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Case Report

Tonsillar Plasmacytoma Treated with Surgery

Shannon Alsobrooks[1], Raju Vaddepally[2], Abhinav B Chandra[3*]

[1]University of Arizona College of Medicine-Phoenix, Phoenix, Arizona, USA.

[2]Department of Oncology, Lee Health Regional Cancer Center, Fort Myers, Florida, USA.

[3]Yuma Regional Medical Center, Yuma Regional Medical Center, Yuma, Arizona, USA.

[*]**Corresponding author:** Abhinav B Chandra, Yuma Regional Medical Center, Yuma Regional Medical Center, Yuma, Arizona, USA.

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Abstract

We present a unique case of tonsillar plasmacytoma in a unique rural population at the Arizona, Mexico border. The patient is a 56 y/o male with significant tobacco use history presented with a complaint of 6 months of right tonsillar enlargement, difficulty swallowing, and food trapping. Biopsy histopathology identified plasma cell neoplasm with demonstrating weak to moderate positivity for CD138 and a subset demonstrates strong aberrant expression of CD56. At the time of diagnosis patient did not meet criteria for multiple myeloma, he had no evidence of hypercalcemia, monoclonal protein, M spike, or renal insufficiency. Absence of clinical evidence of plasma cell myeloma or end-organ damage due to plasma cell dyscrasia and a solitary tumor involving the tonsil, the morphologic and immunophenotypic findings were most consistent with extramedullary plasmacytoma (EMP). This patient underwent full surgical resection without radiation of tumor with PET CT demonstrating no evidence of residual disease.

Keywords: Tonsillar Plasmacytoma; Tonsillar Mass; Non-Hodgkin's Lymphoma; Solitary Plasmacytoma; Lymphoid Mass

Introduction

Extramedullary solitary plasmacytomas (EMPs) are extremely rare form of non-Hodgkin's lymphoma, accounting for around 3% of all plasma cell malignancies [1] EMPs can develop in any site of the body where lymphoid tissue may be found, but are most commonly found in soft tissues of the head and neck region. [2] These lesions often remain subclinical until the fifth decade of life or later at which point patients present with a localized submucosal mass or swelling along with nonspecific symptoms of upper respiratory obstruction. [3] In this paper we aim to highlight a recent patient presentation of solitary plasmacytoma which presented with asymmetric tonsillar enlargement.

Case Presentation

A 56-year-old non-Hispanic male patient sought medical attention as he noticed asymmetric right tonsil enlargement for about 6 months. He denied pain or weight loss. He complained of difficulty swallowing and food being trapped at the right tonsillar fossa. He did endorse a greater than 50-pack-year smoking history. No family history of malignancy. He denied any fevers or night sweats or lymphadenopathy or bone pain. History of successful treatment of previous hepatitis C infection. Current long-term smoker, previous drug use, previous alcohol use.

Evaluation by ENT revealed asymmetric tonsils right greater than left. Marked right tonsillar enlargement 3+. No inflammatory changes or stones palpated. Tonsil was soft with multiple crypts. No trismus no crossbite. The left tonsil was 1+ soft nontender. No uvular shift, no edema or swelling tonsil fossa. Patient underwent direct laryngoscopy and tonsillectomy.

Pathology was reported as-Tonsil, right, tonsillectomy-Plasma cell neoplasm. Microscopic examination of the right tonsil demonstrated an expansile lesion with diffuse sheets of atypical small to medium-sized cells with clock face chromatin pattern, prominent perinuclear halo and scant to moderate eosinophilic cytoplasm. Frequent multinucleation, irregular nuclear contours, Dutcher bodies and mitotic figures were also demonstrated. No

large cell or small cell infiltrates were identified. No definite necrosis is identified. Immunohistochemical stains pankeratin, CD20, CD3, CD138, CD56, cyclin D1 (single antibody procedures with appropriate controls) were performed on a selected tissue block. In situ hybridization studies for kappa, lambda light chains and EBV (EBER) were performed. The neoplastic cells demonstrated weak to moderate positivity for CD138 and a subset demonstrated strong aberrant expression of CD56. Neoplastic cells were negative for pankeratin and cyclin D1. CD20 demonstrated absence of B-cell infiltrates within the lesion. CD3 highlights scattered small reactive T lymphocytes in the background The plasma cells demonstrated kappa light chain restriction and were negative for lambda and EBV. In the absence of clinical evidence of plasma cell myeloma or end organ damage due to plasma cell dyscrasia and a solitary tumor involving the tonsil, the morphologic and immunophenotypic findings were most consistent with extramedullary or extraosseous plasmacytoma. [Figure 1 and 2]

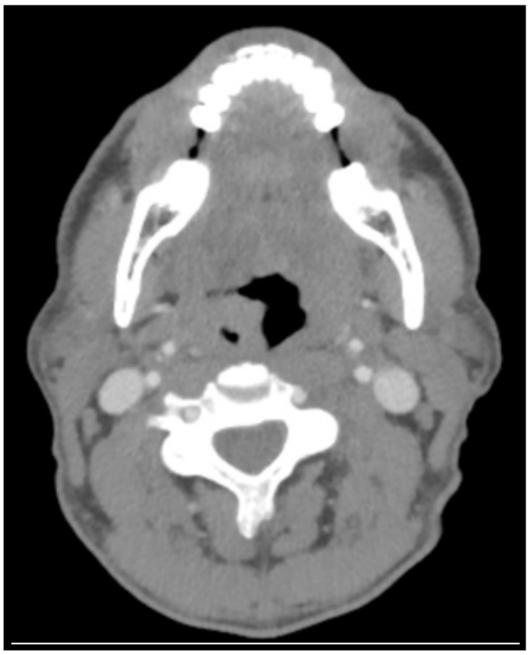

Figure 1: Axial section of contrast-enhanced axial PET/CT scan of oropharyngeal region demonstrating R tonsillar lesion.

