Neurofibrosarcoma of the Subglottis—A Rare Case Report

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Abstract A frazzled man in his early thirties presented with a 4 year long history of frequent progressive airway obstruction. He had undergone two open laryngeal surgeries and multiple endolaryngeal procedures for excision of a subglottic lesion which recurred. Each time the histopathology showed a different entity ranging from hemangioma and fibrihystiocytic lesion and spindle cell carcinoma. Finally the case was proven to be an intermediate grade sarcoma of neurogenic origin. He underwent widefield laryngectomy with placement of Provox-2 prosthesis followed by radiotherapy. Five years following surgery, he is progressing well. The case is being reported for its rarity and also for the histogenetic problems associated with this group of lesions. Authors found only one other case of laryngeal neurofibrosarcoma that was reported 29 years back [3].

Keywords Neurofibrosarcoma · Subglottis · Larynx

Introduction

Non-epidermoid tumors of the larynx represent an extremely diverse group of diseases with varying prognosis and rationales of management. Sarcoma is one of them. The symptoms are nonspecific like dysphagia, pain, hoarseness and stridor. Clinically one commonly sees polypoid submucosal lesions. With the lack of specificity in symptoms and physical findings

lesion characterization depends on high index of suspicion by the clinician as well as accurate histopathology.

Case Report

A 32 year old factory worker presented with history of repeated surgical procedures for the larynx for airway obstruction. He had no other co-morbidities and gave no history of smoking or alcohol consumption. In a span of 4 years, he had undergone two open laryngeal procedures for excision of subglottic masses, with a gap of 2 years between them. The initial episode had concluded with the diagnosis of a subglottic hemangioma and hence his tracheostomy was closed. The second time, following tracheostomy imaging studies with CT scan showed a subglottic mass with mild irregularity of the cricoid cartilage. The histopathology of the excised specimen was reported as spindle cell carcinoma. The patient was advised to undergo adjuvant chemoradiation; but he did not comply and he had got himself decannulated. A year and a half following this, the initial symptoms recurred and he had to be re-tracheostomized for the third time.

Subsequent general examination done at our centre was unremarkable, except for the tracheostomy and healed scars of previous open laryngeal procedures. Laryngoscopy showed normal mobile vocal cords with a pink mass in the subglottis. Microlaryngoscopy and excision of pedunculated mass arising from the posterior aspect of the subglottis was done. It was reported as a benign mass with possibility of fibrohistiocytic lesion and paraganglioma. He was then decannulated for the third time.

He returned within 5 months with a new tracheostomy and occasional bleeding from the stoma. CT Scan showed a homogenous mass extending from the subglottis upto the third tracheal ring.

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A detailed laryngotracheoscopy showed a smooth pink highly vascular mass arising from the posterior aspect of the subglottis with attachment to the cricoid cartilage extending into the tracheal lumen. Superiorly it was involving the posterior commissure. Biopsy revealed an intermediate grade sarcoma of probable neurogenic origin. It expressed S100 protein. It was negative for CK, EME, HMB, CIS, SMA, Desmin, Calponin, Alk1, Synaptophysin, Chromogranin, CK7, CK20, CD24, CD23, CD60 and CD30. A thorough metastatic work up showed no signs of local or distant metastasis (Figs. 1, 2 and 3).

The patient underwent widefield laryngectomy with placement of speech prosthesis. The surgical specimen confirmed the diagnosis. Disease was staged as stage 1 (rP T4 N0 G2 M0 R0). He progressed well and underwent adjuvant radiotherapy.

Five years post surgery, patient has reported doing well.

Discussion

Neurofibrosarcomas have appeared spasmodically in medical literature since the time of Virchow. They constitute 5 % of soft tissue sarcomas [1] and 20 % of these are seen in the head and neck region [2]. About 50 % of neurofibrosarcomas are associated with neurofibromatosis type I, and the rest are sporadic.

Less than 1 % of laryngeal cancers are sarcomas [3]. Histologically they are classified as fibrosarcomas, chondrosarcomas, rhabdomyosarcomas, osteosarcomas, leiomyosarcomas and

Fig 2 Spindle shaped cells arranged in whorls

neurofibrosarcomas. Of these, chondrosarcomas are the commonest and fibrosarcomas follow. Even after thorough search of various medical journals, authors could find only one previous case of laryngeal neurofibrosarcoma that was reported in 1980 [3]. The patient was a 73 year old male who had a lump on the right thyroid ala for which he underwent right hemilaryngectomy. Post surgery he developed both regional and distant metastasis and within 6 months succumbed to the latter.

There are numerous synonyms used for neurofibrosarcomas, of which malignant schwannoma, malignant neurofibroma and

Fig 1 CT Scan Neck showing mass in the subglottis extending into trachea, eroding cricoid

Fig 3 Laryngectomy specimen showing mass in the subglottis

malignant nerve sheath tumors are commonly used [4]. The tumor is derived from connective tissue of the peripheral nervous system. The term neurogenic sarcoma is misleading as it is not ectodermal in origin. The term fibrosarcoma of the nerve sheath expresses more accurately the pathologic concept of the tumor.

Majority of the spontaneous cases of neurofibrosarcomas occur in the age range of early thirties to fifties, the mean age of diagnosis being 10 years younger in neurofibromatosis type II patients. There is equal gender preponderance. The most common presentation in the head and neck is painless neck mass.

Histopathology is the mainstay of diagnosis. The differential diagnosis includes malignant fibrous histiocytoma, fibrosarcoma and leiomyosarcoma, but the differentiation is difficult. Earlier, one relied on the presence of pre-existing neurofibroma or origin from a nerve trunk to reach diagnosis. Now immunohistochemistry plays a vital role in histological differentiation of sarcomas. While fibrosarcomas are marker negative except for Vimentin, neurofibrosarcomas are positive for both Vimentin and S100 protein. Frequent mitosis and tissue invasion helps differentiate neurofibrosarcomas from neurofibromas. Neurofibrosarcomas are graded into low, intermediate and high grade tumors on the basis of cellularity and nuclear atypia.

Management of neurofibrosarcomas includes wide local excision with an attempt to resect the involved nerve proximally and distally and obtain clear margins. Since lymphatic spread is rare, prophylactic neck dissection is not done unless node involvement is seen clinically or radiologically [5, 6]. Adjuvant radiotherapy is best tailored by multi-disciplinary

tumor board based on individual patient and tumor factors. One third of the patients do develop distant metastasis especially in the lungs. However the role of chemotherapy remains poorly defined.

Disease free survival in neurofibrosarcomas is 35 and 24 % at 5 and 10 years respectively and the overall survival is 47 to 61 % at 5 years and 29 to 52 % at 10 years [2, 7]. Patients with high grade tumors, positive tumor margins, neurofibromatosis and tumors larger than five cms tend to have worse survival.

Conclusion

Sarcomas are a rare, but important differential diagnostic possibility in laryngeal submucosal soft tissue masses. An awareness of their existence, common clinical behaviour and high index of suspicion are essential for early diagnosis of the lesion and institution of appropriate treatment. Expert pathological review and classification are critical in reaching proper diagnosis. When examining literature on neurofibrosarcomas arising in other organs, it appears that radical complete surgical excision is the treatment of choice with post operative radiotherapy if indicated [4]. Lymphnode dissection is required only in the presence of node involvement.

Due to limited number of cases so far reported in the larynx, elective treatment can only be tentatively indicated. Further contributions will help bring to focus the treatment and predict prognosis of laryngeal sarcomas of neurogenic origin.

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