The Journal of Laryngology & Otology (2010), ***124***, *1333–1336.*

doi:10.1017/S0022215110001362

Clinical Record

Tracheotomy bleeding from an unusual tracheo-arterial fistula: involvement of an aberrant right subclavian artery

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Abstract

Introduction: The development of a tracheo-arterial fistula is a rare but severe complication of tracheotomy. The reported patient presented with such a fistula involving an aberrant right subclavian artery, termed an arteria lusoria, an aortic arch abnormality which is usually asymptomatic.

Case report: A 30-year-old man was admitted to our institution for bleeding through his tracheotomy. He had been treated for advanced Duchenne muscular dystrophy, involving invasive assisted ventilation, since the age of 10 years. Surgical exploration and computed tomography scanning revealed a tracheo-arterial fistula involving an aberrant right subclavian artery, associated with severe scoliosis. Emergency, transient haemostasis was achieved by over-inflation of the tracheostomy tube cuff. Aneurysm ablation was successfully achieved as the result of an endovascular interventional radiology procedure.

Discussion: Arteria lusoria is one of the most common aortic arch abnormalities. The occurrence of an aneurysm of this artery in a tracheotomised patient has not previously been described. In Duchenne muscular dystrophy patients, spinal deformities may result in thoracic compression, which may alter the anatomical relations of mediastinal vessels. Such deformities are slowly progressive. Thus, vigilance is required in the long term management of Duchenne muscular dystrophy patients with a tracheostomy.

Key words: Tracheotomy; Fistula; Subclavian Artery; Duchenne Dystrophy

Introduction

The development of a tracheo-arterial fistula is an unusual but severe complication of persistent tracheostomy, with high morbidity and mortality rates. The survival rate following tracheo-arterial fistula bleeding has been reported as 14.3 per cent.[1]

An aberrant right subclavian artery, termed an arteria lusoria, is the most common aortic arch abnormality and is usually asymptomatic.

Herein, we describe a case of major bleeding from an arteria lusoria aneurysm via a tracheotomy wall fistula, in a patient with muscular dystrophy disease. This unique occurrence emphasises the need for long term vigilance in the care of tracheostomised patients, particularly those with significance comorbidity.

Case report

A 30-year-old man presented to the emergency department with bleeding from his tracheotomy. He had previously received long term treatment for Duchenne muscular dystrophy, and had required invasive assisted ventilation for chronic respiratory failure since the age of 10 years. Hitherto, this treatment had been well tolerated. The patient's mother performed tracheostomy tube placements every week. The last change had been performed using a low pressure cuff tube, without difficulty. Prior to presentation, the patient had developed an abrupt tracheotomy haemorrhage after an acute coughing episode.

On examination, there was no active bleeding, but a bulky blood clot around the tracheostomy tube was noticed. Flexible fibre-optic endoscopic examination of the tracheal tube was unremarkable, with no active bleeding or mucosal erosion. However, removal of the tracheostomy tube induced massive bleeding; this was stopped by over-inflation of the cannula cuff.

Surgical exploration under general anaesthesia was immediately undertaken. The brachiocephalic trunk and carotid arteries were exposed. No vascular injury was seen. Through the tracheotomy, bleeding was identified emerging from an erosion of the posterior tracheal wall. A diagnosis of posterior tracheo-arterial fistula was suggested. A new cuffed tracheostomy tube was placed, and the cuff over-inflated.

A thoracic computed tomography (CT) scan with contrast injection showed an aberrant right subclavian artery (arteria lusoria) adherent to the damaged tracheal posterior wall. Tracheostomy cuff inflation had achieved emergency haemostasis by crushing the aneurysm against the adjacent dorsal vertebra (Figures 1 and 2). A scoliosis deformity was also observed, causing unusual contact between the trachea, the arteria lusoria anomaly and the spine (Figure 3). As a result of these radiological features, a diagnosis of tracheo-arterial fistula from an aberrant right subclavian artery was highly suspected.

A surgical mediastinal approach was not considered in the first instance, due to the risks of the procedure and the patient's weak general condition. Instead, a vascular interventional radiology procedure was performed via a

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Accepted for publication: 22 March 2010. First published online 11 June 2010.

