CASE REPORT

Young man with dyspnoea for 6 months; presenting with subacute tracheal obstruction due to leiomyoma

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SUMMARY

Tracheal leiomyoma is a rare benign tumour with less than a 100 cases reported in the literature. We report a case of leiomyoma initially presenting and being treated as asthma until it lead to life-threatening tracheal obstruction, requiring immediate surgical intervention.

BACKGROUND

Primary tracheal tumours occur rarely and <20% are benign. Tracheal leiomyoma constitute only 1% of the primary tracheal tumours.^{1 2} About 40 cases have been reported in the literature. The majority presents with long-standing symptoms; most commonly dyspnoea. In others, it may emerge as an incidental finding on an imaging study. We report a case that presented with stridor requiring immediate surgical intervention.

CASE PRESENTATION

A 37-year-old man presented in the emergency room with progressive dyspnoea and cough for 6 months. His symptoms had worsened in the last 3 days. He had no known prior comorbid or a surgical history. He reported scanty amount of haemoptysis, usually streaks of fresh blood mixed with saliva when he had episodes of violent cough, which he attributed to throat irritation. Occasionally he would also notice an audible wheeze in the morning. He resided in a rural area without advance medical facilities but denied any history of exposure to animals. The only other people in his house were his wife and two children, all of whom were healthy. He had visited a local physician when his symptoms became bothersome and was being treated for asthma for the last 4 months. He was initially prescribed bronchodilators and then took two short courses of oral steroids (prednisolone) lasting <10 days each, but his shortness of breath persisted. His serial chest X-rays never showed any infiltrates.

On examination in the emergency, he was afebrile, had a pulse of 98 bpm, a blood pressure of 118/74 mm Hg and respiratory rate of 29 breaths/min. He had a stridor with maximum severity heard at the base of the neck, the rest of the examination being normal. He was maintaining an oxygen saturation of 95% on room air.

INVESTIGATIONS

A chest X-ray was carried out which was normal. However, due to the presence of stridor and a

possibility of airway obstruction, an urgent CT scan was performed, while the patient awaited an ear, nose and throat consultation (which was later cancelled). The CT scan showed a rounded polypoid lesion partially obstructing the trachea at the D2 level (figure 1A, B).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis was

- Carcinoid (neuroendocrine tumour), due to history of asthma, haemoptysis and a polypoid mass.
- Endobronchial tuberculosis; Pakistan being an endemic area and a history of haemoptysis.

TREATMENT

He was admitted urgently for bronchoscopy, followed by resection of the tumour and tracheal repair. Under general anaesthesia, the patient was intubated without any complications. A transverse incision was given at the middle of the neck and the platysma was divided followed by the isthmus. The trachea was dissected after which an intraoperative fibre-optic bronchoscopy was performed, showing a well-defined rounded mass at the level of thoracic inlet which was adherent to the posterolateral wall. Trachea was then opened and the mass identified, aided by the bronchoscope. A second reinforced endotracheal tube (a flexible type of endotracheal tube) was passed through the trachea, and the mass was excised along its margins from the posterolateral wall of the trachea. The trachea was then repaired and the wound closed in layers with a drain placed.

OUTCOME AND FOLLOW-UP

The resected polypoidal mass measured 3×2×2 cm. On gross examination and sectioning, it appeared as a pale pink fleshy tumour. Microscopic examination from the attached tracheal margins revealed fragments of fibrocollagenous tissue partly lined by stratified squamous epithelium. The underlying stroma revealed mucus glands arranged in clusters. The hyaline cartilage was also seen with no evidence of malignancy. Sections examined from the tracheal mass revealed a neoplastic lesion composed of spindle cells with elongated nuclei and pale cytoplasm (figure 2A). The nuclei revealed mild-to-moderate nuclear atypia and 4 mitosis per 10×HPF. The neoplastic cells were arranged in interlacing and whorling pattern. No area of necrosis was seen.

