A 54 years female presented with 6 months history of dyspnea and 2 months history of haemoptysis. She was evaluated with CT thorax which showed 22×16×15 mm sized hypodense soft tissue lobulated lesion in right lower trachea extending inferiorly into right side of carina, up to origin of right main bronchus. Bronchoscopic guided biopsy was done which was S/O adenoid cystic carcinoma, grade 1. Patient was treated with definitive concurrent radiotherapy (IMRT 66Gy/33#/6.3 weeks) and weekly chemotherapy with Cisplatin. Post treatment CT thorax showed 16×15 mm sized heterogeneously enhancing soft tissue enhancing lesion at right side tracheal bifurcation. She was started on tab Gefitinib 250 mg once a day, tolerating it well since 19 months with minimal residual disease as per CT thorax. Tracheal tumors are very rare with an incidence less than 0.2 per 100,000 persons per year. Squamous cell carcinoma is most common followed by adenoid cystic carcinoma. It’s a very indolent tumor with prolonged clinical course with a tendency for local recurrence and late metastasis. Adenoid cystic carcinoma is not associated with smoking as causative factor unlike squamous cell carcinomas. It has got better prognosis than squamous cell carcinomas. Most of these tumors are detected in middle age in 4th and 5th decade and it has got almost equal distribution in both sexes. It is asymptomatic initially when the size is small, but as the size grows, it may cause hoarseness, cough, haemoptysis, wheezing, chest pain, dysphagia etc. Most of these patients are treated for bronchial asthma or bronchitis for a long time, before getting detected.

INTRODUCTION

Tracheal tumors are very rare tumors and adenoid cystic carcinoma is second common after squamous cell carcinoma. It has an indolent presentation and clinical course with various symptoms which mimic benign respiratory diseases leading to its late presentation and diagnosis. High index of suspicion is required for early diagnosis of adenoid cystic carcinoma.

Case report

A 54 years non-smoker female presented in January 2016 with 6 months history of dyspnea and 2 months history of haemoptysis off and on. She was treated in periphery with bronchodilators and steroids, considering her a case of chronic bronchitis. As she started having haemoptysis off and on, she was referred here for further evaluation. She was evaluated with CT thorax which showed 22×16×15 mm sized hypodense soft tissue lobulated lesion in right lower trachea extending inferiorly into right side of carina, up to origin of right main bronchus (Fig. 1). Bronchoscopic guided biopsy was done which was S/O Adenoid cystic carcinoma, grade 1 with tubular and cribriform pattern (Fig. 2). There was no evidence of disease elsewhere in body. Multidisciplinary discussion was done in view of rarity of the tumor and it was labelled unresectable because of involvement of carina. Patient was treated with definitive concurrent radiotherapy (IMRT 66Gy/33#/6.3 weeks) and weekly chemotherapy with Cisplatin.
Fig. 1. CT thorax showing tracheal tumor in right lower trachea

Fig. 2A. Biopsy showing invasive tumor comprising of basaloid cells arranged in cribriform pattern S/O adenoid cystic carcinoma (H&E,x10)

Fig. 2B. Biopsy showing invasive tumor comprising of basaloid cells arranged in cribriform pattern S/O adenoid cystic carcinoma (H&E,x40)

Fig. 3. CT thorax showing minimal residual tracheal tumor in right lower trachea with calcification

She is asymptomatic, on regular follow up and tolerating Gefitinib well since 19 months with minimal residual disease as per CT thorax.

DISCUSSION

Tracheal tumors are very rare with an incidence less than 0.2 per 100,000 persons per year (Yang, 2005). Squamous cell carcinoma is most common followed by adenoid cystic carcinoma (Li, 1990). It’s also called ‘Cylindroma’ and was first reported by Bilroth in 1859. It’s a very indolent tumor with prolonged clinical course with a tendency for local recurrence and late metastasis. The site of origin of most of these tumors is distal third of trachea. Adenoid cystic carcinoma is not associated with smoking as causative factor unlike squamous cell carcinomas. It has got better prognosis than squamous cell carcinomas. The median survival is 9 years vs. 9 months in squamous cell carcinomas (Bhattacharyya, 2004). The etiology is unknown but some possible carcinogens have been reported. Dohyung Kim et al & Mancuso et al showed possible role of Nitroso compounds found in rubber fumes in exposed workers in rubber industry (Kim et al., 2013; Mancuso, 1970). Most of these tumors are detected in middle age in 4th and 5th decade and it has got almost equal distribution in both sexes (Bhattacharyya, 2004). Adenoid cystic carcinoma is primarily a tumor of salivary glands but also occurs with low frequency in cervix, skin, lungs, breast, and trachea etc (Albers et al., 2004).

