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CASE REPORT

A rare cause of recurrent wheeze and seizures

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SUMMARY

A 34-year-old woman presented with recurrent attacks of breathlessness and seizures. The patient's condition worsened during the course of her hospital stay, as a result of which she had to be intubated twice. Radiological studies showed a mass in the trachea and the subsequent biopsy of the mass revealed an infiltrating carcinoma with morphological features of adenoid cystic carcinoma.

BACKGROUND

Neoplasms of the trachea with the trachea as primary origin are extremely rare, they occur in every 2.6 per one million people per year.[1] Malignant tumours comprise of 86% of all primary tracheal tumours, the most common of which are squamous cell carcinomas, followed by adenoid cystic carcinomas. Anatomically, adenoid cystic carcinomas originate from the submucosal glands that are embedded in the epithelial lining of the trachea.[2] Conventionally, adenoid cystic carcinomas are slow growing tumours that metastasise haematogenously and by direct extension through the submucosa. Metastatic spread to the lymph nodes is uncommon.[3] Surgery followed by radiation has shown to have the best outcome. Early detection of the disease, followed by surgical resection of the tumour with regular follow-ups is imperative for securing a good prognosis.[4]

CASE PRESENTATION

A 34-year-old woman presented to the emergency department, with a 1- day history of shortness of breath, fever and two episodes of unresponsiveness. She reported having a 2-month history of episodic, sudden shortness of breath at rest, along with the twitching of facial muscles, jerking of the limbs and up rolling of the eyes, followed by loss of consciousness. She also reported faecal and urine incontinence during these episodes. She was placed on non-invasive ventilation due to hypoxaemia, and was treated as having a case of acute asthma exacerbation with salbutamol and ipratropium bromide nebulisation, along with fluticasone and salmeterol inhaler. She was given lamotrigine, phenytoin and valproic acid for her seizures. She was treated with clarithromycin and ceftriaxone for a possible infection. The antibiotics were stopped when negative blood and urine cultures were obtained.

The patient was diagnosed with asthma a year prior to her presentation, based on spirometry, which showed a forced vital capacity (FVC) of 63%, forced expiratory volume in 1 s (FEV1) of 43% and FEV1/FVC ratio of 67%. Her condition was being monitored by a general practitioner in her village. Two months prior to admission, she had reported having at least 15 episodes of fits, each of which was followed by asthma exacerbation. The patient's EEG and MRI were normal; she was prescribed lamotrigine, phenytoin and valproic acid to control her seizures. For her asthma, she was given intravenous methylprednisone, albuterol inhaler, fluticasone and montelukast; however, her condition did not improve.

A physical examination revealed a young woman in no respiratory distress. She was alert and responsive. Vital signs revealed a body mass index of 27.6 kg/m^2, blood pressure 170/110 mm Hg, pulse 144 bpm, respiratory rate 36/min, O_2 saturation of 98% and temperature 37.6°C. Chest examination revealed bilateral harsh vesicular breath sounds with an audible wheeze. Motor examination revealed a power of 3/5 in all four limbs. Mild bilateral pitting oedema was noted in both legs.

INVESTIGATIONS

Complete blood cell count showed a white cell count of 16.9 with neutrophils of 91.7%. Arterial blood gas showed pH of 7.34, pCO_2 of 74.10, pO_2 of 213.30 (on supplemental oxygen), bicarbonate of 40.60 and oxygen saturation of 99.70% (on bi-level positive airway pressure). The patient's blood and urine cultures were both negative. Chest X-ray showed no significant abnormality (figure 1).

DIFFERENTIAL DIAGNOSIS

A diagnosis of acute asthma exacerbation, hypercarbic respiratory failure and complex partial seizures

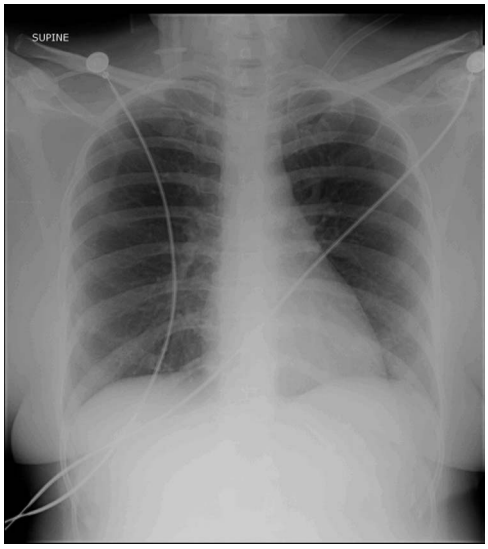

Figure 1 Normal chest X-ray, no significant abnormality is seen.

