CASE REPORT

Intratracheal inflammatory myofibroblastic tumour mimicking severe acute asthma

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SUMMARY

A 3-year-old boy presented with severe airway obstruction which was diagnosed as asthma. He improved but had repeated episodes of severe airway obstruction. On clinical examination, he had a tracheal cough and monophonic wheezing. Imaging revealed a large lesion in the distal part of the trachea which was confirmed by bronchoscopy. The lesion was surgically removed. Histology revealed features characteristic of an inflammatory myofibroblastic tumour. Following the resection there is no recurrence of the lesion.

BACKGROUND

Severe asthma not responding to treatment is a relatively common clinical problem. This rare cause of large airway obstruction in children highlights the importance of carefully evaluating children who present with lower airway obstruction mimicking severe asthma with episodes of relapse. This case highlights the importance of identifying clinical signs which are more in keeping with fixed large airway obstruction which do not respond to a β_2 -agonist.

CASE PRESENTATION

A 3-year-old boy presented with the clinical picture of severe lower airway obstruction which was diagnosed as a life-threatening attack of asthma requiring admission to an intensive care unit (ICU). This was his first attack of airway obstruction. On examination, he had severe respiratory distress with chest wall indrawing. He had a prominent brassy cough, and on auscultation, bilateral monophonic wheezing with bilateral crepitations was audible. He was treated with an inhaled B2 agonist, oral corticosteroids and intravenous magnesium sulfate. He responded to initial treatment and was discharged from the ICU. He was continued on asthma maintenance therapy, but 5 days later he had another severe attack requiring re-admission to ICU.

INVESTIGATIONS

Biochemical and haematological investigations were normal. Rhinovirus was cultured from nasopharyngeal aspirates. Chest X-ray showed hyperinflated lung fields with perihilar streakiness. Chest CT scan demonstrated a large intraluminal lesion in the trachea at the level of the carina. The axial views gave the impression that this lesion had grown out of the tracheal wall, and was obstructing at least 75% of the tracheal lumen (figure 1). There were no other airway or intraparenchymal lesions visible.

Figure 1 Coronal chest CT scan reconstruction demonstrating a lesion in the trachea located just above the carina. The lesion appears to adhere to the right wall of the trachea.

On chest CT scan complicating small pneumomediastinum was also present (figure 2). Flexible bronchoscopy, under general anaesthesia with a 4.0 mm video bronchoscope confirmed a large pedunculated tumour arising from the posterior wall of the distal trachea obstruction approximately 80% of the tracheal lumen (figure 3). The distal part of the tumour obstructed the opening to the right main bronchus.

DIFFERENTIAL DIAGNOSIS

Most cases of children presenting with severe acute asthma respond to medical treatment. A small group of children will require ventilation during the acute exacerbation. Children not responding to

Figure 2 Chest CT scan demonstrating that lesion has infiltrated the wall of the trachea.

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Figure 3 Postresection: large tumour nearly filling the whole lumen of the airway.

treatment are more likely to have anatomical abnormalities of the large airways. External compression of trachea is commonly caused by duplication cysts of the foregut or vascular rings. Tracheal stenosis presents mostly early in life but rarely can have a delayed presentation. Large airway obstruction in children older than 1 year is mostly caused by the inhalation of foreign bodies. The symptoms in these children are mostly persistent and do not respond to bronchodilators and corticosteroids. Acquired tracheal obstruction is rare in children. In areas with a high burden of tuberculosis, intraluminal obstruction or external compression of the trachea must be considered. Intraluminal tracheal tumours are very rare.

TREATMENT

By a right-sided thoracotomy the tumour which stretched across three cartilage rings was exposed. Ventilation was maintained by endotracheal intubation of the left main bronchus and the tumour completely resected.

OUTCOME AND FOLLOW-UP

Histological examination confirmed an inflammatory myofibroblastic (IF) tumour. The histological appearance was that of a polypoid tumour arising from the posterior wall of the trachea and was composed of fascicles and a storiform pattern of bland spindle phased cells (figure 4). The resection border was tumour free. The patient was discharged home after 5 days. A follow-up bronchoscopy carried out 6 weeks, 6 and 12 months after the surgery demonstrated no tumour recurrence.

Figure 4 Histology slide demonstrating proliferation of spindle cells in fascicles and a variable mixture of lymphocytic and plasma cells diagnostic of an inflammatory myofibroblastic tumour.

