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CASE REPORT

Radioresistant Solitary Extramedullary Plasmacytoma of Larynx

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ABSTRACT

Solitary extramedullary plasmacytomas (SEP) of larynx are rare tumors and are radiosensitive. However, a case of radioresistant SEP of larynx was managed by endoscopic CO_2 excision.

Keywords: Plasmacytoma, Radiotherapy, CO_2 Laser.

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INTRODUCTION

Solitary extramedullary plasmacytomas (SEP) are focal proliferation of plasma cells in soft tissues.[1] 85 to 90% of the SEP's arise in the head and neck region of which the nose and paranasal sinuses being the most common sites.[2] Other sites in the head and neck are larynx, oropharynx, salivary glands, thyroid, orbit and skin.[3] Most of the SEP are radiosensitive.[4] A unique case of radioresistant SEP is being reported.

CASE REPORT

A 40-year-old farmer, with a history of tobacco chewing for the past 20 years and no other comorbidities, presented in the outpatient department with chief complaint of hoarseness of voice for 6 years and dyspnea on exertion for 3 years. There was no history of dysphagia, stridor, aspiration or weight loss. There were no other ENT complaints. The patient had undergone microlaryngoscopy and biopsy thrice which had been reported as round cell tumor, plasmacytoma and plasmacytoma with kappa chain restriction respectively.

On fiber optic laryngoscopy a smooth pink mass involving the left AE fold, arytenoids and medial wall of pyriform fossa was seen (Fig. 1). The epiglottis was pushed to right side and the left vocal cord was not seen. Right AE fold, arytenoid, vocal cord and pyriform fossa were normal. No palpable neck nodes were present.

Review of blocks and slides showed plasmacytoma with kappa chain restriction. Hemogram, liver function test, kidney function test, urine for Bence Jones protein was normal. X-ray chest, X-ray skull, X-ray spine, X-ray pelvis and USG abdomen were all normal. CT scan neck showed an ill-defined enhancing mass in the left supraglottic region measuring 4.20 × 3.96 cm, abutting epiglottis and displacing it to the right side (Fig. 4). There was effacement of left pyriform fossa by the mass. The left paraglottic fat was infiltrated. There was no cartilage involvement or cervical lymphadenopathy. Serum electrophoresis showed no M-protein. Bone marrow aspiration was normal with no evidence of multiple myeloma. Bone scan (Tecnitium 99) did not show any bony metastasis.

These features were suggestive of a SEP. The patient underwent radiotherapy (56Gy/28Fr/37dy), however no regression was seen (Fig. 2). Endoscopic surgery was therefore offered to the patient for the removal of the radioresistant mass. Tracheostomy and endoscopic CO_2 assisted (10W continuous mode) excision of the tumor was done. Postoperative period was uneventful and decannulation of the tracheostomy was done by 7th postoperative day.

HPE was reported as a plasmacytoma with kappa chain restriction. The patient is under follow-up with no recurrence (Fig. 3) and has a normal voice. There are no signs of multiple myeloma.

DISCUSSION

Solitary extramedullary plasmacytoma of larynx account for 10% of extramedullary plasmacytoma of head and neck.[4] They represent <0.2% of laryngeal neoplasms. Peak age of incidence is the 6th decade with a male predisposition of 3:1.[4] Most common subsite is the supraglottis, of which epiglottis is the commonest. Among 10 to 20% incidence of multiple myeloma is seen at time of diagnosis. About 15% have cervical metastasis at diagnosis. About 20% will eventually

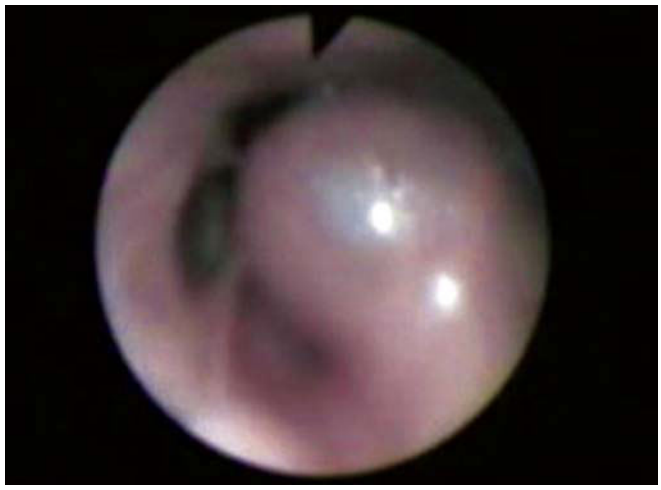

Fig. 1: Fiber optic laryngoscopy at initial presentation

develop distant metastasis (multiple myeloma) within 2 years of therapy, regardless of control of primary disease.[5] Two-third of the patients survive for more than 10 years.

Investigations recommended are biopsy (deep biopsy has to be taken as tumor is submucosal) and immuno-histochemical staining (local amyloid deposits are seen in 11-38%). HPE examination can not distinguish multiple myeloma from an extramedullary plasmacytoma, therefore the following investigations are recommended to rule out systemic disease—complete blood count, bone marrow biopsy, liver function tests, kidney function tests, serum and urine electrophoresis, skeletal survey and imaging (CT scan or MRI).[6]

Solitary extramedullary plasmacytomas have been staged as:[6]

- *Stage I:* Localized and controllable disease
- *Stage II:* Local extension and involvement of the lymph nodes
- *Stage III:* Disseminated disease.

