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CASE REPORT

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Adenoid cystic carcinoma of the trachea mimicking asthma

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Abstract

Adenoid cystic carcinoma (ACC) is primarily a salivary gland tumour that rarely involves the respiratory tract. A 58-year-old lady was admitted with worsening dyspnoea, cough and wheezing for 2 days. CT pulmonary angiogram was done due to persistent dyspnoea which revealed a 12 mm mass protruding into the posterior aspect of the trachea with multiple enlarged nodes. There was a complete collapse of the left lower lobe and right middle lobe with right upper lobe pulmonary embolism which was thought to be contributing to her hypoxia. She was struggling with secretion clearance and initial measures to clear her secretions were not successful. She was treated with a tracheal stent, followed by an interval endoscopic ultrasound-guided biopsy of the tracheal wall lesion which revealed ACC. She was referred to cardiothoracic surgeons for excision of the tumour after discussing in MDT. Surgery followed by radiotherapy is advised in cases with incomplete resection margins.

KEYWORDS

adenoid cystic carcinoma, radiotherapy, stent, surgery, trachea

INTRODUCTION

Adenoid cystic carcinoma (ACC) is primarily a salivary gland tumour that rarely involves the respiratory tract. Accurate diagnosis and timely intervention are essential in complicated cases as it has a favourable outcome compared to other tracheal tumours.

CASE REPORT

A 58-year-old lady was admitted with worsening dyspnoea for 2 days. This was associated with mild wheezing and cough. She was diagnosed to have bronchial asthma since childhood with good control without needing any hospital admission or physician review for more than 10 years. She denied having weight loss or chest pain. On examination, she was tachypnoeic but was speaking in sentences. There was no stridor. Auscultation of the chest was recorded as demonstrating diffuse wheeze. There was no salivary gland or lymph node enlargement.

She was initially managed as an exacerbation of bronchial asthma, but she did not respond to treatment. As there was ongoing dyspnoea with persistent hypoxia, CT pulmonary angiogram was done to exclude a pulmonary embolism (PE) which revealed a 12 mm mass protruding into the posterior aspect of the trachea with multiple enlarged nodes in the pre-carinal and subcarinal stations (Figure 1). There was a complete collapse of left lower lobe and right middle lobe with right upper lobe PE which was thought to be contributing to her hypoxia. She was struggling with secretion clearance and initial measures to clear her secretions were not successful.

She was electively intubated and referred to cardiothoracic surgeons and treated with a tracheal stent followed by interval endoscopic ultrasound-guided biopsy of the tracheal wall lesion. She was noted to have a significant amount of secretions at the time of tracheal stent insertion which was cleared. The case was further complicated as she needed a histological diagnosis with an intratracheal stent. Tracheal lesion was on the posterior wall and after reviewing the images it was decided to approach the lesion through endo-oesophageal ultrasound-guided fine needle aspiration which was successful. Cytopathology revealed ovoid tumour cells with bland appearing nuclei forming nests, trabeculae and cribriform spaces with hyaline materials. Immunohistochemistry highlighted both luminal/ductal epithelial cells positive for CK7 and CD117 and basal/myoepithelial cells positive for p63, p40 and Sox 10, and the

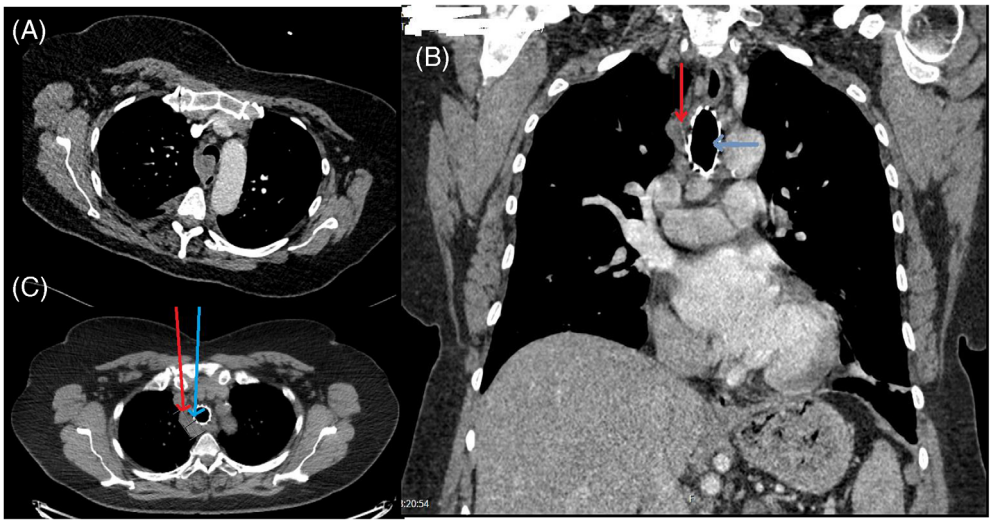

FIGURE 1 (A) CT chest showing a lesion arising from the posterior tracheal wall causing narrowing of the trachea. (B, C) Blue arrow—tracheal stent in-situ, Red arrow—paratracheal lymph nodes