DISCUSSION

In children acute severe asthma exacerbations is a common reason for emergency department visits, yet only a small percentage (15%) of hospital admissions will require admission to a paediatric ICU. Of these patients, 8–33% will require mechanical ventilation.^{1–4} Most children will respond to medical treatment for their severe obstruction. We report this rare case of IF tumours in a child presenting with symptoms and signs of suggestive of life-threatening asthma, who initially partially responded to treatment. On initial presentation, the importance of the brassy cough and monophonic wheeze indicative of large airway obstruction requiring further investigation were not recognised (figure 5).

IF tumours are benign tumours of the respiratory system, accounting for 0.04–1% of all primary lung neoplasms. IF tumours were first described by Brunn in 1939.^{5–12} The lung parenchyma is the most commonly affected site,^{5–12} with only a few published reports relating to tracheal involvement.¹³ A report of 15 cases of IF tumours, of which four cases with tracheal involvement, described 52.3% of cases involving lung parenchyma and 46.7% the airways.¹⁴ IF tumours are rare in children with Bumber *et al*¹⁵ reporting eight cases in children under the 16 years of age.

IF tumour has been referred to by various names including: inflammatory pseudotumour, pseudosarcomatous myofibroblastic proliferation, plasma cell granuloma, xanthomatous pseudotumour, inflammatory myofibrohistiocytic proliferation, inflammatory fibrosarcoma and recently, IF tumour.⁹

Most children with IF tumours are asymptomatic, but common symptoms include fever, cough, pain, haemoptysis and weight loss.¹⁶ Recently Dhouib *et al*¹⁷ have described a case of atelectasis of a lobe due to an endobronchial lesion.

Treatment of choice is complete resection with a rim that is completely tumour free, to reduce the risk of recurrence. The prognosis is good if the tumour is completely resected and recurrences rates are very low.^{12 18} IF tumours confined to the lung have a recurrence rate of only 1.3%. Tumours extending beyond the confines of a single organ at the initial presentation, however, have a high chance of recurrence despite what appears to be adequate resection. These children require frequent post-operative evaluation to detect recurrence at an early stage.

In infants and children with airway obstruction, not or inadequately responding to treatment, a cause other than asthma must be considered. A barking cough, monophonic wheezing audible in inspiration and expiration, decreased unilateral ventilation are clinical clues that you are dealing with large airway obstruction requiring further investigation. A clinician's suspicion is further raised if the wheezing is of sudden or recent

Figure 5 Bronchoscopy pictures: (A) preoperatively demonstrating large intraluminal tumour in distal trachea adhering on the right to the lateral tracheal wall; obstructing most of the airway lumen. (B) Six months postresection; no signs of recurrence.

onset in a previous asymptomatic child. The most common cause of this clinical scenario is foreign body aspiration. Other possibilities include acquire tracheal stenosis following endotracheal intubation, lymph node compression of the airways due to tuberculosis and lymphoma, congenital abnormalities of the foregut or malignancies of the lung or large airways. The initial investigation is a chest radiograph, where the radiological picture will lead to the choice of either a chest CT scan or flexible bronchoscopy as the next level of investigation. This approach will rapidly lead to an accurate diagnosis and correct therapy.

CONCLUSION

We describe a young boy with a large IF tumour in the distal trachea, which mimicked an attack of acute severe asthma. The clinical findings were those of large airway obstruction and presenting with bilaterally monophonic wheezing due to the close proximity of the tumour to the carina. Although rare, children who do not respond to treatment for acute asthma should be evaluated for other causes of airway obstruction, including large airway pathology, even if the symptoms are bilateral.

Learning points

- ▶ In children with an atypical presentation or response to treatment for asthma, intraluminal airway obstruction should be considered subsequently.
- ▶ A brassy cough, monophonic wheezing and unilateral ventilation are clinical clues of an anatomical cause of airway obstruction.
- ▶ Bronchoscopy is necessary in severe acute asthma. If the patient does not respond to treatment, exclude intraluminal obstruction.
- ▶ If intratracheal lesions are present, an inflammatory myofibroblastic (IF) tumour must be considered.
- ▶ Surgical resection of IF tumours is required

Contributors P G is the main author. PG was involved in the diagnosis and management of the patient, which included conducting the bronchoscopy and the follow-up of the patient. PG and RG were responsible for writing the paper. R G played a major role in the management of the patient, since he was present during

the different time period of the child's management and was the leading decision maker. J J was the leading surgeon and was responsible for the surgery; he was present during the follow-up bronchoscopy. P S was the pathologist who made the histological diagnosis of this patient.

Competing interests None.

Patient consent Obtained.

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