CASE REPORT

Ortner's syndrome: Cardiovocal syndrome caused by aortic arch pseudoaneurysm

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72-year-old hypertensive presented with two weeks history of left sided chest pain and hoarseness. Workup demonstrated a pseudoaneurysm in the lesser curvature of the distal aortic arch opposite the origin of the left subclavian artery from a penetrating atherosclerotic ulcer. Following a left carotid-subclavian bypass, endovascular stenting of the aorta was performed excluding the pseudoaneurysm. Patient had excellent angiographic results post-stenting. Follow up at 12 weeks demonstrated complete resolution of his symptoms and good stent position with no endo-leak.

Ortner's syndrome describes vocal changes caused by cardiovascular pathology. It should be included in the differential diagnosis of patients with cardiovascular risk factors presenting with hoarseness. This case demonstrates the use of endovascular stents to treat the causative pathology with resolution of symptoms. In expert hands, it represents low risk, minimally invasive therapeutic strategy with excellent early results in patients who are high risk for open procedure.

Keywords: Ortner's syndrome, Cardio-vocal syndrome, Pseudoaneurysm, Endovascular stent

Introduction

In 1897, Norbert Ortner [1] described hoarseness caused by recurrent laryngeal nerve paralysis in patients with a large left atrium due to mitral valve stenosis. Since then, the term *Ortner's syndrome* has been used to describe cardiovocal syndrome caused by left recurrent laryngeal nerve (LRLN) paralysis from various cardiac or aortic pathology.

We present a patient with cardiovocal syndrome caused by aortic pseudoaneurysm from a penetrating atheroseclerotic ulcer that was successfully treated with thoracic endovascular aortic repair (TEVAR).

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Case presentation

A 72-year-old man with long standing, poorly controlled, hypertension presented with left-sided chest pain at rest of 2 weeks' duration. The patient described the pain as sharp and moderate in severity and denied any similar episodes in the past. Further review of the systems revealed changes in his voice for about the same duration, which he attributed to his heavy smoking. Blood pressure at presentation was 200/120 mmHg and pulse regular at 88 beats/min. Workup for acute coronary syndrome with echocardiography and serum troponin was negative. Chest computed tomography angiography (CTA) was ordered to rule out an acute aortic pathology given the history of long-standing, poorly controlled hypertension and the nature of the pain. The CTA demonstrated a pseudoaneurysm in the lesser curvature of the distal aortic arch just opposite the origin of the left subclavian artery, most likely from a penetrating atherosclerotic ulcer (PAU; Fig. 1). It measured 14 mm at the base and 16 mm deep. The location of the pseudoaneurysm was very close to the anatomical location of the left recurrent laryngeal nerve and probably explained the patient's hoarseness.

The patient was initially managed medically with aggressive antihypertensive medication to control the high blood pressure. Given the patient's age and comorbidities, it was decided that he was high risk for a conventional open repair and should undergo an endovascular repair.

A minimum of 2 cm proximal landing zone is required to properly deploy an endovascular stent and avoid endoleaks. Since the pseudoaneurysm was in close proximity to the origin of the left subclavian artery, the origin of the left subclavian artery needed to be covered during the procedure for adequate landing zone. To avoid any neurological or vascular complications from covering the left subclavian artery, the patient initially underwent a left carotid-subclavian bypass with an 8 mm polytetrafluoroethylene graft through a left supraclavicular incision stent. The bypass procedure was successful and 24 hours later he underwent TEVAR. The stent's diameter was sized to be at least 15% larger than the native vessel to be covered. Also, adequate proximal and distal seal to the area of interest should be at least 2 cm each. A single 28/161 mm Cook Zenith TX2 stent (Cook Medical Inc., Bloomington, IN, USA) was deployed via the right femoral artery covering the pseudoaneurysm (Fig. 2A). Poststent completion angiogram demonstrated isolation and lack of flow in the pseudoaneurysm (Fig. 2B).

Earlier examination of the vocal cords demonstrated palsy of the left vocal cord. The patient was treated conservatively in anticipation of possible spontaneous recovery of the LRLN palsy with regression of the pseudoaneurysm. At 12-week follow-up, the patient had regained his normal voice while examination showed normal movement of the left vocal cord. CTA of the aorta demonstrated no endovascular leak and resolution of the pseudoaneurysm (Fig. 3).

