LARYNGEAL LEIOMYOSARCOMA: A RARE MALIGNANT TUMOUR

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INTRODUCTION

Laryngeal malignancies account for approximately one percent of all malignancies diagnosed annually in men. The incidence is roughly 5 per 100,000 populations. Most of these are squamous cell carcinomas, sarcomas or malignant tumours of mesenchymal origin account for only 1 percent [1].

Leiomyosarcoma is a malignant tumour of smooth muscle origin with the reported incidence of 5-6 percent of all soft-tissue sarcomas. Majorities (eighty-five per cent) develop in the extremities and approximately three per cent occur in the head and neck region [2]. This low incidence is due to the scarcity of smooth muscle in the head and neck, where it is limited to vessel walls, the erector pili muscle of the skin, the posterior wall of trachea, and the muscular wall of the middle and lower third of the oesophagus [3]. Mindell et al [4] found that the most common site of presentation in the head and neck were the scalp and superficial soft tissues. Frank reported the first case of laryngeal leiomyosarcoma in 1941 [5-7]. Since then, in the literature there have been approximately 52 reported cases of this laryngeal tumour [8]. We present a case of leiomyosarcoma of the larynx, treated by surgery alone and without any local or regional recurrence, 36 months after the surgery. This is second such case from Pakistan. The first was reported by Abbas et al of Aga Khan University Hospital, Karachi [8].

CASE REPORT

A 55-year-old man presented in ENT department in May 2004 with history of progressive hoarseness of voice of one year and breathlessness of 2-month duration. Dysphonia and breathlessness were insidious in onset and slowly progressive. There was no history of dysphagia, pain, cough or haemoptysis. He denied any history of smoking, pan / betelnut chewing and taking alcohol. His nose, oral cavity, oropharynx and nasopharynx were normal. Indirect laryngoscopy revealed large, smooth surfaced mass involving right vocal cord and obliterating the glottis. The vocal cord was not mobile. Both the aryepiglottic folds were normal and there was no pooling of saliva in any pyriform fossa. Laryngeal crepitus was intact and no cervical lymph node was palpable. Thoracic examination did not reveal any abnormality.

The possible risks and progressive nature of the mass was discussed with the patient and his attendants; and direct laryngoscopy under general anaesthesia planned. During anaesthesia, endotracheal tube could not be passed and emergency tracheostomy was performed. At direct laryngoscopy, a mucosa covered, firm to hard mass was found involving the right vocal cord and almost completely occluding the airway. Biopsy was taken and debulking of the mass done. Left vocal cord and pyriform fossae were normal. Subglottic extension could not be ascertained. Histopathology of the specimen revealed a malignant neoplasm, possibly sarcoma or sarcomatous variant of carcinoma. At follow up indirect laryngoscopy, mass was found slightly reduced in size and the right vocal cord was immobile. Left vocal cord was normal. Computerized tomographic scan revealed a 2.5 cm high, transglottic mass involving right side of the larynx, crossing the midline anteriorly and almost completely occupying the lumen of the infraglottic larynx (fig). Right aryepiglottic fold was found thickened with possible erosion of the right arytenoid cartilage. Thyroid cartilage was intact and thyroid gland was not infiltrated. Total laryngectomy was performed on 20th July 2004. Post-operative recovery remained uneventful. The resection specimen consisted of total larynx with pedunculated mass arising from the whole right vocal cord and going down into the subglottis. The mass was

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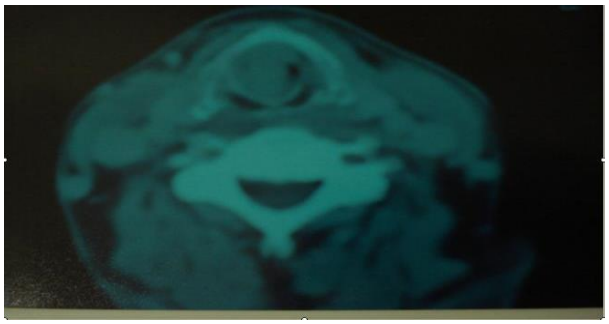

Fig: Axial CT scan demonstrating the soft tissue mass almost completely occluding the lumen of the infraglottic larynx

firm in consistency and measured 3x2.5x1 cm. Histopathology confirmed the diagnosis of leiomyosarcoma. The resection margins were free of tumour. Immunohistochemistry showed tumour cells positive for actin and negative for cytokeratin AE1, AE3, MFN116. Follow up of the patient did not reveal any local or regional recurrence 36 months after the operation.

DISCUSSION

Laryngeal leiomyosarcoma is extremely rare. It occurs at an average age of fifth to sixth decade of life. Smoking and alcohol consumption are not risk factors [6, 9]. Review of 26 published cases by Marioni et al [5] showed that 24 were men and 2 women. The present case and the other one reported by Abbas et al of Aga Khan Hospital Karachi were also males. This may be indicative of that the tumour is relatively more common in men than women. The presentations of laryngeal leiomyosarcoma do not differ from that of other laryngeal malignant tumours. Common symptoms are hoarseness, and dyspnoea. They rarely present with lymph node metastasis. Histological diagnosis of this tumour is very difficult by conventional light microscopy and immunohistochemical studies are necessary [9]. The tumour cells are positive for vimentin, smooth muscle actin, but negative for keratin. Electron microscopic examination is also a useful aid to the diagnosis. The differential diagnosis includes leiomyoma, spindle cell sarcoma, myofibrosarcoma, rhabdomyosarcoma, malignant melanoma, fibrosarcoma, malignant schwannoma and sarcomatoid carcinoma [5, 9, 10].

The treatment of leiomyosarcoma of the head and neck has generally been a wide local excision. Radical neck dissection is usually withheld unless there is regional lymph node metastasis. The role of post-operative radiotherapy remains to be defined. Wadhwa et al [9] reported a local recurrence rate of 35-50 % whereas our case remained free of local or regional recurrence 36 months after the operation. Because of the small number of reported cases, survival statistics for laryngeal leiomyosarcoma are inconclusive. Data from previous studies show that patients with head and neck leiomyosarcoma generally have a 5-year survival of 35-50% [11].

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