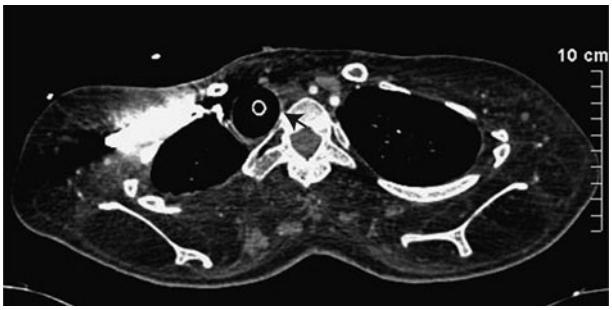

FIG. 1

Axial, contrast-enhanced computed tomography scan of the thorax (mediastinal window setting). The aberrant right subclavian artery (arrow) is located lateral to the trachea and behind the oesophagus, next to the spine.

femoral approach. Initial angiography confirmed the presence of an arteria lusoria aneurysm, with extravasation of contrast material (Figure 4). The aneurysm was ablated by percutaneous implantation of a 7×40 mm Fluency Plus® stent graft (Bard Peripheral Vascular, Tempe, Arizona, USA). The self-expanding, covered stent (7 mm in diameter and 40 mm in length) was advanced along a guide wire and positioned at the aneurysm site. Thereafter, no residual bleeding was seen. All adjacent vessels remained permeable, with good patency of the right aberrant subclavian artery (Figure 5).

The patient's post-procedure course was uneventful, with no recurrent bleeding or right arm ischaemia. He was admitted to the intensive care unit for medical supervision.

The patient was discharged six weeks later, and the previous home management of his respiratory failure was resumed.

Discussion

Post-tracheotomy bleeding from a tracheo-arterial fistula is unusual, but is one of the most devastating long term complications of tracheotomy. The incidence of tracheo-arterial fistula ranges from 0 to 1 per cent, with a mean of 0.3 per cent.[2–4] The brachiocephalic (innominate) arteries and the carotid arteries are often involved.

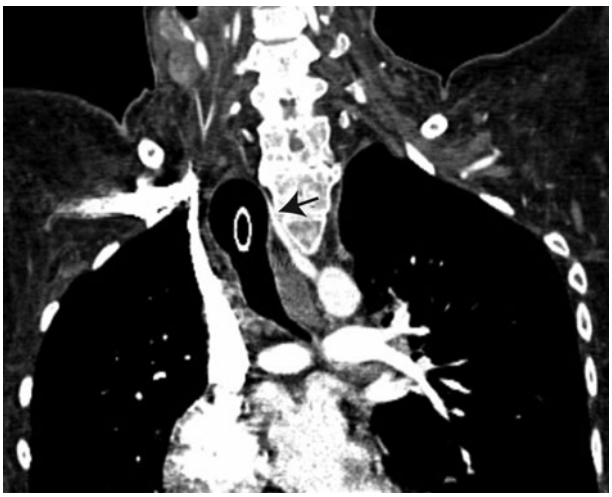

FIG. 2

Coronal, contrast-enhanced computed tomography scan of the thorax (mediastinal window setting). The arteria lusoria (arrow) arises from the aortic arch and is pushed back onto the adjacent dorsal vertebra by the tracheostomy cuff projecting into the trachea.

