Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 10, Issue, 02, pp.65120-65122, February, 2018

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

ISSN: 0975-833X

CASE STUDY

ADENOID CYSTIC CARCINOMA OF TRACHEA- A RARE CASE WITH REVIEW OF LITERATURE

[1,*]Amitabh Kumar Upadhyay, [2]Pankaj goyal, [3]Nidhi Gupta, [4]Rakesh kumar Gupta and [5]Sandeep kukkar

[1]Consultant Medical Oncologist, Grecian hospital, Mohali
[2]Consultant Medical Oncologist, RGCIRC, Delhi
[3,4]Consultant Radiation Oncologist, Grecian hospital, Mohali
[5]Consultant Medical Oncologist, Mohali

ARTICLE INFO

Article History:
Received 20th November, 2017
Received in revised form
23rd December, 2017
Accepted 15th January, 2018
Published online 18th February, 2018

Key words:
Adenoid Cystic Carcinoma,
Tracheal Tumor.

ABSTRACT

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. Brochoscopic 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, hemoptysis, wheezing, chest pain, dysphagia etc. Most of these patients are treated for bronchial asthma or bronchitis for a long time, before getting detected. There are 2 definitive treatment modalities, surgery and radiotherapy. Surgery is the treatment of choice with adjuvant radiotherapy in resectable tumors. Radiation is used in multiple indications like adjuvant after surgical procedure, definitive treatment of unresectable tumors and palliative treatment of tumor for symptomatic relief.

Citation: Amitabh Kumar Upadhyay, Pankaj goyal, Nidhi Gupta, Rakesh kumar Gupta and Sandeep kukkar. 2018. "Adenoid cystic carcinoma of trachea- A rare case with review of literature", *International Journal of Current Research*, 10, (02), 65120-65122.

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). Brochoscopic 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.

Corresponding author: **Amitabh Kumar Upadhyay,**
Consultant Medical Oncologist, Grecian hospital, Mohali.

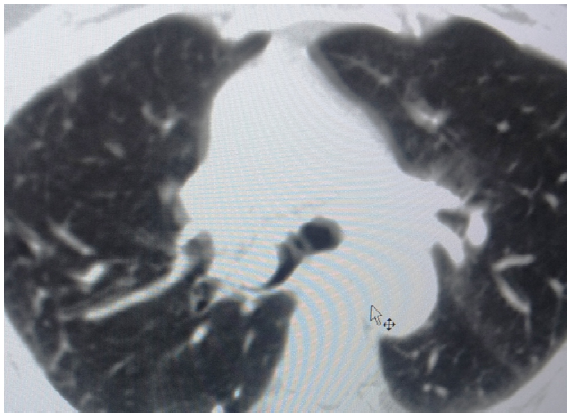

Fig. 1. CT thorax showing tracheal tumor in right lower trachea

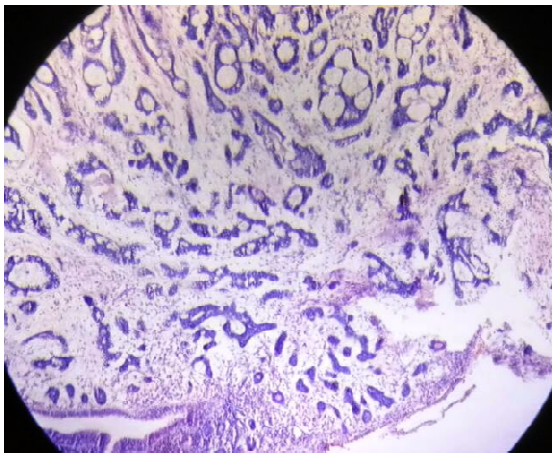

Fig. 2A. Biopsy showing invasive tumor comprising of basaloid cells arranged in cribriform pattern S/O adenoid cystic carcinoma (H&E,x10)

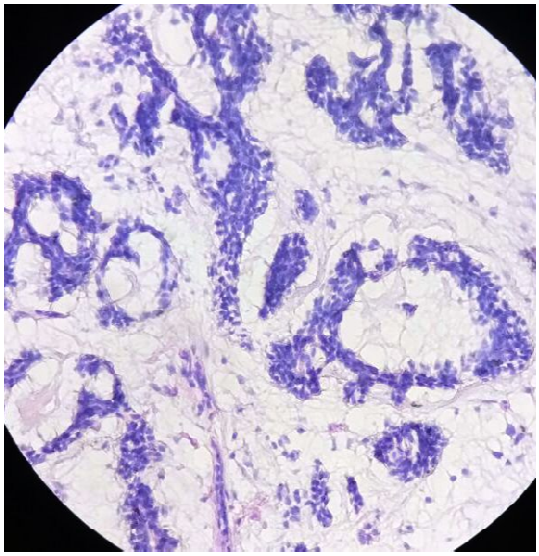

Fig. 2B. Biopsy showing invasive tumor comprising of basaloid cells arranged in cribriform pattern S/O adenoid cystic carcinoma (H&E,x40)

Post treatment CT thorax showed 16×15 mm sized heterogeneously enhancing soft tissue enhancing lesion at right side tracheal bifurcation (Fig. 3). Surgical option was discussed with patient in view of residual disease but refused by the patient. She was started on tab Gefitinib 250 mg once a day, in view of rarity of disease, non-willingness for surgery and no robust data about any role of chemotherapy or targeted therapy.

