

Case Report

Definitive radiotherapy for inoperable adenoid cystic carcinoma of the trachea: A rare case report

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ABSTRACT

Adenoid cystic carcinoma (ACC) of the trachea is rare; it represents 1% of all respiratory tract cancers. It is generally considered as a slow-growing, with prolonged clinical course. Most patients present with dyspnea, and the symptoms often mimic those of asthma or chronic bronchitis. Surgical resection is the mainstay of treatment often combined to radiotherapy because of close surgical margins. When surgery is not possible, most tumors respond to radiotherapy alone which often results in long periods of remission. There is no consensus on the best treatment for locally advanced inoperable ACC of trachea. This case report describes a 51-year-old woman unresectable ACC of trachea due to comorbid conditions, successfully managed by intensity modulated radiotherapy. At 8 months follow-up, the patient is healthy and asymptomatic.

KEY WORDS: Adenoid cystic carcinoma, radiotherapy, trachea

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INTRODUCTION

Adenoid cystic carcinoma (ACC) is a malignant tumor of salivary glands. The incidence of tracheal tumor ranges from 0.09% to 0.2% of all cancers and are mainly squamous cell carcinomas. Only 10% of all tracheal carcinomas are ACC.^[1,2] These patients present with dyspnea and wheezing. Long latency is usually observed before presentation because of its slow growth rate. It infiltrates locally and can be life-threatening due to central airway obstruction. Surgical excision is the primary treatment, but mostly it is unresectable due to large tumor size, local extension, or other comorbidities. The standard treatment for inoperable ACC of trachea is radiotherapy. Here, we present a case of inoperable tracheal ACC managed with Radiotherapy.

CASE REPORT

A 51-year-old female presented in March 2015 with chronic cough and breathlessness since 18 months.

She was being treated for bronchial asthma without any relief in the last 9 months. Presently, her symptoms worsened with increasing shortness of breath for which she was investigated. Her computed tomography (CT) scan thorax [Figure 1] showed an exophytic mass from mid-trachea to 3.4 cm proximal to carina causing marked luminal obstruction. It is indenting on adjacent esophagus, brachiocephalic artery with loss of fat planes. There is no enlarged lymph node. On bronchoscopy, a pinkish polypoidal tracheal growth measuring 1 cm × 1.5 cm in the lower third of trachea was seen with near total luminal obstruction. Histopathology confirmed it as ACC [Figures 2a and b]. No distant metastasis was seen.

Surgical intervention was not possible due to extensive local disease; hence, the patient was planned for local radiotherapy. A total dose of 54 Gy/30 fractions over 6 weeks using 6 MV photons with intensity-modulated

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radiotherapy was given limiting the dose to adjacent normal organs. During the first 2 weeks, the patient was treated with oxygen support. Later on, the treatment was well tolerated with manageable side effects.

At 8 months postradiotherapy, the patient is asymptomatic, and her CT scan shows lobulated moderately enhancing wall thickening in the left lateral wall of mid-trachea 2.1 cm × 1.5 cm causing mild luminal narrowing [Figure 3]. Her bronchoscopy did not show any mass in the lumen of trachea.

DISCUSSION

ACC of the trachea is rare. The incidence is <0.2/100,000 people per year and only 10% of these cases are ACC. ACCs have been reported without sex predilection in



Figure 1: Computed tomography scan thorax showing growth in trachea lumen

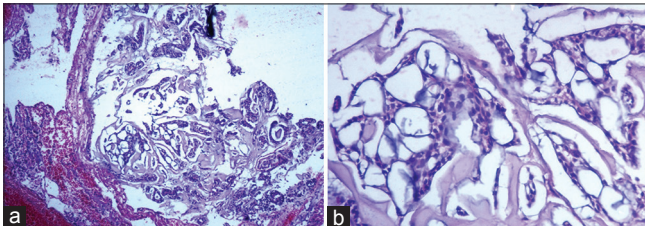


Figure 2: Tracheal biopsy shows the adenoid cystic carcinoma (a) the tumor has a cribriform (arrows) and tubular pattern (×10). (b) The pseudoglandular spaces contain basophilic basement membrane-like material (×20)

