Extramedullary Plasmacytoma of the Nasopharynx Treated with Surgery and Adjuvant Radiation: Case Report and Review of the Literature

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ABSTRACT

Objectives
Extramedullary plasmacytoma (EMP) is a rare neoplasm of plasma cell origin that arises due to soft tissue infiltration of mononuclear plasma cells, in the absence of systemic disease. Although the head and neck is the most common location of EMP, there are only a handful of cases reported in the nasopharynx. This report conveys our recent experience with EMP of the nasopharynx in the context of the limited literature on this topic.

Study Design
Report of a case and review of the literature

Results
A 50 year-old healthy male presented with a year-long history of increasing bilateral nasal congestion unresponsive to medical therapy. Nasal endoscopy revealed a large exophytic mass filling both posterior nasal cavities. Imaging demonstrated a 3X3 cm mass arising from the nasopharynx extending into the posterior nasal cavities bilaterally. A biopsy was performed, demonstrating extramedullary plasmacytoma of the nasopharynx. Metastatic work-up was negative. The patient underwent surgical debulking followed by radiation therapy to the primary site and bilateral necks. He has not recurred to date and does not have evidence of progression to multiple myeloma.

Conclusions
This report adds to the scant literature on EMP of the nasopharynx, which may mimic many benign nasal and sinus conditions. Although surgery is usually considered second line treatment for EMP of the head and neck, this case demonstrates the utility of surgical debulking in symptom palliation before definitive treatment with radiation.

INTRODUCTION

Extramedullary plasmacytoma (EMP) is a rare neoplasm of plasma cell origin that arises due to soft tissue infiltration of mononuclear plasma cells in the soft tissue in the absence of systemic disease. EMP is characterized as a plasma cell tumor, along with solitary plasmacytoma of bone and multiple myeloma, which is considered a more advanced stage of disease. EMP accounts for 3-4% of all plasma cell tumors.

EMP is non-destructive to bone and soft tissue, but produces symptoms due to local tumor effects. They include nasal congestion, anosmia, hyposmia, epistaxis, rhinorrhea, pain, and neck swelling. There are only a handful of reports specifically describing EMP arising in the nasopharynx. This report conveys our recent experience with EMP of the nasopharynx in the context of the limited literature on this topic, with an emphasis on multimodality treatment.

DISCUSSION

The diagnosis of extramedullary plasmacytoma (EMP) is made when a localized collection of monoclonal plasma cells exists in the absence of plasma cell proliferation elsewhere in the body (i.e., bone marrow, blood). Diagnostic criteria, as recommended by the UK Myeloma Forum include:

1. Single extramedullary mass of clonal plasma cells, histologically normal bone marrow aspirate, normal skeletal survey, no evidence of anemia, hypercalcemia, or renal impairment due to myeloma, and absent or low serum and urinary mononuclear immunoglobulin.
2. In less than 25% of patients, a monoclonal paraprotein can be detected in the blood or urine.
3. Systemic evaluation should include CBC, ESR, Serum and urine protein electrophoresis, immunoglobulin determination, bone marrow biopsy, and skeletal survey.

Others recommend spinal MRI for staging to evaluate for bony lesions. The goal of testing is to exclude systemic involvement characteristic of multiple myeloma. Some authors also recommend pan-endoscopy for suspected cases of EMP, including nasal endoscopy, laryngoscopy, bronchoscopy, and esophagoscopy/gastroscopy.

CT or MRI is critical to delineate the extent of local disease; however, there are no characteristic imaging criteria. Due to the submucosal nature of the tumor, deep biopsies or excisional biopsies are sometimes required for diagnosis; fine needle aspiration may be inadequate. Histologically, EMP may mimic other conditions, such as lymphoma, undifferentiated carcinoma, reactive plasmacytoma, and plasma cell granuloma; these are best distinguished by immunophenotyping that are CD138+ and positive for monoclonal cytoplasmic light chain expression of malignant plasma cells.

SURGERY

Traditionally, surgery alone has had a limited role in the treatment of EMP, and its use remains controversial. Although surgery can be curative, there is significant morbidity associated with adequate resection. Further, it is not always possible to obtain adequate margins in the head and neck. Given these limitations, as well as the responsiveness of EMP to radiation, surgery is currently a second line treatment. In a review of 714 cases of EMP of the head and neck, Alexiou et al. found that 44.3% of cases were treated with radiation alone, while 26.9% were treated with surgery & radiation, and 21.9% with surgery alone. They found a statistically significant increase in overall or recurrence free survival in patients treated with surgery and radiation. When resectable, they recommend surgical resection; adjuvant radiation is utilized in cases of close margins or incomplete resection. This position is supported by other authors.

In terms of the nasopharynx there are no reports directly addressing surgical resection, although some authors have utilized endoscopic or open methods to resect such lesions. Our case is unique in that in our patient, the necks were included in the radiation fields due to concern for neck involvement.

PROGNOSIS

Between 1905 and 1997, 131 cases of EMP involving the nasopharynx were reported. Approximately 20% of EMP cases in the nasopharynx are associated with or arise from the nasopharynx. They tend to present at an earlier age than solitary plasmacytoma of bone or multiple myeloma, with a 3:1 male dominance. EMP accounts for 3-4% of all plasma cell tumors. Between 1905 and 1997, 131 cases of EMP involving the nasopharynx were reported. The mean age at presentation was 53 years with a range from 18 to 91 years. EMP occurs more commonly in the 6th and 7th decade of life, with a 3:1 male dominance. EMP accounts for 3-4% of all plasma cell tumors. The most common location is the head and neck, with estimates ranging up to 80-90%.

EMP is a non-destructive to bone and soft tissue, but produces symptoms due to local tumor effects. They include nasal congestion, anosmia, hyposmia, epistaxis, rhinorrhea, pain, and neck swelling. There are only a handful of reports specifically describing EMP arising in the nasopharynx. This report conveys our recent experience with EMP of the nasopharynx in the context of the limited literature on this topic, with an emphasis on multimodality treatment.

EMP of the Nasopharynx (NP)

Between 1905 and 1997, 131 cases of EMP involving the nasopharynx were reported, accounting for approximately 18% of cases of the head and neck. More recently, Wein et al. reviewed 21 cases of EMP of the NP, reporting a myeloma progression rate of 9.5%. Unfortunately, these reports do not specify the exact treatment for the nasopharynx.

RADIATION

Given its radiosensitivity, radiation is the first line treatment for EMP, with local cure rates of 80-100% of the norm. However, given the heterogeneous nature of the literature, there is no evidence based dosing regimen, with most larger series reporting the use of 35-60 Gy.

CONCLUSIONS

This report adds to the scant literature on EMP of the nasopharynx, which may mimic many benign nasal and sinus conditions, and must be included on the differential diagnosis of refractory nasal obstruction and congestion. Although surgery is usually considered second line treatment for EMP of the head and neck, this case demonstrates the utility of surgical debulking in symptom palliation before definitive treatment with radiation.

REFERENCES