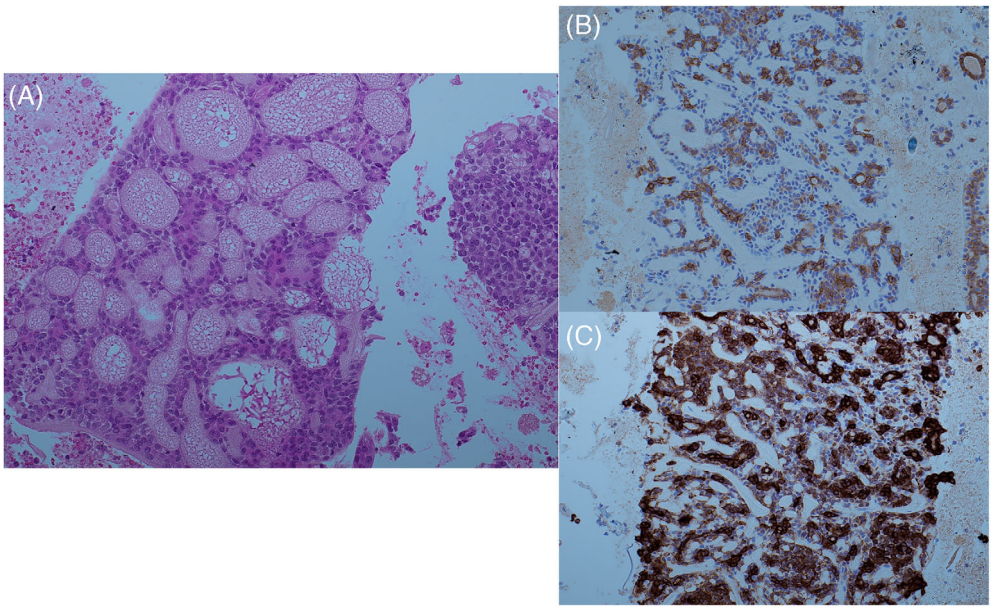

FIGURE 2 (A) H&E staining: malignant cells arranged in cribriform pattern with microcystic spaces containing hyaline/basophilic mucoid material. (B) CK7 positivity within ductal/luminal cells and negative staining within myoepithelial/abluminal cells. (C) CD117 positivity within ductal/luminal cells and negative staining within myoepithelial cells

overall diagnosis favoured a salivary gland origin tumour leading to ACC (Figure 2).

She was referred to Oncology for radiotherapy as cardiothoracic surgeons felt surgery was not possible due to surrounding infiltrative nature of the tumour.

DISCUSSION

ACC are primarily salivary gland tumours. ACC of the trachea are rare slow growing tumours that arise from mixed seromucinous glands in the trachea with an incidence of 0.04%–0.2%.[1] Two thirds of all tracheal tumours are malignant. Of these, 75% are squamous cell carcinomas (SCCs) and 15% are ACC.[2,3] Most of these patients are in the fourth to sixth decade of their life as in our case. Compared to squamous cell carcinoma ACC has a favourable prognosis and responds very well to radiotherapy and some are operable. Our patient was further evaluated with head and neck MRI followed by a PET-CT which did not show salivary gland or cervical lymph node abnormality.

ACC grows primarily in the tracheal lumen with a slow growth rate. It is rarely associated with lymph node enlargement and distant metastases. Bhattacharyya proposed a modified TNM classification system based primarily on the tumour size as the decisive factor of survival.[4] Current data suggests a favourable overall 5-year survival of >70%.[5,6] In one study it even reached 95% with a 10-year overall survival of 81%.[1] A complete resection is considered the gold standard in treating tracheal ACC. Due to the infiltrative growth of ACCs into the surrounding tissue, incomplete resection margins are often observed following surgery. According to the literature, positive resection margins after surgery occur in 8%–82% of all cases.[5,7,8] Surgery followed by radiotherapy is advised in cases with incomplete resection margins given the favourable response to radiotherapy.

Our patient presented with dyspnoea and monophonic wheeze which did not respond to initial measures. Her management was further complicated by the fact that she had a history of asthma leading to a possible differential of asthma on admission. Poor response to initial measures was a clue to address an alternative diagnosis which led to the suspicion of a PE, which was confirmed by an angiogram with an additional diagnosis of tracheal tumour. As she was symptomatic, she was treated with a tracheal stent as an emergency procedure by thoracic surgeons to which she had a dramatic response. As the primary tumour was treated with a stent, endotracheal route was not considered safe to obtain a biopsy. This alternative oesophageal approach was convenient, and it could even be used for tumour staging as we could view the left adrenal and partly the liver as well.

Disease conditions including tracheal tumours, viral/bacterial pneumonia, sinusitis, foreign bodies, allergic broncho-pulmonary aspergillosis, acid reflux can mimic asthma. Poor response to treatment should lead to further evaluation to rule out the mimics which can be life threatening in certain situations. Detailed history, appropriate radiological imaging, lung function tests, endoscopic assessment are needed for further evaluation. Optimisation of the underlying condition will improve the breathlessness and wheezing which was seen in our case.

Our patient was referred to Thoracic surgeons with expertise in tracheal surgery however, it was felt that the tumour was not operable and the decision to proceed with radiotherapy was made given the favourable outcome after discussion in MDT.

AUTHOR CONTRIBUTIONS

All authors contributed to the conception of the case report. Sugeesha Wickramasinghe and Mohommed Munavvar wrote the first draft. All authors revised subsequent versions and approved the final version.

CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

not applicable

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

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