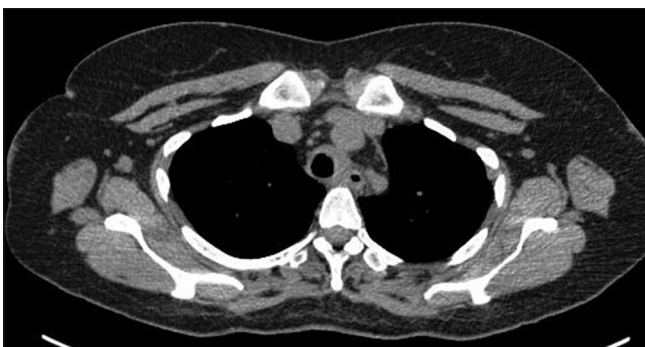


Figure 3: Postradiotherapy (4 months) regression of growth in computed tomography scan thorax

the fifth decade of life and smoking does not affect the incidence. These patients present with cough and expectoration, wheeze, dyspnea, and are often treated as asthma.^[1] ACC is a nonencapsulated tumor and spreads by direct extension, submucosal, or perineural invasion in transverse and longitudinal planes. Lymphatic spread is uncommon. More than 50% of patients have hematogenous metastases. Pulmonary metastases are most common. Metastases to the brain, bone, liver, kidney, skin, and heart are reported.^[1-3] On CT scan, the tumor infiltration in the submucosa manifests as an intraluminal mass with extension through the tracheal wall with a diffuse or circumferential wall thickening^[2] as is also seen in our case. Magnetic resonance imaging can better define the extent of submucosal infiltration and local mediastinal invasion which can influence resectability. Treatment options include surgery, radiotherapy, or a combination. The ideal treatment of ACC is primary resection and end-to-end anastomosis. The median survival time of resected patients was reported to range from 90 to 118 months.^[4] Because they tend to infiltrate along the airways, ACCs are often incompletely resected. Negative surgical margins are difficult to obtain because of relative inability to resect more than 6 cm of trachea.^[5] No significant difference in survival is seen in completely or incompletely resected ACCs.^[6] Maziak showed 5-year survival rates of 82% and 77% for completely and incompletely resected patients.^[7] In contrast, Gaissert *et al.*^[8] showed significantly longer survival with negative margins. The lack of statistical significance on survival benefit for negative margins is likely due to lower incidence and inadequate follow-up. ACC is a radiosensitive tumor as long periods of remission is seen after radiotherapy.^[3] ACCs often traverse along tracheal wall in vertical direction, a margin of 4–5 cm in craniocaudal axis was suggested by Kaminski *et al.*^[6]

Chow *et al.*^[9] cautions against giving doses higher than 60 Gy, as 50% of his cases had tracheoesophageal fistula, esophageal stricture, and severe tracheal crusting requiring surgical intervention when dose in excess of 60 Gy was given, whereas none of the patients treated with <60 Gy had late side effects. Other authors have corroborated the concern of high doses contributing to the risk of complications; hence, we gave a dose of 54 Gy to our patient with a good local control.

In the largest series, Gaissert *et al.*^[8] reviewed 135 patients with tracheal ACC and showed a 52% and 29% in resected but only a 33% and 10% 5- and 10-year survival, respectively, in unresectable cases although 40% of resected cases had positive cut margins. Maziak *et al.*^[7] reported a mean survival time of 37 months (4 months - 7 years) in patients receiving radiotherapy.

Neutron radiotherapy has been shown to be effective in advanced unresectable ACC of the trachea in a recent study by Bittner *et al.*,^[10] they showed a 5-year overall survival rate of 89% and a 5-year locoregional control rate of 54% in their cohort of twenty patients.

The role of postoperative adjuvant radiotherapy remains uncertain. Some advocate a postoperative radiotherapy to all, whereas others offer this treatment when margins are invaded. With limitations of randomized trials, it is reasonable to recommend adjuvant radiotherapy for all patients undergoing resection. Radiotherapy should be started 1 month after surgery and also perform a bronchoscopy before radiotherapy to ensure healing. Postoperative radiotherapy is recommended at 45–65 Gy depending on the quality of margins.

CONCLUSION

ACC is a rare primary tracheal malignancy commonly misdiagnosed initially as asthma. Surgical resection followed by radiotherapy is widely recommended protocol for treatment of localized tracheal tumors and provides the best chance of prolonged survival. Patients with unresectable disease can benefit long periods of remission with radiotherapy and there is no role of chemotherapy.

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Conflicts of interest

There are no conflicts of interest.

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