As it basically a salivary gland tumor, so the clinicopathological characteristics as well as management guidelines are based mainly on the lines of salivary gland tumors. CT thorax is investigation of choice. MRI may show better soft tissue and neural invasions. X-rays are seldom useful. Adenoid cystic carcinoma usually spreads by submucosal and perineural routes. At the time of diagnosis, less than 10% of patients have local or distant metastasis, possibly because of low vascularity and scanty lymphatics in trachea as well as low aggressiveness of tumor. Lung metastasis is most common but metastasis in brain, bone, liver, kidney, skin and heart are also reported (Gaissert ha, 2006). It is asymptomatic initially when the size is small, but as the size grows, it may cause hoarseness, cough, hemoptysis, wheezing, chest pain, dysphagia etc. Most of these patients are treated for bronchial asthma or bronchitis for a long time, before getting detected. Another cause for late detection is large functional reserve of trachea, as there are no
or minimal symptoms before it occludes approximately half to two third of tracheal lumen. There are 2 definitive treatment modalities, surgery and radiotherapy. Surgery is the treatment of choice followed by adjuvant radiotherapy in resectable subset of tumors. The criteria for unresectability are extension to carina, mediastinal extension, long length of involved trachea and multiple mediastinal lymphadenopathies (Grillo, 1981). The role of postoperative adjuvant radiotherapy is uncertain. Some studies have used adjuvant radiotherapy to all, whereas others have used when margins are involved or close. In the absence of randomized trials, it is reasonable to give adjuvant radiotherapy for all patients undergoing resection. Radiotherapy should be started at least 1 month after surgery and a bronchoscopy before radiotherapy to be done to ensure healing. Postoperative radiotherapy is recommended in a dose range of 45–65 Gy depending on the risk factors and margin status (Gaissert ha, mark, 2006). Radiation is also used in definitive treatment of unresectable tumors and palliative treatment of tumor for symptomatic relief. Gaissert et al has published large case series of 135 patients during 40 years. Most of these patients were treated by surgery followed by adjuvant radiotherapy in 70%. In unresectable patients, radiotherapy alone was given.

The 5 year survival in surgical group was 52% vs. 30% in radiation alone group (Gaissert ha, mark, 2006). Concurrent chemotherapy can be given, but there is no concrete evidence to support it. We too have given concurrent Cisplatin with radiotherapy in view of unresectable disease, which was tolerated well. Neutron radiotherapy has been shown to be useful in advanced adenoid cystic carcinoma of the trachea in a study by Bittner et al showing 5-year overall survival of 89% and 5-year locoregional control rate of 54% out of 20 patients (Bittner, 2008). In management of metastatic disease, there is very limited role as well as scanty data on chemotherapy. There are small series in patients of adenoid cystic carcinoma of salivary glands in which drugs like Cisplatin, Mitoxantrone, Epirubicin, Doxorubicin, 5-Fluorouracil have shown some response while drugs like Docetaxel, Paclitaxel, Cyclophosphamide have shown very poor results. There are data about combination chemotherapy with Cisplatin plus 5-Fluorouracil, Cyclophosphamide, Vincristine plus 5-Fluorouracil which has showed some success (Papaspyrou, 2011; Dodd, 2006). A variety of targeted agents have been tried in metastatic disease like Bortezomib, Imatinib, Lapatinib, Cetuximab, Panitumumab, Sorafenib, Gefitinib etc. these drugs haven’t shown much response, with their role limited to disease stabilization in some patients (Dodd, 2006; Alcedo et al., 2004; Locati, 2009; Agulnik, 2007; Argiris, 2006).

REFERENCES