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Figure 1 (A) Transverse section of CT scan of the chest showing a rounded polypoid mass causing partial obstruction of the trachea. (B) Longitudinal section through the same CT scan showing the mass in the trachea.

Immunohistochemistry showed reactivity with α smooth muscle actin (ASMA) (figure 2B) while S-100 and desmin were negative. On the basis of all these features, it was diagnosed to be a leiomyoma. The drain and stitches were removed 2 weeks after surgery. He reported no dyspnoea or haemoptysis. He has been followed up since 2 years without recurrence.

DISCUSSION

Tracheal leiomyoma is a rare neoplasm and accounts for <1% of all tracheal tumours. They occur more commonly in men unlike pulmonary parenchymal and endobronchial leiomyoma which are more common in women. Rarely, they occur at the carina or even in an accessory bronchus. They originate from the smooth muscle cells, from the membranous part of the trachea and usually grow as polypoidal masses frequently obstructing the trachea partially or even completely.²

The patient usually presents with dyspnoea, wheezing, haemoptysis or weight loss.^{3 4} In few, it may present as an incidental finding on an imaging study. Owing to the rarity of these lesions, they are frequently misdiagnosed and treated initially as asthma.

They are usually missed on a chest X-ray (anteroposterior or posteroanterior views). Tracheal tumours that lie in the neck region may be seen, however, if they lie 'too low' in the neck (such

Figure 2 (A) Sections examined from the tracheal mass revealing a neoplastic lesion composed of spindle cells with elongated nuclei and pale cytoplasm. The nuclei reveal mild-to-moderate nuclear atypia. The neoplastic cells are arranged in interlacing and whorling pattern. (B) Immunohistochemistry showing reactivity with α smooth muscle actin.

as the thoracic inlet in our patient), they may not be visualised unless a lateral view chest X-ray is obtained.^{5 6} This was not performed due to the emergent nature of this case and a CT scan was preferred as the next imaging modality. CT scan of the chest is the best modality to detect the lesion, its location, size and the extent of invasion. Leiomyoma presents as a homogeneous nodule on unenhanced CT scans and enhances on contrast.^{7 8} Bronchoscopy can visualise the mass in the tracheobronchial tree.

Although benign but owing to its potential life-threatening complication of airway obstruction, its complete resection is the mainstay of treatment. There have been a few published case series demonstrating local resection via bronchoscopy using YAG-laser, electrocautery and cryotherapy.^{9 10} Surgical approaches that have been used frequently involve segmental sleeve resection and end-to-end anastomosis, carinal resection and even thoracotomy according to the location and size of the mass.^{11 12} In this patient endoscopic resection was not performed due to the risk of respiratory compromise, as the mass was occupying more than two-thirds of the diameter of the trachea. Moreover, the location of the tumour at the thoracic inlet made the intervention via the neck a safer and easier surgical approach.

Histopathology, such as leiomyoma from other organ systems, shows spindle-shaped smooth muscle fibres.⁵ Leiomyomas

usually are positive for immunohistochemical stains ASMA and desmin. In our case it was negative for desmin, however, large-scale studies on immunohistochemistry patterns in leiomyomas do report desmin negative cases¹³ and ASMA as a superior marker for smooth muscle tumours.^{14 15} Long-term prognosis is good. There is one published case report of recurrence.

Learning points

- Dyspnoea and wheezing not responding to conventional therapies for asthma should be investigated for other causes.
- Respiratory symptoms persisting for more than 2 weeks should be investigated with advance radiographic techniques; a CT scan, if possible. In our patient, the tumour was missed on all chest X-rays as it is not a good diagnostic modality for tracheal stenosis or tumours.
- Any suspicion of upper airway or tracheal obstruction needs prompt management.

Contributors FI took initiative in writing the entire article, searching the references and putting down all the information together. Also, involved in the management of this patient as a resident and obtained the history, consent and imaging details. SHF provided all details for the surgical intervention, reviewed and co-wrote the manuscript. SP provided the histopathology details, co-wrote and reviewed the manuscript.

Competing interests None declared.

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