

SOLITARY EXTRAMEDULLARY PLASMACYTOMA - LARYNX

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ABSTRACT : *Solitary Plasmacytoma of the larynx is a rare tumor with male predominance and most patients are between 50-70 years of age. The paper reports a case of Solitary Extramedullary Plasmacytoma (EMP) Larynx in subglottic region, in a 65-year-old man successfully treated with complete response to radiotherapy.*

Key Words : *Extramedullary Plasmacytoma, Laryngeal Tumors.*

INTRODUCTION

Plasmacytoma is a rare neoplastic disorder from B cell series lymphocytes. It can develop in three clinical variants (a) Multiple Myeloma; (b) Solitary Plasmacytoma of the Bone; (c) Extramedullary Plasmacytoma (EMP) (Tesei, 1995). EMP are rare malignant neoplasms, 90% of which occurs in the Head and Neck area but generally representing only 1% of tumors of that location (Navarro Cunchillos, 1999). The authors present a case of solitary EMP of the larynx, a very rare laryngeal tumor.

CASE REPORT

A 65-year-old man presented with a 6 months history of dry cough, hoarseness of voice with discrete dysphagia and shortness of breath (on exertion and at night). He was a known tobacco chewer since 40 years. No past history of medical illness or surgery.

On general examination, mild stridor was present. On indirect and direct suspension laryngoscopic examination, a polypoidal growth was found in the subglottic region. Biopsy was taken. Both vocal cords appeared normal. After the procedure patient was developed respiratory distress, hence tracheostomy was performed. Histopathological examination of the biopsy specimen revealed pseudostratified columnar epithelium (Respiratory epithelium) with underlying stroma. It showed proliferative sheets of mature and immature plasma cells. Many large mononucleated as well as binucleated cells resembling RS cells were also seen. Nuclear and cytoplasmic inclusion were present. The whole picture was suggestive of ? Plasmacytoma,?? Lymphoma,??? Mucosa associated plasmacytoma. The diagnosis EMP was confirmed by

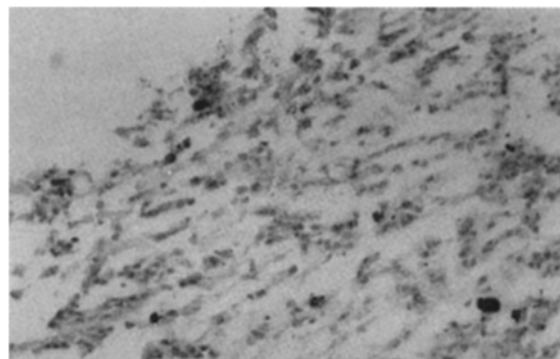


Fig. 1: Microphotograph showed immunohistochemical cytoplasmic staining for kappa antigen.

Immunohistochemical (IHC) examination, which showed LCA - negative, kappa chain - positive (in 70% - 80% of cells), λ chain - focal positive, CD 30 - negative (Fig. 1).

All laboratory and radiological examinations such as urinary BJ proteins, bone marrow aspirate and biopsy, skeletal survey, ultrasonographic (USG) examination of the abdomen and pelvis were normal. This ruled out systemic evidence of plasmacytoma and multiple myeloma. CT Scan of the Neck showed infiltrating as well as nodular thickening on subglottic region from midline to left side.

The patient received local external radiotherapy (60 Gy in 30 fraction for 6 weeks) and was doing well for two years following therapy with evidence of disease.

DISCUSSION

Extramedullary plasmacytoma is an uncommon tumor, comprising 4% of all plasma cell neoplasms (Nowak-Sadzikowska, 1998). Solitary EMP of the larynx

represents a rare category of malignant disease. Review of literature gave limited information regarding diagnosis, staging and natural history (Liebross, 1999). It is predominantly found in males between 50 -70 years. Most patients present with hoarseness and chronic cough. The epiglottis is the site most commonly involved, followed by the ventricles, true vocal cords and false vocal cords (Schwartz, 1999). But in our case the polypoidal growth was in subglottic region.

Biopsy and histological examination establish diagnosis but confirmed by IHC. Large number of the plasma cells in the tumor are positive for kappa light chains on IHC examination (Rakover, 2000). Complete work-up to rule out systemic evidence of plasmacytoma is necessary. Microscopically, solitary plasmacytoma must also be distinguished from plasma cell granuloma.

Radiotherapy is the treatment of choice. Although surgery is generally considered as a diagnostic tool, local disease always be removed surgically when surgery produces low morbidity. Surgical debulking of the lesion can increase the probability of local radiotherapeutic control (Tesei, 1995).

Long-term follow up is necessary as 20% of patient eventually may develop distant disease after years.

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