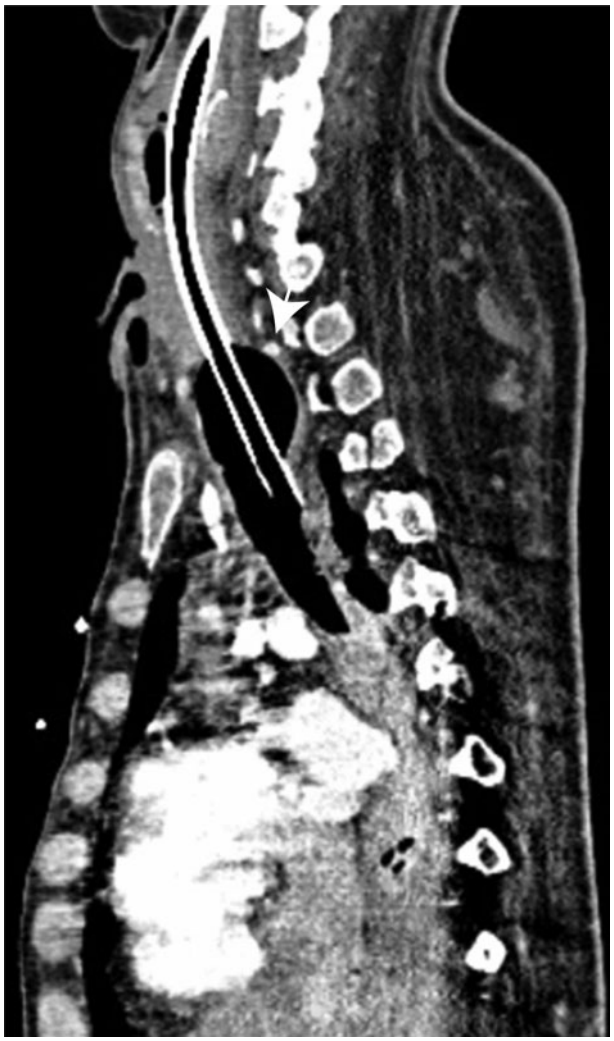

FIG. 3

Sagittal, contrast-enhanced computed tomography scan of the thorax (mediastinal window setting), showing a curved indentation of the posterior trachea wall caused by the tracheostomy cuff, allowing haemostasis of the arteria lusoria anomaly (arrow).

An aberrant right subclavian artery, termed an arteria lusoria, is the most common aortic arch abnormality, occurring in 0.5 to 2.5 per cent of the general population.[5] In this vascular malformation, four vessels arise sequentially from the aortic arch; the arteria lusoria is the last one, after the left subclavian artery. The aberrant right subclavian artery courses alone from the left side to the right side of the chest, usually behind the oesophagus (in 80 per cent of cases), as in our patient. In the remaining cases, it crosses between the oesophagus and the trachea (15 per cent), or in front of the trachea (5 per cent).[5]

This vascular malformation is usually asymptomatic, occurring as an isolated anomaly. However, in rare cases it can be associated with vascular or cardiac abnormalities, with clinical consequences. Atherosclerosis can induce 'dysphagia lusoria', with oesophageal compression.[6] Dyspnoea or chronic coughing can occur in cases of tracheal compression. Finally, aneurysmal dilation of this artery with oesophageal fistulisation has been described.[5,7] There have been no previous reports of arteria lusoria fistulisation into the trachea after previous tracheotomy. Thus, an

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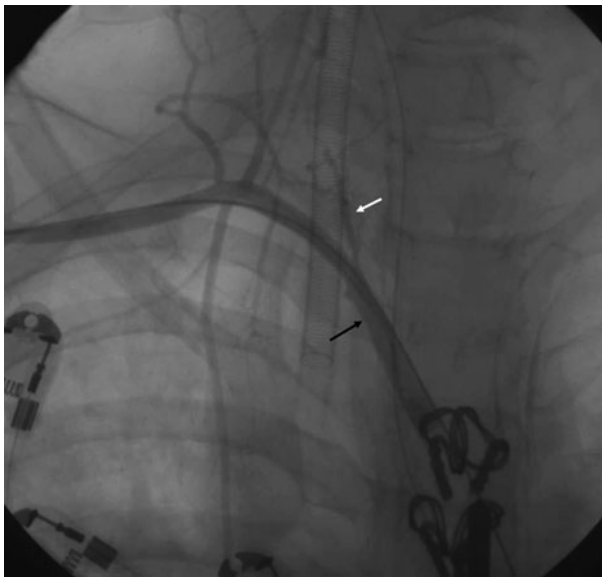

FIG. 4

Pre-treatment arteriography of the supra-aortic branches. Opacification indicates the aberrant right subclavian artery arising from the left. The eroded aneurysm (black arrow) causes active bleeding (white arrow).

aberrant right subclavian artery should be kept in mind as a potential cause of tracheotomy bleeding, similar to brachiocephalic and carotid arteries.

