A Rare Case of Oropharyngeal Sarcoma

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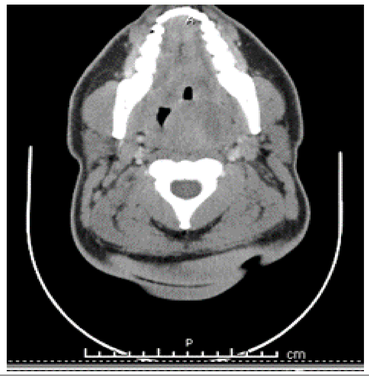
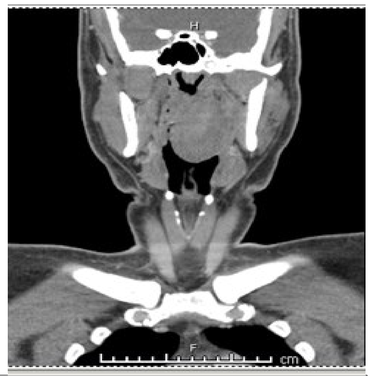
Introduction:

- Soft tissue sarcomas of the head and neck are rare, mesenchymal malignant neoplasms that account for 2-15% of all soft tissue sarcomas and for approximately 1% of all head and neck malignancies.[1,2,3,4]
- There are several histologically and morphologically different sarcoma subtypes The differential diagnosis includes Ewing sarcoma (ES), but also undifferentiated spindle cell sarcomas, such as synovial sarcomas (SS) and malignant peripheral nerve sheath tumors (MPNST).
- Improved genetic technologies have enabled identification of a BCOR-fusion sarcoma (BCOR-CCNB3). This is a type of undifferentiated unclassified sarcoma described as a 'Ewing-like' sarcoma.[5]
- Here we present the first documented case in the literature of a BCOR-rearranged head and neck sarcoma originating from the oropharynx.

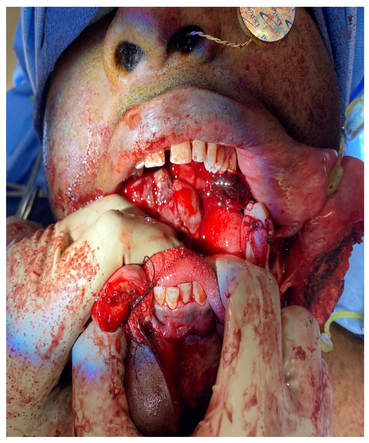
Case Presentation:

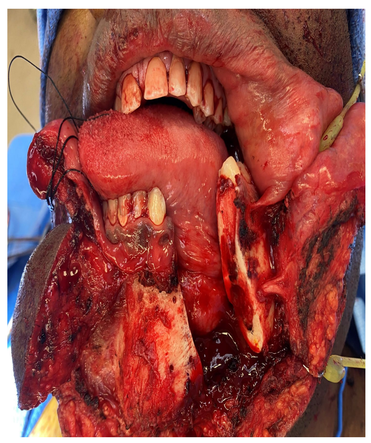
- History: 23 year-old man with no PMH presented to the emergency department (ED) at an academic medical center complaining of left-sided throat discomfort and dysphagia. He was diagnosed clinically with a left peritonsillar abscess. Needle aspiration yielded scant return and he was discharged with oral antibiotics. He re-presented to ED 2 weeks later with worse dysphagia, odynophagia and muffled voice.
- Our otolaryngology team was consulted upon his re-presentation. Physical examination demonstrated an enlarged, irregular-appearing left tonsillar mass without effacement of the anterior pillar. Nasopharyngolaryngoscopy (NPL): enlarged left tonsillar mass with oropharyngeal crowding but patent airway.
- CT Head & Neck: Tumor extended laterally involving the superior constrictor muscle, superiorly to the level of the nasopharynx, and inferiorly involving the posterior pharyngeal wall (**Figures 1-2).**
- Pre-op: Patient was admitted to the hospital for airway monitoring and underwent an excisional biopsy in the operating room (OR) the following day.
- Pathology: High-grade sarcoma composed of round to ovoid cells with a high mitotic rate originating from oropharynx, immunopositive for BCOR, (-) SS18-SSX fusion protein, (-) BCOR internal tandem duplication.
- Patient was uninterested in surgical resection at that time and could not be reached for follow-up until 2 weeks later when he re-presented to ED with increasing dyspnea and globus sensation. NPL: gross enlargement now measuring ~5cm from left tonsillar region. Patient underwent an open tracheostomy in OR for airway control. Subsequently, he underwent a lip-split mandibulotomy with left lateral oropharyngectomy and primary closure of the defect **(Figures 3-5)**. Gross margins were negative. He has begun aggressive systemic chemotherapy.[6]





Figures 1 and 2: Well-circumscribed heterogenous 4.5 x 3.7 x 5.3 cm exophytic soft tissue mass with subtotal effacement of nasopharynx and oropharynx.





Figures 3 and 4

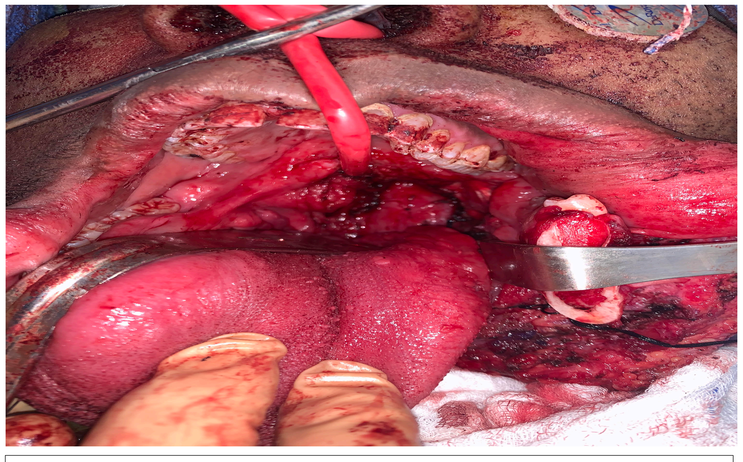


Figure 5

Discussion:

- The BCOR-fusion sarcoma is a rare and relatively recently genetically defined undifferentiated round cell sarcoma.[1-5]
- The usual management of localized Ewing Sarcoma includes locoregional control with surgery and/or radiotherapy, followed by chemotherapy.[6]
- As this tumor was classified as a Ewing-like sarcoma, our management specifically included local resection with negative margins (which we achieved) using a paramedian lip-split mandibulotomy approach to gain access to the oropharynx and posterior pharyngeal wall. The inferior alveolar nerve was preserved using this approach.
- Complete resection is associated with improved survival compared with incomplete resection.[7]
- Age and stage are the most important prognostic factors for clinical outcome; however, response to systemic chemotherapy is one of the most important prognostic factors affecting disease-free survival and overall survival. Mortality is most often due to metastatic disease (~50%). [8,9,10,11]
- No difference in overall survival is found when comparing patients treated with surgery, radiotherapy or combined surgery and radiotherapy.[11]
- Surveillance for recurrence should include regular history and physical exam including NPL. Role of PET/CT for detection of recurrence is controversial, but some studies have found PET/CT most helpful for surveilling high-grade sarcomas.[12,13]

Conclusions:

- Our case is unique in that it is the first documented case in the literature of a BCOR-rearranged head and neck sarcoma originating from the oropharynx.
- Management of this rare entity includes locoregional control with complete resection in addition to aggressive systemic chemotherapy.
- Mortality is most commonly due to metastatic disease.
- Treatment of rare and aggressive head and neck sarcomas requires complex care and a multidisciplinary approach.

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