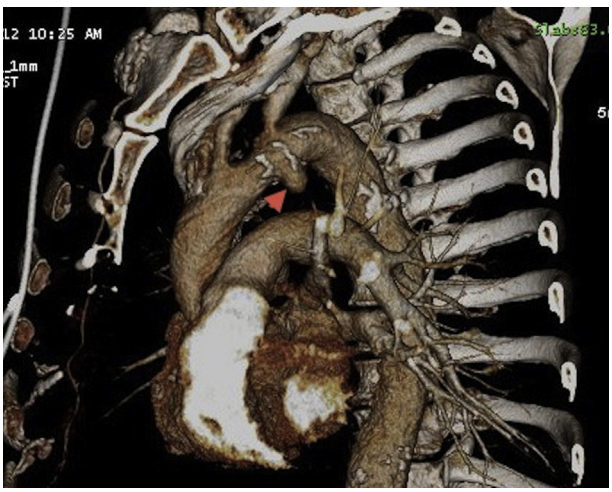

Figure 1. Sagittal images of the thoracic aorta demonstrating a pseudoaneurysm (arrow head) in the lesser curvature of the distal aorta.

Discussion

Norbert Ortner [1] deduced that an enlarged auricle caused compression of the LRLN against the aorta resulting in nerve palsy. In 1904, Alexander [2] described enlargement of the left pulmonary artery causing compression of the nerve against the aorta. Fetterolf and Norris [3], in 1911, after reviewing 37 cases with careful anatomic studies of frozen specimens, concluded that dilatation of the left auricle caused the left pulmonary vein to press against the pulmonary artery, the latter being forced against the aorta causing compression of the LRLN between the left pulmonary artery and aorta or ligamentum arteriosum.

Currently, cardiovocal syndrome is used to describe LRLN palsy caused by any cardiovascular pathology. Reports have included left heart failure, patent ductus arteriosus, Eisenmenger syndrome, primary pulmonary hypertension, and various other cardiac pathologies [4]. As more cardiovocal syndrome cases were reported,

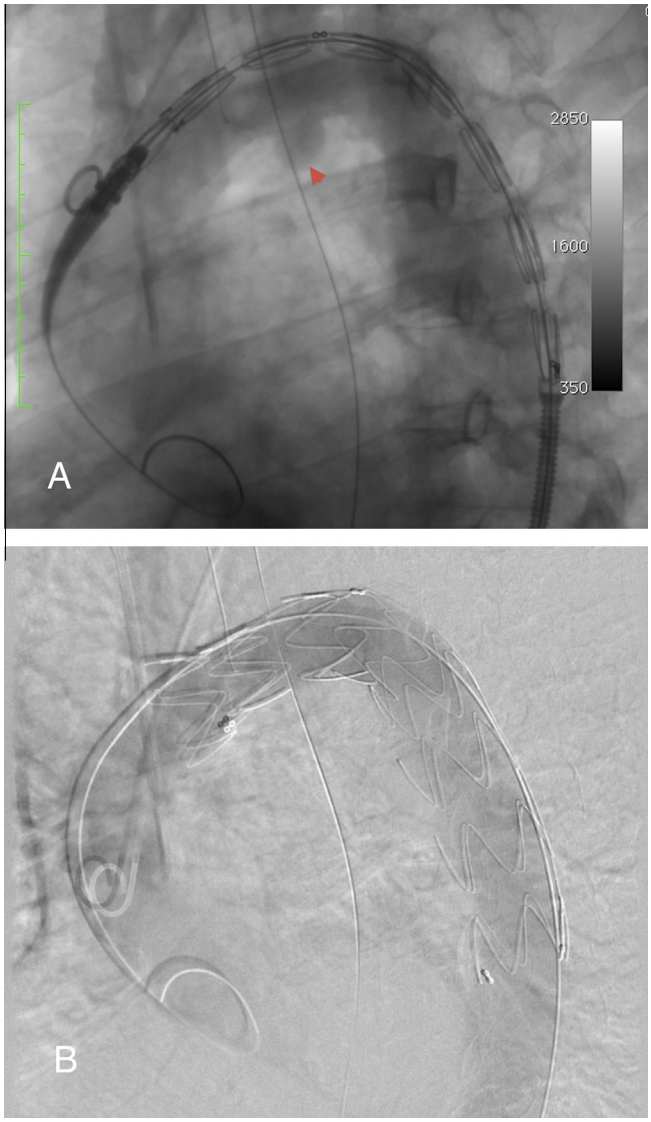

Figure 2. (A) Angiogram demonstrating the pseudoaneurysm (arrowhead). (B) Angiogram poststent deployment demonstrating obliteration of the pseudoaneurysm.

various new explanations of the cause have emerged such as lymphadenitis and scarring in the aortic window causing nerve fixation, pressure from the left bronchus, right ventricular hypertrophy, or pulmonary artery atherosclerosis [5].