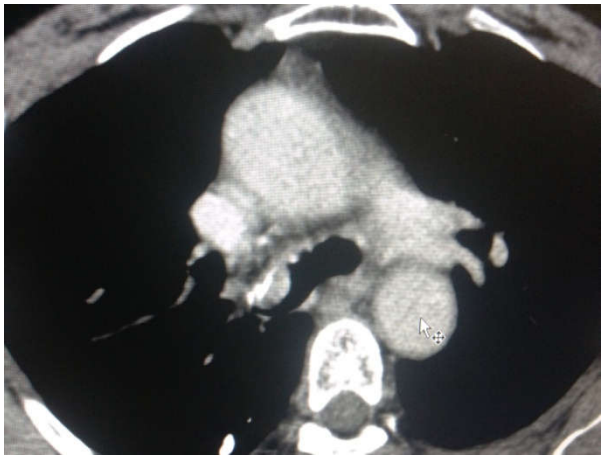

Fig. 3. CT thorax showing minimal residual tracheal tumor in right lower trachea with calcification

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 4^{th} and 5^{th} 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 clinico-pathological 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

Agulnik M, Cohen EW, Cohen RB. *et al*. 2007. Phase ii study of lapatinib in recurrent or metastatic epidermal growth factor receptor and/or erbb2 expressing adenoid cystic carcinoma and non adenoid cystic carcinoma malignant tumors of the salivary glands. *J clin oncol.,* 25:3978– 3984.

Albers E, lawrie T, harrell Jh, Yi ES 2004. tracheobronchial adenoid cystic carcinoma—a clinicopathologic study of 14 cases. Chest 125:1160–1165

Alcedo JC, fa´brega JM, arosemena JR, Urrutia A. 2004. Imatinib mesylate as treatment for adenoid cystic carcinoma of the salivary glands: report of two successfully treated cases. Head neck., 26:829–831

Argiris A, goldwasser MA, burtness B, deconti R, Axelrod R, van waes C. 2006. A phase ii of ps-341 (bortezomib) followed by the addition of doxorubicin at progression in incurable adenoid cystic carcinoma of the head and neck: an eastern cooperative oncology group study. Proc am soc clin oncol 24 (suppl 298s):5573

Bhattacharyya N. 2004. Contemporary staging and prognosis for primary tracheal malignancies: a population-based analysis. Otolaryngol head neck surg 131:639–42

Billroth T. 1859. Beobachtungen fiber geschwulste der speicheldriisen. Virchow arch path anat.17(5):357–75. Http://dx.doi.org/10.1007/bf01930503

Bittner N, Koh WJ, laramore GE. 2008. Neutron radiotherapy of tracheal acc. Int j radiat oncol biol phys.72:410–4.

Dodd RL, slevin NJ. 2006. Salivary gland adenoid cystic carcinoma: a review of chemotherapy and molecular therapies. *Oral oncol.,* 42:759–769.

Gaissert HA, mark EJ. 2006.Tracheobronchial gland tumors. Cancer control.13:286–94.

Grillo HC 1981. Tumors of the cervical trachea. In: suen jy, myers en (eds) cancer of the head and neck. Churchill livingstone, new york, pp 500–522

Kim D, hwang Y, choi S, park C, lee N, kim EA 2013. A case of tracheal adenoid cystic carcinoma in a worker exposed to rubber fumes. Ann occup environm med 25:22

Li W, ellerbroek NA, libshitz HI. 1990. Primary malignant tumors of the trachea: a radiologic and clinical study. Cancer. 66(5):894–9. Http://dx.doi. Org/10.1002/1097–0142

Locati Ld, bossi P, perrone F. *et al*. 2009. Cetuximab in recurrent and/or metastatic salivary gland carcinomas: a phase ii study. *Oral oncol.,* 45:574–578

Mancuso TF. and brennan MJ 1970. Epidemiological considerations of cancer of the gallbladder, bile ducts and salivary glands in the rubber industry. *J Occup Med.,* 12(9):333–341

Papaspyrou g, hoch s, rinaldo a, rodrigo jp, takes rp, van herpen c, werner ja, ferlito a. 2011. Chemotherapy and targeted therapy in adenoid cystic carcinoma of the head and neck: a review. Head neck, 33(6):905-11. Doi:10.1002 /hed.21458.

Yang PY, liu MS. and chen CH. 2005. Adenoid cystic carcinoma of the trachea. *Chang gung med j*. 28(5):357–63.

****