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CASE REPORT

Multiple Myeloma Presenting as Respiratory Stridor

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ABSTRACT

Extramedullary plasmacytoma occurs in 18% of patients with multiple myeloma. Laryngeal involvement in multiple myeloma is rare, and only a few cases have been reported. We present a case of a 44-year-old women with multiple myeloma who presented with stridor due to a mass involving the larynx which was initially proven to be plasmacytoma on biopsy. She had evidence of multiple myeloma of IgA lambda subtype. She was treated with bortezomib containing chemotherapy followed by lenalidomide as maintenance therapy. She attained complete remission and is alive in remission at 3 years of treatment.

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Introduction

Plasmacytoma is a discrete solitary mass of monoclonal neoplastic plasma cells occurring either in bone (solitary plasmacytoma of bone) or in soft tissues (extramedullary plasmacytoma/EMP). EMP occurs in 18% of patients with multiple myeloma.[1] The most common sites of EMP are head and neck region, upper respiratory tract, gastrointestinal tract, and central nervous systems. EMP accounts for less than 1% of all malignant head and neck tumours. Multiple myeloma (MM) usually presents with anemia, bone pain, and renal failure. Laryngeal involvement in multiple myeloma is rare, and only a few cases have been reported.[2-7] We present a woman with multiple myeloma who presented with stridor due to plasmacytoma involving the larynx.

Case Report

A 44-year-old woman presented with hoarseness of voice for 3 months and a period of two weeks of stridor. There was no dysphagia. Computed tomogram scan of neck showed a soft tissue mass (3 x 3.3 x 3.4 cm) in the left pyriform sinus and aryepiglottic fold encasing hyoid bone, thyroid cartilage, and vocal cord on left side along with bilateral regional lymphadenopathy (figure 1). She underwent emergency tracheostomy. Indirect laryngoscopy revealed a mucosa-lined growth involving the left arytenoids, aryepiglottic fold and pyriform sinus with fixed left hemilarynx (figure 2). A biopsy from the mass showed plasmacytoid cells loosely arranged in a fibrocollagenous stroma. On immunohistochemistry, the tumor cells were strongly positive for CD138 and negative for cytokeratin, thus a preliminary diagnosis of plasmacytoma was made for the patient (figure 3A & 3B). Skeletal survey revealed multiple lytic bone lesions and bone marrow aspiration showed a population of about 45% immature plasma cells which resulted in final diagnosis

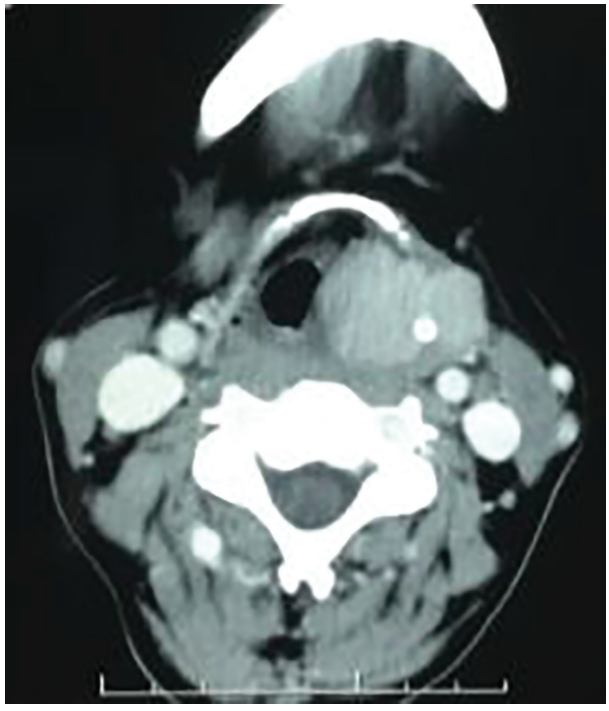

Figure 1: CT-scan of the neck showing a soft tissue mass in left pyriform sinus and aryepiglottic fold encasing hyoid bone, thyroid cartilage and vocal cord on the left.

of MM, since bone and bone marrow involvement were detected. The 24-hr urine protein was 2.49 gm/day, and β2-microglobulin was 14.5 mg/dl. Serum electrophoresis showed M-band with M protein level of 7.8 gm/dl. Serum Immunoglobulin (Ig) A was 11955 mg/dl, IgG was 333 mg/dl, IgM was <25mg/dl, free kappa chain was 15.5 mg/L. and free lambda chain was 114.3 mg/L. Immunofixation electrophoresis demonstrated the presence of IgA lambda monoclonal gammopathy. She received bortezomib and dexamethasone for 6 cycles. A repeated CT-scan after 6 cycles of chemotherapy showed complete resolution of the laryngeal lesion. She also achieved complete remission of MM at this point. She was planned for consolidation with high-dose melphalan and autologous peripheral stem cell transplantation, however she refused. She was scheduled to receive maintenance with lenalidomide for

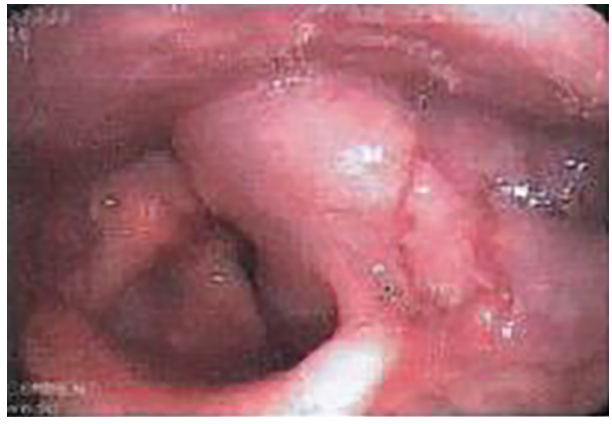

Figure 2: Indirect laryngoscopy showing a mucosa covered growth involving the left arytenoids, aryepiglottic fold and pyriform sinus.

