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Central airway obstruction caused by adenoid cystic carcinoma in pregnancy: a case report and review of the literature

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Abstract

Malignancy complicates one in a thousand pregnancies. The most frequently diagnosed of these are breast, cervical, melanoma, ovarian, and haematological neoplasms. Tumours of respiratory origin are very uncommon during pregnancy. We present a case of tracheal adenoid cystic carcinoma (ACC), a rare type of primary airway tumour, diagnosed in a pregnant woman. To our knowledge, this is the third reported case of tracheal ACC complicating pregnancy. We discuss potential barriers to timely diagnosis of malignancies during pregnancy and consider optimal management strategies, taking into account the potential harm to the mother and foetus in a field with a limited evidence base.

Introduction

Primary tracheal cancers are very rare, representing less than 0.2% of all malignancies of the respiratory system [1]. Following squamous cell carcinoma (SCC), adenoid cystic carcinoma (ACC) is the most common histopathological subtype, accounting for approximately 7% of cases [2]. Compared to tracheal SCC, ACC, a tumour of salivary gland origin, usually occurs in younger non-smoking individuals [1]. Tracheal ACCs are slow-growing, and patients frequently present with very large tumours causing obstructive symptoms. The presentation can be commonly initially mistaken for asthma or bronchitis [3]. Bronchoscopy allows direct visualization of the lesion, and imaging modalities such as computed tomography (CT) or positron emission tomography (PET) can demonstrate the local and distant extent of the disease [4].

Treatment of tracheal ACC is based on management strategies for head and neck salivary gland tumours, where ACC is a more common entity. Surgical resection is the treatment of choice for local tumours [5]. However, there is a high rate of positive surgical margins, as well as a high likelihood of regional recurrence or late distant metastases due to early perineural invasion and extension along vascular structures [5,6]. Adjuvant radiation therapy is usually utilized for pathologies with positive surgical margins. Radiation therapy alone or concurrent chemoradiotherapy with a platinum-based regimen is suggested in unresectable cases [7]. Studies show the 5- and 10-year survival rates for tracheal ACCs are 73% and 57%, respectively [8].

Case Report

A 27-year-old, G2P1, non-smoking female of Southeast Asian background presented to her local emergency department at 26 weeks gestation with a one-month history of chest discomfort, cough, and haemoptysis, with a background of asthma diagnosed three years previously. The pregnancy was otherwise uncomplicated. Chest X-ray (CXR) was not performed initially to avoid foetal radiation

exposure. She was commenced on steroid inhalers and oral antibiotics to treat a presumed mild chest infection. The patient represented five days later with persistent symptoms. Chest X-ray showed collapse/consolidation in her left lung (Fig. 1A). She was admitted to the hospital for further investigation and was commenced on intravenous antibiotics, bronchodilators, and steroids for management of possible pneumonia. She underwent diagnostic video-bronchoscopy due to persistent symptoms. This revealed an obstructing lesion partly pearly in appearance and soft in consistency, visualized at the entrance of the left main bronchus (Fig. 1B). Biopsy revealed an ACC.

Bronchoscopic snare resection to provide symptomatic relief was originally planned, but her symptoms improved and stabilized, and a repeat CXR showed spontaneous re-expansion of the left lung, although air entry remained poor. On weighing the risk-benefit of bronchoscopic resection in the antepartum period, it was agreed through multidisciplinary discussion between her obstetrician, respiratory physician, cardiothoracic surgeon, and anaesthetist that such an intervention was not required but that definitive resection following elective caesarean section at 35 weeks gestation would be preferable, assuming she remained clinically stable. Following an uncomplicated C-section at 35 weeks, a pulmonary CT scan demonstrated a large endobronchial lesion within the left main bronchus extending into the distal trachea and measuring approximately 24 × 12 × 20 mm (Fig. 1C, D). No significant collapse or consolidation of the left upper or lower lobe was seen, and there was no mediastinal or hilar lymphadenopathy. Local extension of the tumour to the oesophagus was excluded with gastroendoscopy. At two weeks post-partum, the patient underwent an elective thoracic resection of the tumour. The procedure was performed via sternotomy with the use of extra-corporeal circulatory support. Resection of the carina plus a significant resection of the left main bronchus was required in order to macroscopically clear the tumour (Fig. 1E). Reconstruction involved anastomosis of the right main bronchus to the distal trachea and of the left main bronchus to the left side of the bronchus intermedius (Fig. 1F). Both lung hila were fully mobilized to decrease tension on the anastomoses. In addition, intraoperative frozen section histological examination of the resection margins was performed. This was reported as showing clear margins. The patient made an uneventful recovery and was discharged home on day 7. Postoperative pulmonary CT scan demonstrated patent airways and no residual tumour.

