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CASE REPORT

Laryngeal Extra Medullary Plasmacytoma

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Abstract

Extramedullary plasmacytoma (EMP) is a rare form of plasma cell neoplasm that occurs outside of the bone marrow. Although it represents less than 1% of all head and neck tumors, 80% of EMPs occur within the head and neck region, with most of them found submucosally in the paranasal sinuses and nasal cavity. Laryngeal EMP is especially rare, accounting for only 0.2% of all laryngeal neoplasms. We present a case of a patient presenting with dysphagia, dysphonia, and cough found to have laryngeal EMP. The patient subsequently received radiotherapy alone with favorable outcome at one year follow up.

1 | CASE REPORT

A 57-year-old male with past medical history of diabetes, hypertension, and heart transplantation presented with a chief complaint of dysphonia for 1 year. He noted that his voice was worse in the morning but did not fatigue with use throughout the day. His symptoms were gradual in onset. He denied dysphagia, odynophagia, cough, globus sensation, or weight loss. He had been treated with pantoprazole 40 mg without improvement. He denied current or previous tobacco or alcohol use. Flexible laryngoscopy with stroboscopy revealed a papillomatous lesion with leukoplakia involving both vocal folds and involving the anterior commissure (Figure 1) The remainder of his head and neck exam was normal.

Figure 1. In office Laryngoscopy Exam

Direct operative laryngoscopy with biopsy was performed. Hematoxylin and eosin staining demonstrated mucosa with a plasma cell rich infiltrate which upon immunohistochemical staining was positive for CD138. Kappa monotypic plasma cells represented greater than 85% of the total cell population. CD3 and CD20 staining highli -ghted scattered T-cells and B cells, respectively which was consistent with a diagnosis of an extramedullary plasmacytoma.

The patient was evaluated by the Hematology/ Oncology and Radiation Oncology services. PET/ CT showed no evidence of FDG uptake, and the patient was subsequently treated with 5000 cGy of radiation in 25 fractions. He has had an unremarkable post-treatment course with no signs of recurrence 1-year post treatment.

2 | DISCUSSION

Extramedullary plasmacytoma (EMP) is a rare form of plasma cell neoplasm which occurs outside of the bone marrow. Although it represents less than 1% of all head and neck tumors, 80% of EMPs occur within the head and neck region, with the majority of them found submucosally in the paranasal sinuses and nasal cavity.(1), (2) EMP of the larynx is extremely rare and accounts for less than 0.2% of malignant laryngeal tumors.(2) The most common presenting symptoms of laryngeal EMP is dysphonia, dysphagia, and cough. In a recent literature review of laryngeal EMPs, the supraglottis was found to be the most common site.(3) Patients presenting with cervical lymphadenopathy or involvement of multiple sites were more likely to recur or have distant metastasis.

Pathological diagnosis of EMPs is made by identifying monoclonal plasma cells on histology and CD138 on immunohistochemical staining. Computed tomography usually demonstrates a homogeneous mass with well-defined margins. Additional workup including complete blood count, lactic dehydrogenase, urine protein electrophoresis, serum protein electrophoresis, serum immunofixation electrophoresis, serum free light chain assay, and serum quantitative immunoglobulins to rule out concomitant multiple myeloma.(4)

EMPs are highly radiosensitive and treated with radiotherapy alone while multiple myeloma requires the addition of systemic chemotherapy. Regular follow-up is necessary since approximately 16 to 23% of EMPs of the head and neck have been noted to progress to multiple myeloma, usually within the first two years.(2),(5)

Supplementary information The online version of this article (https://doi.org/10.52845/JORR/2022/3.4.1) contains supplementary material, which is available to autho-rized users.

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3 | CONCLUSION

Laryngeal EMP accounts for less than 0.2% of malignant laryngeal tumors. They generally present with non-specific symptoms including dysphonia, dysphagia, and cough. When EMP occurs in the larynx, they most often involve the supraglottis. Radiotherapy alone is an effective

treatment for EMP. However, close follow-up is necessary to monitor for progression to multiple myeloma.

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