Tracheo-oesophageal fistula in a patient with chronic sarcoidosis

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ABSTRACT

Sarcoidosis is a common multisystem granulomatous condition of unknown aetiology, predominantly involving the respiratory system. Tracheal stenosis has been described but we believe that we present the first case of a tracheo-oesophageal fistula secondary to chronic sarcoidosis. A 57-year-old woman with sarcoidosis, a known tracheal stricture and a Polyflex[®] stent in situ presented with stridor. Bronchoscopy confirmed in-stent stenosis, by exuberant granulation tissue. The stent was removed and the granulation tissue was resected accordingly.

Postoperatively, the patient was noticed to have an incessant cough and video fluoroscopy raised the suspicion of a tracheo-oesophageal fistula. A repeat bronchoscopy demonstrated marked granulation tissue, accompanied by a fistulous connection within the oesophagus at the mid-lower [middle of the lower] third of the trachea. Three Polyflex[®] stents were sited across the entire length of the trachea.

Sarcoidosis presents with varying clinical manifestations and disease progression. Tracheal involvement appears to be a rare phenomenon and usually results in stenosis. To date, there has been little or no documented literature describing the formation of a tracheo-oesophageal fistula resulting from sarcoidosis. Early reports documented the presence of sarcoidosis induced weakening in the tracheal wall, a process termed tracheal dystonia. Weaknesses are more apparent in the membranous aspect of the trachea.

Despite the rare nature of such pathology, this case report highlights the need to consider the presence of a tracheo-oesophageal fistula in sarcoidosis patients presenting with repeat aspiration in the absence of an alternate pathology.

KEYWORDS

Tracheo-oesophageal fistula – Sarcoidosis – Sarcoid – Trachea – Fistula

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Sarcoidosis is a multisystem granulomatous condition affecting predominantly the respiratory system. Tracheal involvement of the disease resulting in stenosis has rarely been described. We believe this to be the first report of a tracheo-oesophageal fistula secondary to chronic sarcoidosis.

Case history

A 57-year-old woman with a previous tracheal stent in situ for tracheal stenosis presented with stridor secondary to in-stent restenosis. There was a marked reduction in her forced expiratory volume in one second (FEV1) from a baseline of 75 to 53% of predicted. Her past medical history included sarcoidosis, asthma, diabetes mellitus and depression. She was admitted on co-amoxiclav, flucloxacillin and clarithromycin for a presumed community acquired pneumonia and then transferred to our unit. She had also

been commenced on a prolonged course of prednisolone for her underlying sarcoidosis.

Initially, the patient underwent a bronchoscopy, which confirmed in-stent stenosis as well as a narrowed distal trachea. Tissue debris was cleared from inside the stent to fully restore patency. However, extubation failed and she was transferred intubated to the intensive care unit. A further bronchoscopy undertaken the following day demonstrated marked occlusion secondary to thickened secretions. Initial review by the otolaryngology team elicited normal vocal cord function, with no obvious paralysis. Despite this, a follow-up review showed the patient to be freely aspirating fluid. A subsequent video fluoroscopy demonstrated appearances that were consistent with the presence of a tracheo-oesophageal fistula (Fig 1).

The patient therefore underwent a further bronchoscopy, which, in addition to demonstrating marked granulation

Figure 1 Initial bronchoscopy demonstrating a tracheo-oesophageal fistula in the mid to lower third of the trachea

Figure 3 Postoperative bronchoscopy demonstrating a proximal Polyflex® stent in the trachea

tissue and significant secretions, confirmed the presence of a mature fistula within the oesophagus at the mid-lower [middle of the lower] third of the trachea, with surrounding granulomatous tissue. Multiple biopsies were obtained, and a total of three Polyflex (Boston Scientific, Natick, MA, US) stents were used across the entire length of the trachea (Figs 2 and 3).

The patient's postoperative recovery was unremarkable until three days after surgery, when she developed marked chest pain and hypoxia. Computed tomography pulmonary angiography demonstrated an extensive pulmonary embolism in the right pulmonary artery as well as bilateral basal pulmonary emboli. Unfortunately, despite the initiation of anticoagulation therapy, she was found collapsed two days later and repeated resuscitation attempts were deemed futile. The most likely cause of her death was cardiac arrest secondary to thromboembolic disease.

Figure 2 Postoperative bronchoscopy demonstrating a distal Polyflex® stent in the trachea at the level of the carina

Discussion

Sarcoidosis is a relatively common systemic granulomatous disease of unknown aetiology, which can present with varying clinical manifestations and disease progression. Although granulomas may occur in any organ or system, they primarily affect the respiratory tract in just over 90% of cases.¹ The disease can be characterised by mucosal erythema, granularity/cobblestoning, plaques, nodules, bronchiectasis, bronchiolitis, anatomical distortion and stenosis. In the instances of airway involvement, airflow limitation may be present and may manifest as acute airway obstruction. Involvement of the upper aerodigestive tract may lead to hoarseness, dysphonia and dysphagia.

Tracheal involvement of sarcoidosis appears to be a less than well understood phenomenon with varying levels of presentation. Although more commonly associated with bronchial stenosis, narrowing in the trachea secondary to sarcoid has been documented in the literature since the early 1980s.²⁻⁴ However, although documented in the literature, stenotic involvement of the large airways is an unusual complication of sarcoidosis.⁴ Such cases have been shown to respond variably to courses of oral corticosteroids and serial dilations.

To date, there has been no documented literature describing the presence of sarcoid induced tracheo-oesophageal fistulas. One probable explanation for a potential fistulous connection in our case would be the presence of a tracheal dystonia. Early reports demonstrated sarcoid induced weakening in the tracheal wall, with the size of such weaknesses dependant on the extent of intratracheal pressure.⁵ Such a condition would lead to an inherent weakness in the membranous tracheal wall, most probably secondary to chronic inflammation and subsequent destruction of the supportive elastic fibres as well as cartilaginous structures.² This then essentially represents a form of end-stage tracheomalacia.

Conclusions

Tracheal involvement of sarcoidosis and its tendency to result in stenosis is a phenomenon that has been described extensively in the literature. To date, there has been an absence of data to support the pathogenesis of a tracheo-oesophageal fistula in such patients. Despite its rarity, this particular case highlights the need for a high index of clinical suspicion in any sarcoidosis patient with evidence of repeat aspiration with no other underlying cause.

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