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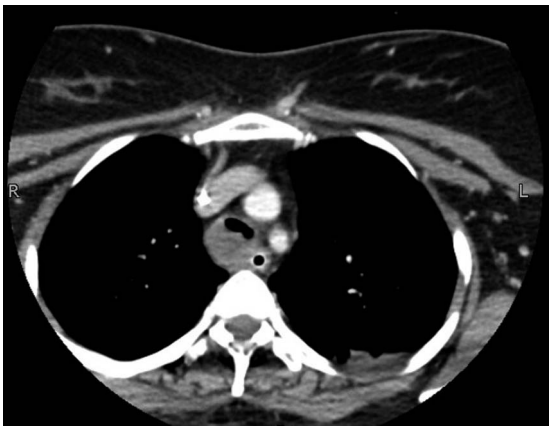

Figure 2 Soft tissue mass, 35×19 mm, located 15 mm above the carina. The lesion appears closely adherent to the oesophagus and prevertebral tissues.

was made. The patient was treated with a combination of steroids, bronchodilators and antibiotics, along with non-invasive ventilation. She was weaned off the non-invasive ventilation and did well for a day. The next day, she developed respiratory distress, hypoxia and a low Glasgow Coma Scale. The arterial blood gases showed pH of 7.12, pCO_2 of 51.9, pO_2 of 126, bicarbonate of 41.3 and oxygen saturation of 69%. She was intubated and shifted to the intensive care unit. The Neurology consult team advised optimisation of the antiepileptics. She remained stable on the ventilator, and hence she was re-extubated the next day. She was re-intubated, secondary to hypercarbia, within 2 h of extubation.

Contrast-enhanced CT of the chest and neck showed an infiltrating oval-shaped soft tissue mass arising from the right lateral wall of the trachea, which was causing significant narrowing of the tracheal lumen; no evidence of metastatic disease was seen (figure 2).

Bronchoscopy revealed a mass impinging on the trachea and its surrounding structures.

TREATMENT

The patient underwent a right posterior lateral thoracotomy, with tracheal mass biopsy and tracheostomy. The operative findings confirmed a large firm mass, adherent to the trachea externally, with significant impingement. The mass was deemed to be non-resectable. Histopathology revealed an infiltrating carcinoma with the morphological features of adenoid cystic carcinoma (figure 3). Two mediastinal lymph nodes were also found to be malignant.

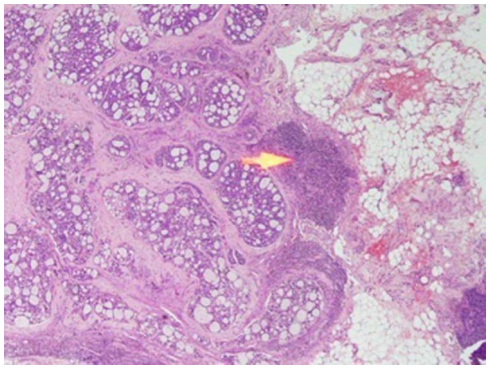

Figure 3 The biopsy revealed fibro-collagenous tissue with a neoplastic lesion composed of cords and clusters of neoplastic cells. Neoplastic cells are seen with a round to ovoid nucleus with hyperchromatic nuclei and eosinophilic cytoplasm. The adjacent stroma shows fibrosis.

OUTCOME AND FOLLOW-UP

The patient was treated with palliative radiation therapy to the mediastinum. At 6-month follow-up, the patient reported to be doing well.

DISCUSSION

Tracheal tumours comprise of 1–2% of all lung masses, and hence have a low incidence.[5] These tumours may either be benign or malignant in nature. Benign tracheal tumours such as lipomas, hamartomas and neurolemmomas are rare. However, malignant tumours such as squamous cell carcinomas and adenoid cystic carcinomas comprise 80% and 16.3% of all tracheal tumours in adults, respectively.[6] These tumours usually present with asthma-like symptoms—due to the impinging of the tumour on the surrounding organs.

An adenoid cystic carcinoma is a rare tumour of unknown aetiology. The tumour has a predilection to grow on several different sites of the body, such as the head and neck region, the breast and the pelvis. Previously, this tumour of the tracheobronchial tree variety was referred to as a cylindroma, due to the cylindrical shape of epithelial cells of mucous glands.[7] It occurs most commonly in the fifth and sixth decades of life, and is not predisposed to a particular gender. Symptomatically, the tumour closely resembles asthma—with cough and dyspnoea being prominent manifestations of the disease.[8] One of the atypical symptoms in our case were recurrent seizures. We found only one study that reported a 13% incidence of seizures in patients with cancer, mainly due to intracranial metastasis, strokes, anticancer medication or metabolic disturbances.[9] However, the patient's brain imaging studies were clear and serum electrolytes were normal, hence ruling out a metabolic disturbance. Postresection the tumour has a high rate of local recurrence, most likely due to the perineural invasion of the tumour.[10] Metastasis most commonly occurs to the bones and lungs, and is usually characteristic of a late stage of disease.

Surgical resection and reconstruction remain the mainstay of treatment. Radiotherapy may be offered as a form of palliative therapy, or it may be offered postoperatively in cases where the tumour is not resectable, or in cases where metastasis of the disease has occurred.[11] Chemotherapy with carboplatinum and paclitaxel, in conjunction with radiotherapy, has shown to be successful in some trials.[12]

Learning points

- A high index of suspicion is required to diagnose tracheal tumours.
- Tracheal tumours must be considered as an alternate diagnosis in cases where a patient does not respond to asthma treatment.
- Not all 'wheezes' are asthma.

Contributors MH was responsible for research on the topic of the case report, and was involved in drafting and revision of the final article. SQ was responsible was

looking into the patient's history and case presentation, and was involved in drafting and revision of the article. ABSZ was involved in drafting and revision of the article and is the guarantor.

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