EXTRA MEDULLARY PLASMACYTOMA OF LARYNX-
A RARE CASE REPORT

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ABSTRACT

Introduction: Extra medullary plasmacytoma (EMP) is a rare neoplasm of plasma cell, described as soft tissue outside the bone marrow. EMP represents 0.04 to 0.45% of malignant tumours of larynx.

Objective: To describe and report a case of extramedullary plasmacytoma of larynx with literature review.

Case report: A 60yr old male presenting with dyspnoea for 4 months worsening progressively with globular mass over right lateral glosso-epiglottic fold and aryepiglottic fold, being subjected to excisional biopsy diagnosed as extramedullary plasmacytoma.

Conclusion: Examedullary plasmacytoma must be considered in the differential diagnosis of tumours of larynx. It is highly curable when radiotherapy is used.

Key words: Extramedullary plasmacytoma, larynx, radiotherapy.

INTRODUCTION

Extramedullary plasmacytoma (EMP) is a rare neoplasm of plasma cell, described in soft tissue outside bone marrow\(^1\). The median age of presentation is 56-59yr\(^2\). It occurs predominantly in males, with male to female ratio of 3:1.\(^3\) The most commonly affected sites are the submucosal lymphoid tissue of nose and paranasal sinuses\(^1\). It has been reported rarely in larynx about i.e 10\(^%\)\(^4\).

The symptoms are mainly dysphonia, dysphagia, cough and dyspnoea when EMP affects the larynx/laryngopharynx. The EMP of larynx are usually submucosal.

The diagnosis of an EMP is primarily histological based on the presence of plasma cells which in the immunohistochemical study shows monoclonality, pointing to its neoplastic nature\(^5,6\). Moreover the diagnosis is based on exclusion of multiple myeloma.

EMP is a localised entity usually associated with a long surveillance\(^2\). Nevertheless in 16\(^%\) cases the disease can progress to multiple myeloma\(^4\). It is highly radio sensitive, so radiotherapy is the treatment of choice.

CASE REPORT

A 60yr old male with a history of smoking (15-16 packs per year) and known case of diabetes mellitus presented to ENT dept of Shreeram Chandra Bhanj Medical College and Hospital, Cuttack because of dyspnoea for 2months which was progressive and not associated with hoarseness of voice, dysphagia or haemoptysis.

He has past history of excision of a globular mass of larynx & posterior pharyngeal wall left side by internal approach under GA in other hospital in 2005 histopathology of which had suggested plasmacytoma/multiple myeloma which was not confirmed by immunohistochemistry\(^\text{(Fig.1)}\).

Clinically there was no external swelling, any scar mark or any pulsation over neck. There was no palpable cervical lymphadenopathy.

The indirect laryngoscopy revealed a globular mass over right lateral glosso-epiglottic fold and right aryepiglottic fold. pyriform fossa of right side was not visible & rest part of larynx was normal. CT/MRI scan was not done as the mass was confined to limited area of larynx and the patient was not able to afford it\(^\text{(Fig.2)}\).

The patient was hospitalised, all the investigations were done and he was prepared for excision of the mass under General Anaesthesia.

First of all under Local anaesthesia tracheostomy was done. GA was then given through PVC cuffed
tube. Horizontal skin incision was given over neck at the level of mid part of thyroid cartilage. An apron flap was elevated up to hyoid bone above and cricoid cartilage below. Sternohyoid and thyrohyoid muscles were cut and larynx was skeletonised. Laryngotomy was done through thyrohyoid membrane on right side and from aryepiglottic & glossoepiglottic fold by dissecting through the mucosa mass was excised and delivered. No lymph node were found involved peroperatively. Haemostasis was maintained, Romovac drain was placed and wound was closed in three layers. Nasogastric tube was inserted. After excision of mass, patient was relieved symptomatically. He was under broad spectrum antibiotic for 10 days. The stitches were removed on 10th day. There was wound dehiscence which was repaired by secondary suturing which was found healed on 20th day. The patient was discharged after decannulation on 25th day.

Post operative histopathology study was consistent with the diagnosis of plasmacytoma. Multiple myeloma was excluded as negative results were obtained both in the blood protein electrophoresis and urine immunofixation studies.

The patient then underwent Radiotherapy after confirmation of diagnosis but he did not come for follow up study and went away to his village.

**DISCUSSION**

A plasmacytoma is a discrete solitary mass of neoplastic monoclonal plasma cell in either bone or soft tissue(extramedullary).

The types of plasmacytoma are as follows:

- Solitary bone plasmacytoma
- Soft tissue/extramedullary plasmacytoma
- Multifocal form of multiple myeloma
- Multiple myeloma
- Plasmablastic sarcoma
Diagnostic criteria for EMP are:9

- Tissue biopsy showing monoclonal plasma cell on histology.
- BM plasma cell infiltration not exceeding 5% of all nucleated cell.
- No evidence of myeloma or osteolytic lesion.
- Absence of hypercalcemia or renal failure.
- Low serum M protein, even if present.

EMP usually presents as a mass growing in aerodigestive tract in 80-90% cases often with spread to lymph node. Most of the presenting symptoms are due to compression or invasion to surrounding structures. Common complaints include swelling, headache, nasal discharge, epistaxis, nasal obstruction, sore throat, hoarseness, dysphagia, dysphonia, dyspnea, hemoptysis and epigastric pain.

The etiology may be related to chronic stimulation of inhaled irritants or viral infection.

The most common sites of laryngeal plasmacytoma are in the decreasing order of frequency are: epiglottis, vocal cord, ventricular band, arytenoid and subglottis.4

Wittshaw classified soft tissue plasmacytoma into three clinical stages:7

Stage I- Limited to an extramedullary site
Stage II- Involvement of regional lymph node
Stage III- Multiple metastasis

In our case the patient was in Stage I.

The accepted treatment for EMP is radiotherapy.1 The optimal dose for local control is 40-50 Gy (depending on tumour size) delivered over 4-6 weeks. Because of high rates of lymph node involvement these areas should be included in radiation field. Adjuvant radiotherapy should be given to patient with positive surgical margin. Chemotherapy should be considered for refractory or relapsed cases. Adjuvant chemotherapy for tumour > 5cm as well as those with high grade histology is needed.

CONCLUSION

EMP should be considered in the differential diagnosis of rare tumours of larynx. It is highly curable when radiotherapy is used. Moderate radiation dose & limited field ensure excellent cosmetic and functional result.

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REFERENCES