In this case, the patient presented with hoarseness caused by pseudoaneurysm from a PAU. The nerve palsy is probably due to stretching of the nerve rather than compression as imaging showed the pseudoaneurysm to be in close proximity to the site where the LRLN loops around the aortic arch.

Treatment with TEVAR resulted in resolution of both the chest pain and hoarseness. It is a useful therapeutic strategy in cases of pseudoaneurysms as it isolates the pseudoaneurysm from the aortic lumen, allowing it to thrombose and regress. It also reinforces the aortic wall as pseudoaneurysms, such as those caused by PAU, lack the natural aortic wall layers and present areas of aortic wall weakness that are at risk of free rupture.

Open repair of thoracic pseudoaneurysms is considered the gold standard. However, it carries an inherently high risk given the nature of the procedure. By contrast, TEVAR has lower procedural risk. However, it has mixed results regarding resolution of symptoms. Stoob et al. [6] demonstrated the first successful use of endovascular repair to treat hoarseness caused by thoracic aortic aneurysm with resolution of symptoms. Ting et al. [7] although successfully excluded a

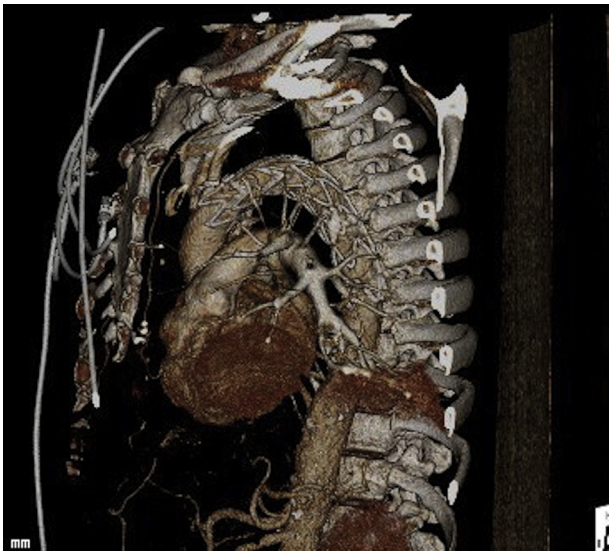

Figure 3. Sagittal computed tomography image of the thoracic aorta poststent deployment confirming obliteration of the pseudoaneurysm, good stent position and lack of endoleak.

mycotic aneurysm with endovascular repair, hoarseness did not resolve at 13-month follow-up.

Conclusion

With careful patient selection, this case demonstrates that TEVAR can be used in the treatment of aortic pseudoaneurysms with resolution of symptoms. It is a minimally invasive therapeutic modality with minimal risk and morbidity, and excellent initial and short-term results. It is an ideal option in patients who are at high risk for conventional open surgical procedure.

The case also demonstrates the importance of including cardiovocal syndrome in the differential diagnosis of hoarseness in patients with cardiovascular risk factors and performing the correct diagnostic investigations. Recovery of the nerve is anticipated with treatment of the causative pathology.

References

[1] Ortner N. Recurrent nerve palsy in patient with mitral stenosis. Wien Klin Wochenschr 1897;10:753–5 [in German].
[2] Alexander A. Ein Fall von Recurrenslähmung bei Mitralstenose. Berl Klin Wochenschr 1904;41:135–9.
[3] Fetterolf G, Norris G. Anatomical explanation of paralysis of left recurrent laryngeal nerve found in certain cases of mitral stenosis. Am J Med Sci 1911;141:625–38.
[4] Van Melle JP, Meyns B, Budts W. Ortner's syndrome, presentation of two cases with cardiovocal hoarseness. Acta Cardiol 2010;65:703–5.
[5] Subramaniam V, Herle TV, Mohammed N, Thahir M. Ortner's syndrome: case series and literature review. Braz J Otorhinolaryngol 2011;77:559–62.
[6] Stoob K, Alkadhi H, Lachat M, Wildermuth M, Pfammatter T. Resolution of hoarseness after endovascular repair of thoracic aortic aneurysm: a case of Ortner's syndrome. Ann Otol Rhinol Laryngol 2004;113:43–5.
[7] Ting AC, Cheng SW, Ho P, Poon JT. Endovascular repair for multiple *Salmonella* mycotic aneurysms of the thoracic aorta presenting with cardiovocal syndrome. Eur J Cardiothorac Surg 2004;26:221–4.