1 year. Currently she is alive in complete remission at 3 years. Patient has given written informed consent for publishing her case details.

Discussion

Neoplasms originating from plasma cells are rare in the head and neck region. Approximately 80% of these EMPs involve the paranasal sinuses, pharynx, nasal cavity or gum and oral mucosa and only 10% of EMP occur in larynx.[8] EMP of larynx represent only 0.04-0.45% of malignant laryngeal tumors. Laryngeal involvement is rare in plasma cell neoplasms and the sites involved are the epiglottis, vestibular fold, arytenoids, aryepiglottic folds, and the subglottis.[2] The peak age of incidence is the 6th decade of life with a male preponderance of 3:1.[8] The clinical presentation varies according to the location of the mass and includes hoarseness, cough, dyspnoea, and stridor.

There are only 2 cases of laryngeal EMP among 22 cases of EMP of the head and neck observed over 20 years.[3] A 65-year-old man with IgG myeloma presenting with stridor has been described.[4] Meanwhile, a 58-year-old man with IgA smoldering myeloma presenting with dysphonia; which was found to be due to laryngeal involvement of MM, has also been reported.[5] A 62-year-

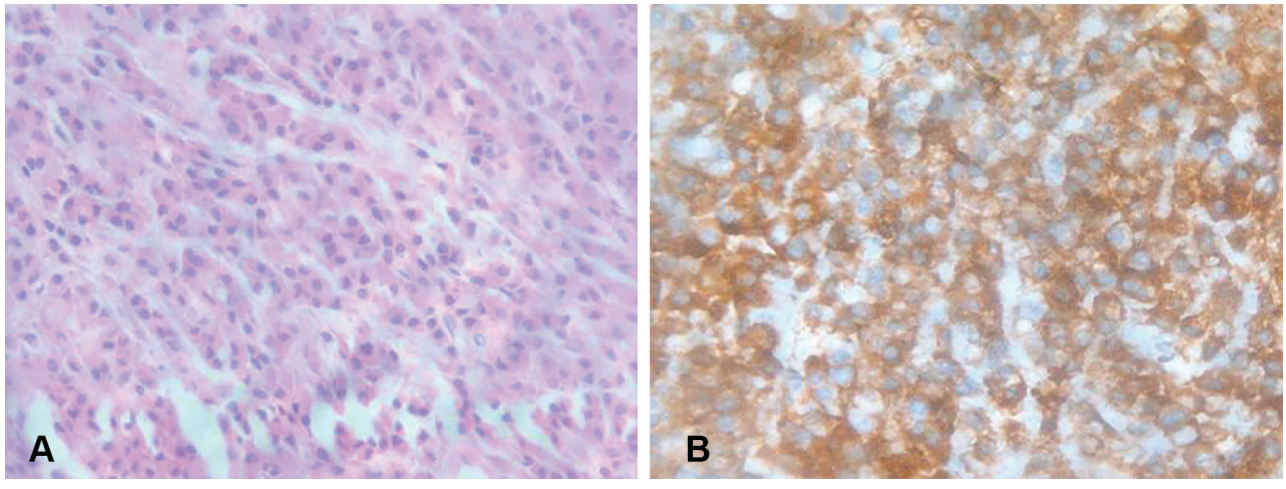

Figure 3: **A)** H&E x40: Section from the laryngeal mass showing plasmacytoid cells. **B)** Immunohostochemistry showing tumor cells positive for CD138.

old woman with dysphonia was also detected to have a plasmacytoma arising from her left true vocal fold through fiberoptic laryngoscopy.[6] There is also a case of plasmacytoma of the larynx being treated with radiotherapy.[7] Our patient presented with stridor whose initial diagnosis of plasmacytoma was positive for CD138; however, she was further diagnosed with MM due to bone marrow involvement with about 45% plasma cells.

Treatment options for isolated EMP of larynx include radiotherapy, laser surgery, endoscopic or conservative surgery and chemotherapy. Plasmacytoma is highly sensitive to radiation and enables voice preservation. The prognosis is related to the location of the tumor and cartilage or bone destruction. Survival is higher in patients with localized disease than in those with MM.[4] Our patient was treated with bortezomib containing chemotherapy in view of the systemic disease. She achieved complete remission and maintenance therapy was continued after achieving remission in place of autologous stem cell transplantation.

Conclusion

Multiple myeloma rarely presents with symptoms related to the primary site of an EMP, as in this case who presented with stridor due to EMP involving the larynx. This entity should be kept in mind in patients with hoarseness and dysphonia.

Conflict of Interest: None declared.

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