor receptor)	Epithelial origin
Desmin	Smooth muscle (leiomyo) origin

left malar hypoesthesia, and intermittent epistaxis. On physical examination, his left eye was proptotic with disrupted conjugate gaze. Nasal endoscopy revealed a solid, left-sided nasal mass. Immediately following his outpatient consultation, the patient was sent to the emergency department for urgent imaging and admission for further work-up.

Contrast-enhanced computed tomography (CT) demonstrated a large, well-circumscribed heterogeneous mass containing an unerupted maxillary molar in the left maxillary antrum. Superior extension of the mass caused displacement of the left orbital floor and inferior rectus muscle. The optic nerve was uninvolved and the extraocular muscles were unremarkable. No bony erosion was identified.

Multi-sequence magnetic resonance imaging (MRI) of the brain, with and without contrast enhancement, demonstrated a lobulated mass within the left maxillary sinus displacing the medial wall and the orbital floor (Fig. 1). The mass appeared to originate from the left maxillary first molar with hypointense signal on T1-weighted imaging and hyperintense signal on T2-weighted and on fluid-attenuated inversion recovery imaging. Orbital structures were intact and intracranial extension was not observed.

Following the imaging work-up, biopsy specimens were obtained endoscopically under image guidance. Frozen sections revealed benign glandular epithelium, fibroepithelial cells, and small, atypical cells on a myxoid background (Fig. 2). The permanent sections demonstrated myxomatous soft tissue with spindle-shaped and stellate cells. Immunochemistry staining was positive for S100 (Fig. 3), vimentin (Fig. 4), GFAP (Fig. 5), and Ki-67

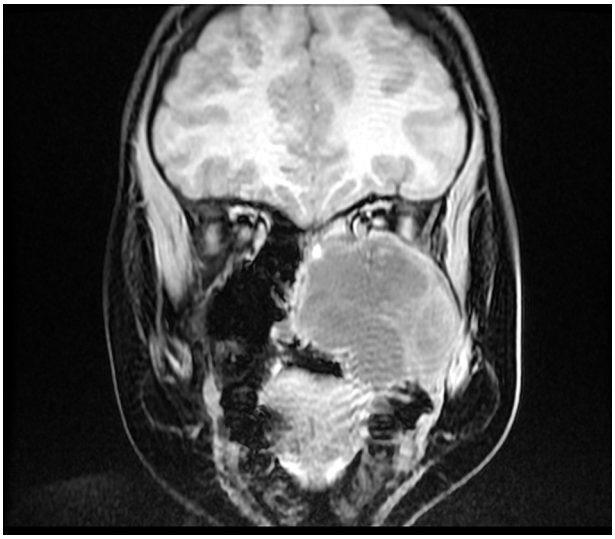

Fig. 1. T1-weighted coronal magnetic resonance image of the face and orbit showing a left maxillary sinonasal tumor.

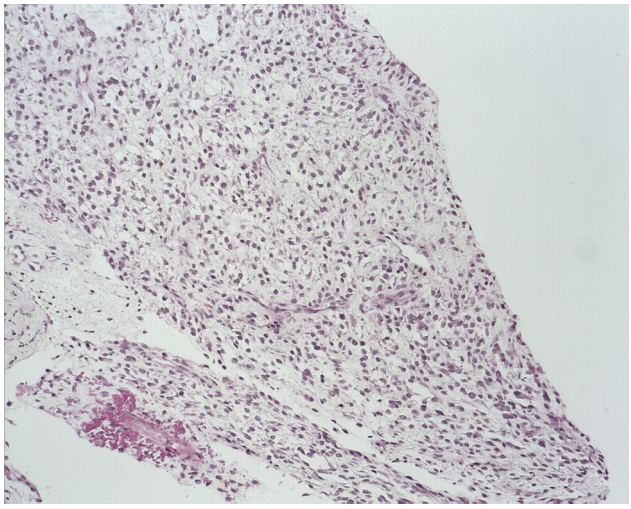

Fig. 2. Standard hematoxylin and eosin stained histopathologic sample showing a myxoid background with atypical cellular areas.

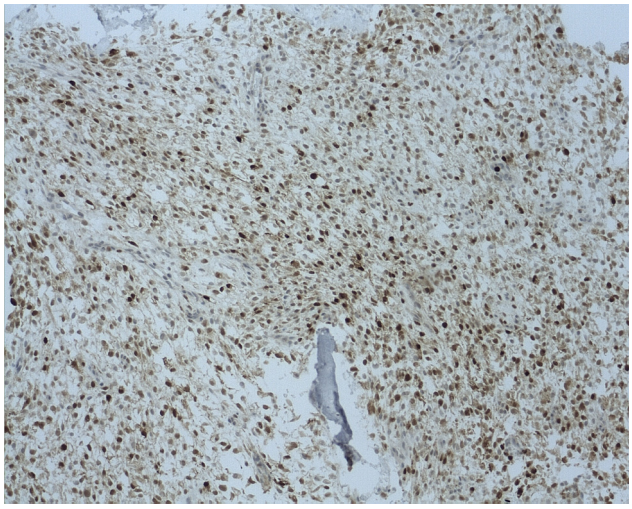

Fig. 3. Immunochemical stain positive for S100.

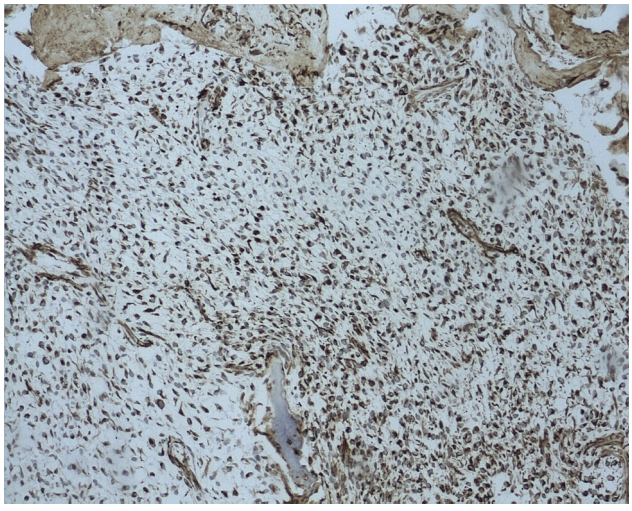

Fig. 4. Immunochemical stain positive for vimentin.