CASE REPORTS

CARDIOVOCAL SYNDROME – AN AORTIC ARCH ANEURYSM AS A RARE CAUSE OF VOCAL CORD PARALYSIS

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Abstract

Introduction: *Cardiovocal or Ortner's syndrome is a rare cause of vocal cord paralysis. Damage to the left recurrent laryngeal nerve may be caused by an aortic arch aneurysm, in even rarer cases.*

Clinical case: *A 60-year-old woman presented with hoarseness lasting for six months. Paralysis of the left vocal cord was confirmed with laryngoscopy and an aortic arch aneurysm was diagnosed on chest CT. Despite correction of the aortic aneurysm, her hoarseness did not improve.*

Discussion: *Mediastinal disease may cause vocal cord paralysis, due to the intrathoracic course of the recurrent laryngeal nerve. The assessment of the superior mediastinum on CT is mandatory in these cases. In cardiovocal syndrome, cardiovascular diseases damage the recurrent laryngeal nerve. Aortic aneurysms are a rare cause of Ortner's, especially when they affect the distal portion of the aortic arch and stretch the left recurrent laryngeal nerve at the aortopulmonary window.*

Keywords: *Cardiovocal syndrome, aortic arch aneurysm, vocal cord paralysis*

INTRODUCTION

Vocal cord paralysis may be caused by mediastinal disease, due to recurrent laryngeal nerve lesion[1]. The recurrent laryngeal nerve is a branch of the vagus nerve that innervates all intrinsic laryngeal muscles except for the cricothyroid. The left recurrent laryngeal nerve has a longer intrathoracic course than the right one, passing through the aortopulmonary window, under the aortic arch posteriorly to the ligamentum arteriosum, which makes it more vulnerable to damage in the setting of mediastinal abnormalities[1,2].

Cardiovocal or Ortner's syndrome is a rare cause of vocal cord paralysis, accounting for less than 11% of the cases of recurrent laryngeal nerve palsy[2,3,4]. In this syndrome, several cardiovascular mediastinal diseases damage the recurrent laryngeal nerve. In rare instances, compression and stretching of the left recurrent laryngeal nerve can result from the presence of an aortic arch aneurysm[2,4]. A timely diagnosis and treatment are essential for hoarseness recovery[4].

CLINICAL CASE

A 60-year-old woman was referred to our institution with complaints of hoarseness lasting for six months as an isolated symptom. She was a smoker (30 pack-year), had an history of hypertension, dyslipidaemia and depression and she had been submitted to bilateral saphenectomy due chronic venous insufficiency. Physical examination showed no abnormalities, revealing normal blood pressure, peripheral pulses, cervical palpation and cardiothoracic auscultation.

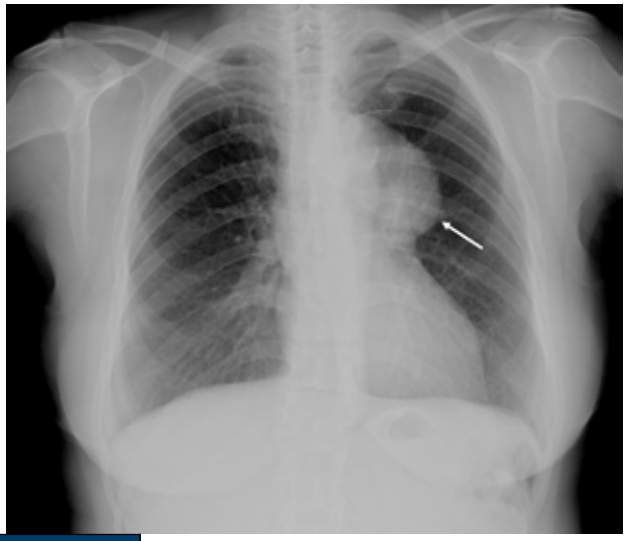

Figure 1 *Chest radiography – There is a large mediastinal mass at the level of the aorto-pulmonary window (arrow). Lung parenchyma does not show any abnormalities.*

Laryngoscopy detected a paralysis of the left vocal cord but no other local abnormalities that could be the cause.

A chest radiography was performed, showing a large mediastinal mass at the level of the aorto-pulmonary window, with no signs of lung disease (Fig. 1), which prompted the execution of a CT of the neck and chest.