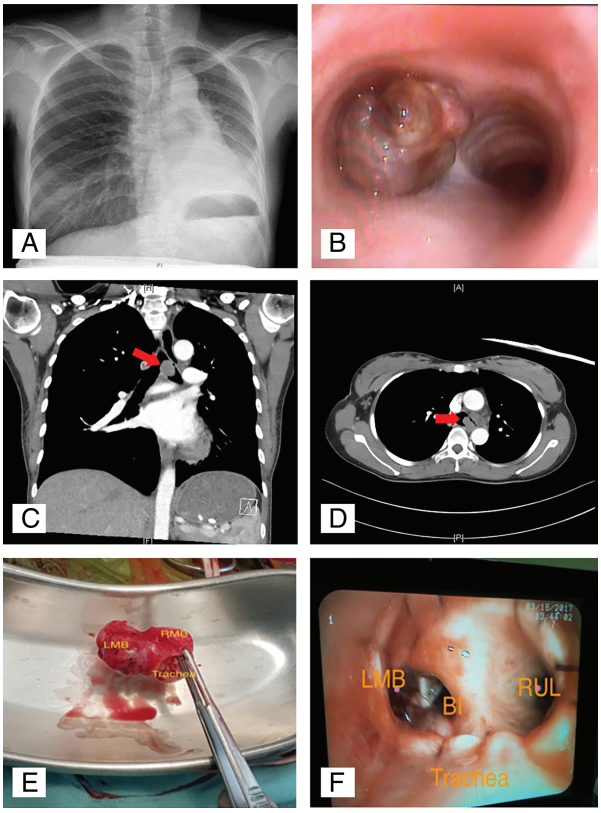

Figure 1. (A) Chest X-ray showed left lower lobe collapse and mediastinal shift towards the left. (B) bronchoscopy demonstrated a pearly, soft lesion obstructing the entrance of left main bronchus. (C, D) Computed tomography scan showed large endobronchial lesion within the left main bronchus extending into the distal trachea (arrows). (E) the resected specimen from the operation (LMB, left main bronchus; RMB, right main bronchus). (F) Broncoscopic view post-carinectomy and bronchial anastomosis (BI, bronchus intermedius; RUL, right upper lobe).

Despite intraoperative frozen section showing clear margins, formal postoperative histology reported positive margins at the trachea and the left main bronchus. Consensus was reached by multidisciplinary team discussion for adjuvant radiotherapy, and the patient received 60 Grey in 30 fractions. Repeat bronchoscopy following radiotherapy demonstrated healthy anastomoses and no evidence of recurrence. Three-monthly clinical reviews with yearly CT scan have been recommended as long-term follow up, complemented by bronchoscopic examination when required.

Discussion

Tracheal tumours, whether benign or malignant, commonly present with obstructive symptoms that may be life-threatening. Relief of obstruction to maintain airway patency is the cornerstone of treatment, with the decision to partly or completely resect dependent on the type and stage of tumour as well as patient performance status and preference [9]. A medical literature search identified 13 cases of primary tracheal tumours during pregnancy,

Table 1. Primary tracheal tumour diagnosed during the pregnancy.

References	Diagnosis	Presentation	Size and site	Treatment
Yamamoto et al. [10]	Malignant tracheal carcinoid	29 weeks pregnant with massive haemoptysis and desaturating	Obstructed 80% of the distal portion of the trachea	Tracheal resection through median sternotomy post-emergency CS
Sanchis Mínguez et al. [11]	Malignant tracheal carcinoid	29 weeks pregnant with acute respiratory insufficiency and massive haemoptysis	Obstructed the distal portion of the trachea	Emergency CS to avoid hypoxic foetal damage
Rajaratnam et al. [12]	Malignant tracheal Kaposi's sarcoma	Severe airway obstruction	N/A	Resection of the mass from the trachea via tracheoscopy
Schmitt et al. [13]	Malignant tracheal adenoid cystic carcinoma metastatic to the placenta	10 weeks pregnant with rapid regrowth of the tumour (6 years prior had resection of the ACC via bronchoscopy + radiotherapy, 1 year prior had recurrence of the tumour requiring tracheostomy, resection of the mass + radiotherapy)	N/A	Pharyngolaringectomy with resection of the first portion of the trachea but developed cervical metastasis CS at 33 weeks gestation Patient died post-partum of respiratory failure
Abike et al. [14]	Malignant tracheal adenocystic carcinoma	28 weeks pregnant with severe respiratory distress causing cardiopulmonary arrest	Obstructed 90% of the tracheal lumen	Emergency thoracotomy + Local resection of the tumour at 28 weeks gestation CS at 39 weeks gestation Radiotherapy postpartum
Shafiee et al. (current case)	Malignant tracheal adenocystic carcinoma	26 weeks pregnant with chest discomfort, cough, and haemoptysis	Obstructed the left main bronchus extending into the distal trachea	CS at 39 weeks gestation Elective thoracic resection of the tumour via sternotomy post-partum Radiotherapy post-partum
Watanabe et al. [15]	Mucoepidermoid tracheal carcinoma	39 weeks pregnant with cough and wheezing	Polypoid lesion narrowing the trachea	Resection of the lesion via fibro-bronchoscopy post emergency CS
Kesrouani et al. [16]	Mucoepidermoid Tracheal Carcinoma	27 weeks pregnant with haemoptysis and severe dyspnoea	Obstructed 90% of the tracheal lumen	Resection of the lesion via bronchoscopy at 27 weeks of gestation + Coagulated of the tumour bed with Argon-Plasma Coagulation (APC) Delivery at 39/40
Dieter et al. [17]	Mucoepidermoid tracheal adenoma	36 weeks pregnant with progressive	Endobronchial tumour above the carina	Right posterior lateral thoracotomy with

