CASE REPORT

An unusual cause of stridor in an adult: mediastinal foregut duplication cyst

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SUMMARY

We present a case of unusual presentation of stridor in an adult man who was identified to have a sudden expansion of a foregut duplication cyst in the mediastinum. This resulted in superior vena caval obstruction and subsequent airway compromise. Following resuscitation, the cyst was surgically removed via a thoracotomy which confirmed the diagnosis and provided definitive management. Foregut duplication cysts are an unusual cause of stridor in adults, with only five cases ever documented worldwide in the literature, presenting in such a manner. This case is a reminder of the importance of thorough clinical examination, the benefits of radiology as an indispensable adjunct and essential role of the multidisciplinary team.

BACKGROUND

Foregut duplication cyst is an extremely unusual differential diagnosis of stridor in an adult. Our case is a timely reminder of the importance of a thorough clinical examination reinforced with appropriate imaging to ensure optimal multidisciplinary management.

CASE PRESENTATION

A healthy 59-year-old man attended his local accident and emergency (A&E) department with a 2-day history of the coryza. He also experienced a 12 h history of chest pain radiating to his back, preceded by 'snap' sensation in his chest. Initial clinical examination was unremarkable, however, a chest radiograph which showed a widened mediastinum in keeping with a mediastinal mass.

Emergency CT imaging to assess the integrity of the aorta showed a large superior mediastinal mass displacing the oesophagus, compressing the trachea and intimately related to both carina and trachea (figures 1 and 2). The mass had a cystic nature which is a feature of a developmental abnormality.

Following discussion with the cardiothoracic team, he was discharged home from A&E department to await outcome of the planned multidisciplinary discussion the following morning.

Shortly after discharge he re-presented with stridor, hoarseness, dysphagia and increasing respiratory distress. He had a newly observed tender rapidly increasing right-sided neck swelling, engorged neck veins and dyspnoea, suggestive of superior vena cava (SVC) obstruction.

He clinically deteriorated rapidly with sudden respiratory decompensation and decreased Glasgow Coma Scale. He was intubated in the A&E

Figure 1 Transverse CT of the superior mediastinum: a large mass containing two visible cysts, compressing and distorting the path of the trachea.

resuscitation room to secure his airway. Repeat imaging showed a soft tissue mass in the retropharyngeal space continuous with the superior mediastinal mass and causing functional SVC obstruction (figures 3 and 4).

Figure 2 Coronal CT of the thorax and abdomen: the large mass intimately related to the oesophagus, trachea and carina.

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Figure 3 Sagittal CT of the thorax and abdomen: the mass's position in the retropharyngeal space continuous with the superior mediastinum.

DIFFERENTIAL DIAGNOSIS

Retropharyngeal abscess rupture into the mediastinum, lymphoma, sarcoma and haemorrhagic transformation of the mass.

TREATMENT

Following emergency intubation his initial management included transfer to theatre where the airway was secured with a surgical tracheostomy by the Ear Nose & Throat team. A multidisciplinary discussion determined that tracheostomy would provide safer management of his airway as the small size of endotracheal tube in place following his emergency intubation would cause difficulties in managing the airway for a thoracic procedure and postoperatively. Insertion of the tracheostomy was technically challenging due to the engorged, oedematous tissues secondary to SVC obstruction. On examination the retropharyngeal swelling was also undertaken and demonstrated grossly oedematous tissue but no evidence of retropharyngeal abscess. An intraoperative flexible endoscopy through the tracheostomy showed external compression to the trachea, confirming the CT findings. Concurrently a video-assisted thoracoscopic surgery was undertaken by the cardiothoracic surgeons, which confirmed a mediastinal cystic lesion with internal haemorrhage, necrotic tissue and pus. A day later the patient returned to theatre for a formal thoracotomy, mediastinal debridement

Figure 4 Coronal CT of the neck: the mass in the retropharyngeal space continuing into the superior mediastinum, and subsequent tissue oedema of the neck.

and resection of the mediastinal cyst and further exploration of the neck and placement of a prevertebral drain to decompress the oedematous tissue.

OUTCOME AND FOLLOW-UP

Following less than a week in intensive care unit he was discharged to the ward and subsequently home. Aside from post-operative scars, the only long-term sequela is a hoarse voice secondary to a medialised left vocal cord.

Histopathological analysis confirmed a cyst lined with ciliated pseudostratified respiratory-type epithelium, and a cyst wall constructed of fibroconnective tissue and smooth muscle bundles with a few bronchial-type glands within the wall. There was no cytological atypia, but associated haemorrhage and inflammation consistent with abscess formation. These histological findings are representative of an inflamed benign cyst, suggestive of a foregut duplication cyst, most likely of bronchial origin.

DISCUSSION

Foregut duplication cysts are uncommon, typically presenting in childhood due to compressive effects on the trachea and oesophagus; respiratory difficulties and dysphagia.¹ Unlike our case study, diagnosis in adulthood is usually incidental with over a third of patients asymptomatic at diagnosis.²

Radiological diagnosis can be challenging, with not all foregut duplication cysts visible on plain chest radiographs. In some cases, the secondary effects of the extrinsic compression from the cyst on bronchial and vascular structures may be the only radiological features.³ CT imaging is the most appropriate investigation allowing characterisation of shape, extent and

relationship with surrounding structures.³ However, despite this the variable position and consistency of the cyst make diagnosis challenging even with adjuncts of oesophagography, echocardiography, 99Tcm-pertechnetate scintigraphy and vascular imaging.³ Scintigraphy (99Tcm-pertechnetate) is used to evaluate the anatomical position of ectopic gastric mucosa for example in meckel's diverticulum, and as up to a third of foregut duplication cysts are lined with gastric mucosa it can add useful diagnostic information, particularly in cases where synchronous multiple duplications are suspected.⁴

Learning points

- ▶ Foregut duplication cysts are uncommon in adults and can present as airway emergencies.
- ▶ If clinical signs of superior vena caval obstruction are present mediastinal pathology needs to be investigated.
- ▶ In complex patients a multidisciplinary approach is essential to optimise patient outcomes.
- ▶ Radiological investigations are an essential adjunct in management of complex head and neck cases, but should not preclude definitive management.

Our literature search identified only five reported cases of airway compromise secondary to mediastinal foregut duplication cyst in over 50 years. Although very few details of the management of these patients is available, the literature agrees surgical removal is mandatory, and should ideally be elective and thoracoscopic in adults as well as children, due to the reduced morbidity in comparison to open thoracotomy.^{2 5}

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