Patients with Duchenne muscular dystrophy and a tracheostomy seem to have a higher incidence of tracheo-arterial fistula. In these patients, nearly 10 per cent of post-tracheotomy haemorrhages are induced by a tracheo-arterial fistula.[8] Some reports indicate that, in this patient population, tracheo-arterial fistula may be associated with the frequent occurrence of spinal deformity.[8] Scoliosis often constricts the chest cavity of such patients, resulting in unusual anatomical relations of the trachea and the brachiocephalic arteries. In our patient, a combination of a retro-oesophageal arteria lusoria and spinal scoliosis appeared to result in increased mechanical forces pressing the artery against the tracheostomy cannula.

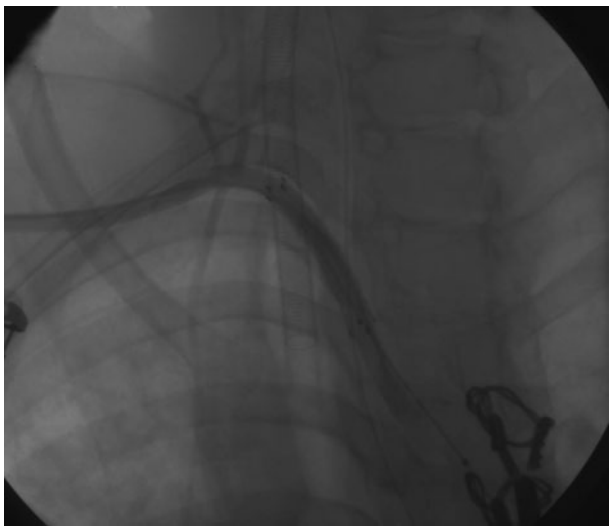

FIG. 5

Post-treatment arteriography of the right subclavian artery. The self-expanding, covered stent is positioned at the aneurysm site, with good patency of the right subclavian artery and distal branches.

The spinal deformities associated with Duchenne muscular dystrophy are gradually progressive; thus, so is the resulting vascular compression. This fact probably explains the long delay (10 years) between our patient's tracheotomy procedure and his bleeding event; this is one of the longest such delays described, and is in agreement with an aetiology of progressive spinal distortion and aneurysm formation.[3,8,9] Thus, a post-tracheotomy haemorrhage can appear several months after the surgical procedure. A tracheo-arterial fistula must be always suspected in cases of tracheostomy bleeding, especially in Duchenne muscular dystrophy patients.

- **Tracheo-arterial fistulae usually involve the brachiocephalic (innominate) or carotid arteries**
- **An aberrant right subclavian artery, termed an arteria lusoria, is the most common aortic arch abnormality**
- **Aneurysmal disruption of an arteria lusoria in patients with spinal deformities should be kept in mind as a potential cause of tracheotomy bleeding in patients requiring long term follow up**

Several precautions have been advised to prevent the development of tracheo-arterial fistula. The tracheotomy should not be performed under the third tracheal ring.[1,8,9] Tracheostomy cuff pressure should be limited to 20 mmHg.[1,4,8] In the intensive care unit, neck hyperextension and repetitive pulling on the cannula should be avoided.[4,8–10] Tracheotomy infections must be treated adequately to avoid tissue necrosis.[10] In patients with myopathy, it is suggested to perform a thoracic CT scan before the tracheotomy in order to clearly establish the anatomical location of blood vessels. The follow up of such patients should include repeated CT scanning; however, no clear scheduling procedure has been described.[9]

Conclusion

Post-tracheotomy bleeding from a tracheo-arterial fistula is unusual. It is generally caused by a brachiocephalic trunk or carotid aneurysm. However, the presence of an aberrant right subclavian artery, termed an arteria lusoria, is not rare, and surgeons should be aware of this potential aetiology for tracheo-arterial fistula. Moreover, the development of tracheo-arterial fistula is probably influenced by spinal deformities such as those found in Duchenne muscular dystrophy patients, which may result in unusual anatomical relations of the mediastinal vessels. Such distortion is gradually progressive, and long term vigilance is hence required in the management of Duchenne muscular dystrophy patients with a tracheostomy.

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Dr G Mortuaire takes responsibility for the integrity of the content of the paper.
Competing interests: None declared