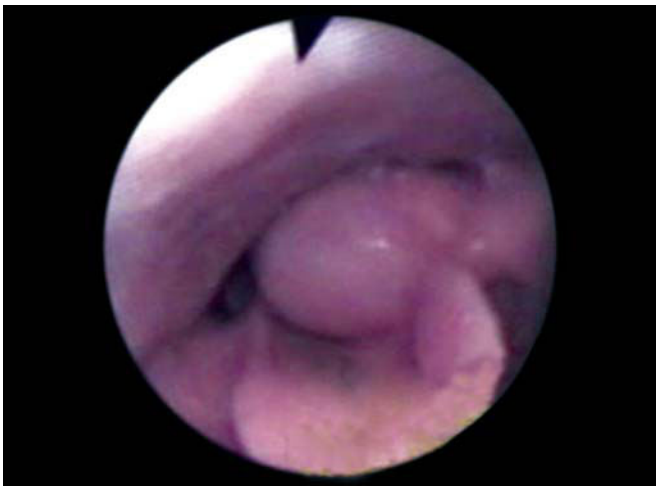

Fig. 2: Fiber optic laryngoscopy postradiotherapy

SEP are highly radiosensitive tumors. Local control rates of 80 to 100% are consistently reported with moderate doses of radiotherapy.[4,8] Radiotherapy dose and anatomical volume should be chosen to minimize early and late side effects and maximize control rates. There is no firmly established dose response relationship, due to small patient series and low local failure rates.[7-9]

The optimal radiation dose therefore appears to be in the range of 40 to 50 gray. SEP <5 cm have an excellent chance of local control with radiation doses in the region of 40 gray in 20 fractions, whereas there is a higher risk of local failure in tumors greater than 5 cm, which require a higher dose in the region of 50 gray in 25 fractions.

The optimal target volume is controversial.[7-9] However, in view of the morbidity of cervical lymph node irradiation and the high local control rates reported without it, the balance of evidence favors a policy of treating the primary site with a margin and including cervical nodes only if clinically involved, or regarded at high risk, as in primary sites involving Waldeyer's ring.

There are differing views about the role of surgery in SEP.[2,6] Surgery is generally only required for diagnosis. Both surgery and radiotherapy give equally good local control rates.[1] Radical surgery with curative intent is generally mutilating and is not indicated as the tumors are generally radiosensitive and the majority of patients are cured by radiotherapy. Radiotherapy therefore remains the mainstay of therapy for most SEP.[4,8,10]

Combination of surgery and radiotherapy has not shown any additional benefit when compared to the individual modalities in isolation[11] and adjuvant radiotherapy was useful only when surgical margins were involved.

Adjuvant chemotherapy is considered in patients with tumors >5 cm and those with high grade tumors and also in refractory and/or relapsed disease.[9]

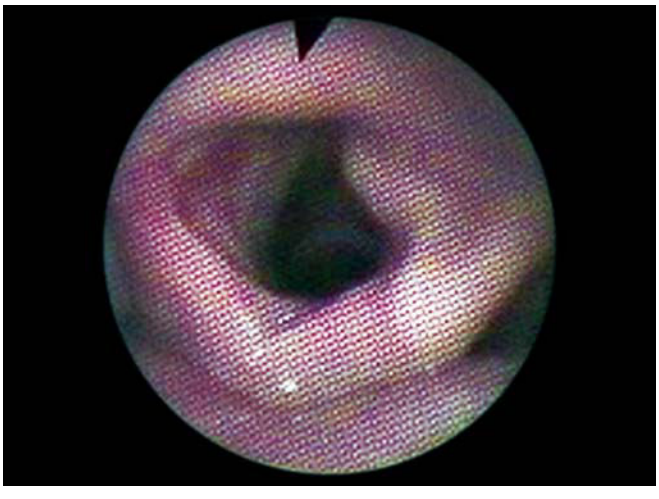

Fig. 3: Postoperative 1 year

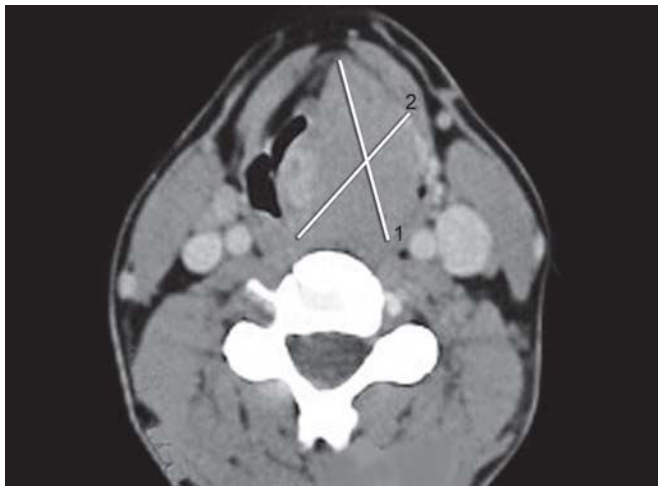

Fig. 4: Preoperative axial CT scan showing an ill defined enhancing mass in the left supraglottis region measuring 4.20 × 3.96 cm

Other treatment approaches that have been tried include thalidomide[12] and bisphoshonates.

CONCLUSION

Solitary extramedullary plasmacytomas of larynx are extremely rare tumors. Further more, they are rarely radioresistant. The diagnosis and the management of SEP are controversial and varying opinions have been given in various textbooks. The consensus probably is that they need to be treated by radiotherapy as they are mostly radiosensitive. However, there are no guidelines mentioned for the management of radioresistant SEP. This case of radioresistant SEP of larynx was managed by endoscopic CO_2 excision. This is a viable option for tumors limited to the larynx. The aim of this case report is to highlight the rarity of radioresistant SEP and the enigma in the management of such cases.

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