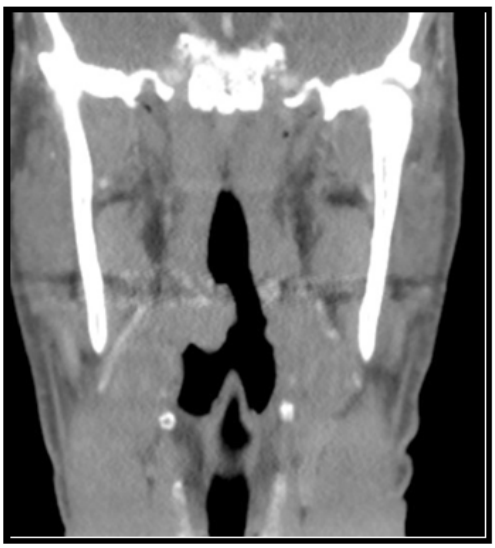

Figure 2: Coronal section of contrast-enhanced axial PET/CT scan of oropharyngeal region demonstrating R tonsillar lesion.

PET CT scan performed after surgery showed no residual mass or metabolic lesions in the oropharyngeal region. There were no metabolic lesions to suggest regional nodal or distant metastatic disease. Beta-2 microglobulin, quantitative immunoglobulins levels were normal. LDH level was normal. Protein electrophoresis did not reveal any monoclonal protein or M spike. Kappa free light chain was mildly elevated at 4.55 with kappa to lambda free light chain ratio also elevated at 4.42. There was no evidence of hypercalcemia or renal insufficiency. Mild anemia was noted. Bone marrow biopsy did not reveal any evidence of involvement with plasma cell neoplasm. Karyotype showed normal male chromosome-46, XY. Clinical findings were suggestive of solitary tonsillar plasmacytoma. No evidence of residual disease after surgery was found and there was no systemic involvement with multiple myeloma. No evidence of end organ damage was discovered. No treatment with chemotherapy and radiation therapy was required. Patient was managed with active surveillance and remained in complete remission one year post surgery.

Discussion

Although cases EMPs of the tonsil continue to be limited in the literature, we are noticing some trends in their incidence and prevalence in general. A large comparative study completed from 2003-2016 found that EMPs occur most frequently in Hispanic Men and non-Hispanic Black men, with higher prevalence of those who lived in non-metropolitan areas. [4]

EMPs vary widely in their symptomatology at presentation. Majority of patients experience pain at the site of the lesion and may also complain of neurologic manifestations as well [5-7] Documented signs and symptoms of tonsillar plasmacytoma noted in previous case reports included, lymphadenopathy, dysphagia, difficulty breathing, hoarseness and even constitutional symptoms such as weight loss. [8-10] There is evidence to support the use of PET CT or MRI to detect plasmacytomas. [7] However, definitive diagnosis of EMPs from other plasma cell dyscrasias requires biopsy. [9] Current literature has shown that there is much variety in immunohistochemical identification of plasmacytoma biopsies. Consensus has not been reached due to the rarity and location of these tumors often making it difficult to obtain adequate specimens. Of note, CD56, CD138 and Ki-67 have been shown to be highly expressed in extramedullary plasmacytomas [4,8,11] Other histological markers include, Vs38c, kappa light chain, lambda light chain, CD20, Cd79a and CD117. [12] Prior to diagnosis of an EMP, multiple other hematological malignancies must be ruled out including; non-Hodgkin's lymphoma, reactive plasmacytosis and plasmablastic lymphoma. [13] Patient's must also meet the following criteria to distinguish EMPs from multiple myeloma; 1) Existence of one or more extramedullary plasma cell tumors; (2) Inconspicuous bone marrow smear with normal plasma cell ratio within the bone marrow, including inconspicuous plasma cell morphology, or less than 10% plasma cell ratio; (3) No radiological evidence of osteolysis; (4) No hypercalcemia or renal failure; (5) None or low M-protein serum concentration [12].

Current literature has not defined a singular treatment modality for EMPs. This is likely due to the rarity of disease, variability in tumor location, nodal involvement, and patient co-morbidities. Plasmacytomas have historically been highly responsive to radiation therapy. Nodal radiation is often considered as well especially when the head and neck is involved. [7,12,14] One study demonstrated 98% overall survival rate after combination radiation and surgery therapy after 5 years. [12] Some studies have even shown that surgery alone in patients with fully resectable disease can show excellent results [14].

Conclusion

This case adds to our current literature on EMPs and provides an example of how these lesions may present somewhat insidiously in younger patients without previous cancer history or family history of multiple myeloma. This case is also unique in that the patient was treated with surgical resection alone.

Although our patient identified as non-Hispanic, the population that we serve is predominately Hispanic. It is important to acknowledge that although rare, this malignancy is very common in Hispanic and non-Hispanic Black men. [4] We also know that many members of the Hispanic community with cancer are unclassified by current our CDC and electronic health record reporting in the United States, so incidence is likely higher than reported. [4] This has large implications to our patient population and stresses the relevance of these demographic considerations [15].

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Ethical Guidelines: None

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