Table 1. Continued

References	Diagnosis	Presentation	Size and site	Treatment
		"asthma" and wheezing		resection of the tracheal tumour post emergency CS
Ipakchi et al. [18]	Tracheal granular cell tumour	16 weeks pregnant with shortness of breath	Narrowed 50% of the tracheal lumen by	Resection of the tumour inside pregnancy
Benisch et al. [19]	Tracheal granular cell myoblastoma	First-trimester pregnant with orthopnoea and paroxysmal nocturnal dyspnoea	Obstructed 90% of the proximal trachea	Tracheotomy via bronchoscopy followed by tracheostomy after one month
Djurhuus et al. [20]	Tracheal paraganglioma	Respiratory impairment and haemoptysis	N/A	Resection of the tumour
Li et al. [21]	Tracheal inflammatory myofibroblastic tumour	34 weeks pregnant with wheezing and haemoptysis	Obstructed 85% of the trachea	Electrocautery snare resection of the tumour via bronchoscopy post emergency CS
Amir et al. [22]	Tracheal inflammatory myofibroblastic tumour	Six weeks pregnant with dyspnoea on exertion and an acute stridor	Obstructed 80% of the subglottic tracheal lumen	Emergency tracheostomy at a level inferior to the mass. Resection of the tumour via subglottiscopy

CS, caesarean section; N/A, not available.

5 cases of which were malignant. Table 1 summarizes the characteristics of these tumours, their clinical manifestations, and adopted treatment plans.

Malignancy in pregnancy is often diagnosed in advanced stages. This can be partly related to the delayed diagnosis due to overlapping symptoms (e.g. shortness of breath, chest or abdominal discomfort, nausea, and vomiting) or blood test derangement during normal pregnancy. In addition, avoidance of diagnostic investigations or operations to minimize the foetal and maternal risks (e.g. exposure to radiation or risk of anaesthesia) may lead to late diagnosis [10]. It is essential for all clinicians providing care for pregnant women to be aware of, and to investigate judiciously, suspicious symptoms of cancer. In all cases of confirmed maternal cancer, placentae should be examined histopathologically post-partum to exclude metastases. If there is evidence of chorionic villous invasion, further investigation of the infant maybe necessary to identify any metastasis [23].

Management of cancers during pregnancy involves the complex balance of maternal and foetal benefits and risks. Expediting the birth depends on maternal condition and gestational age. Surgical intervention can be postponed to the post-partum period, only when the delay does not lead to worsening outcomes. Radiation therapy is relatively contraindicated in pregnancy as the high radiation exposure can cause congenital malformation, foetal death, and risk of malignancy in foetus [24]. Gestational age and type of chemotherapy (e.g. agent, dosage, single-agent vs. multi-agent therapy) should be taken into account when considering administration during pregnancy. Chemotherapy is associated with an increased risk of foetal death and major birth malformations during the first trimester. While it does not seem to increase the incidence of teratogenesis during the second and the third trimesters, there is a substantially higher risk of prematurity, low birthweight, and stillbirth [23,25]. Nevertheless, it is recommended to delay delivery for two to three weeks following exposure to cytotoxic agents for better recovery of foetal myelosuppression caused by maternal chemotherapy [25].

Conclusion

Adenoid cystic carcinoma of the tracheobronchial tree is a rare type of respiratory malignancy. A skilled multidisciplinary team should be involved in assessing the best treatment approach for any type of malignancy during pregnancy, balancing maternal risks and foetal safety. Thorough explanations to the patient and her family regarding the pertinent issues play an important role in providing optimal care.

Disclosure Statement

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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