Contrast-enhanced neck CT confirmed the left vocal cord paralysis and excluded local lesions (Fig. 2). Chest CT, performed before and after intravenous contrast administration, detected a large saccular aneurysm of the aortic arch, at the origin of the left subclavian artery, measuring 41 x 47 mm, with a mural thrombus measuring 2cm of maximum thickness (Fig. 3). No suspicious lung masses nor lymphadenopathies could be identified.

This patient was referred to cardiothoracic surgery and was submitted to aortic arch replacement using the frozen elephant trunk technique. The aneurysm sac was successful excluded and significantly reduced in size (Fig. 4). However, the patient's hoarseness did not improve, probably related to the compression of the left recurrent laryngeal nerve by the residual aneurysm sac and axonal damage from prolonged stretching.

DISCUSSION

Vocal cord paralysis can manifest with hoarseness, dysphagia or dyspnea during speech, although it can be asymptomatic, especially if unilateral, which may delay the diagnosis.

On CT scans, signs of vocal cord paralysis include the dilatation of the ipsilateral piriform sinus, medialization of the aryepiglottic fold and enlargement of the ipsilateral laryngeal ventricle – known as the sail sign – and anteromedial deviation of the arytenoid cartilage[5].

Assessment of the superior mediastinum (including the pulmonary bifurcation) is mandatory when vocal cord paralysis is identified. The most common mediastinal causes of recurrent laryngeal nerve lesions are neoplastic (bronchogenic, thyroid or esophageal cancers, metastatic disease) and post-surgical manipulation[1,6].

Although rare, cardiovascular pathology can also

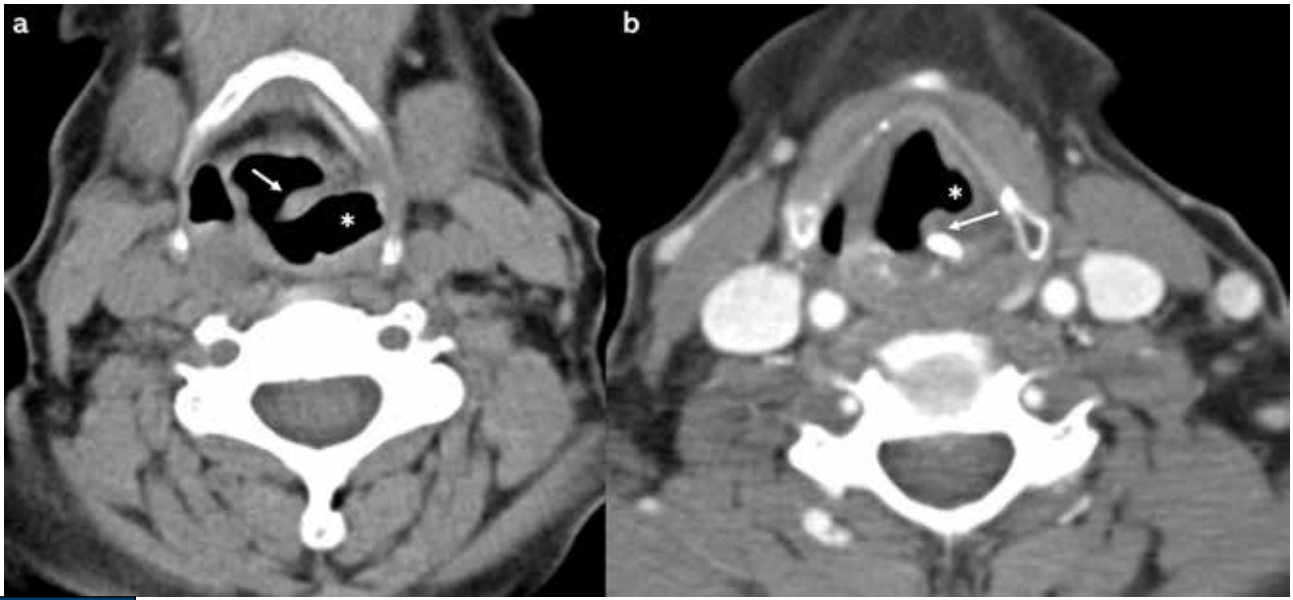

Figure 2 *Contrast-enhanced neck CT (quiet respiration) – a) At the level of the hypopharynx, there is distention of the left piriform sinus with air (*) and medial rotation of the ipsilateral aryepiglottic fold (arrow) b) Inferiorly, at the level of the vocal cords, the left aryepiglottic cartilage is anteromedially rotated (arrow) and the ipsilateral laryngeal ventricle is enlarged - sail sign (*). These are consistent with left vocal cord paralysis.*

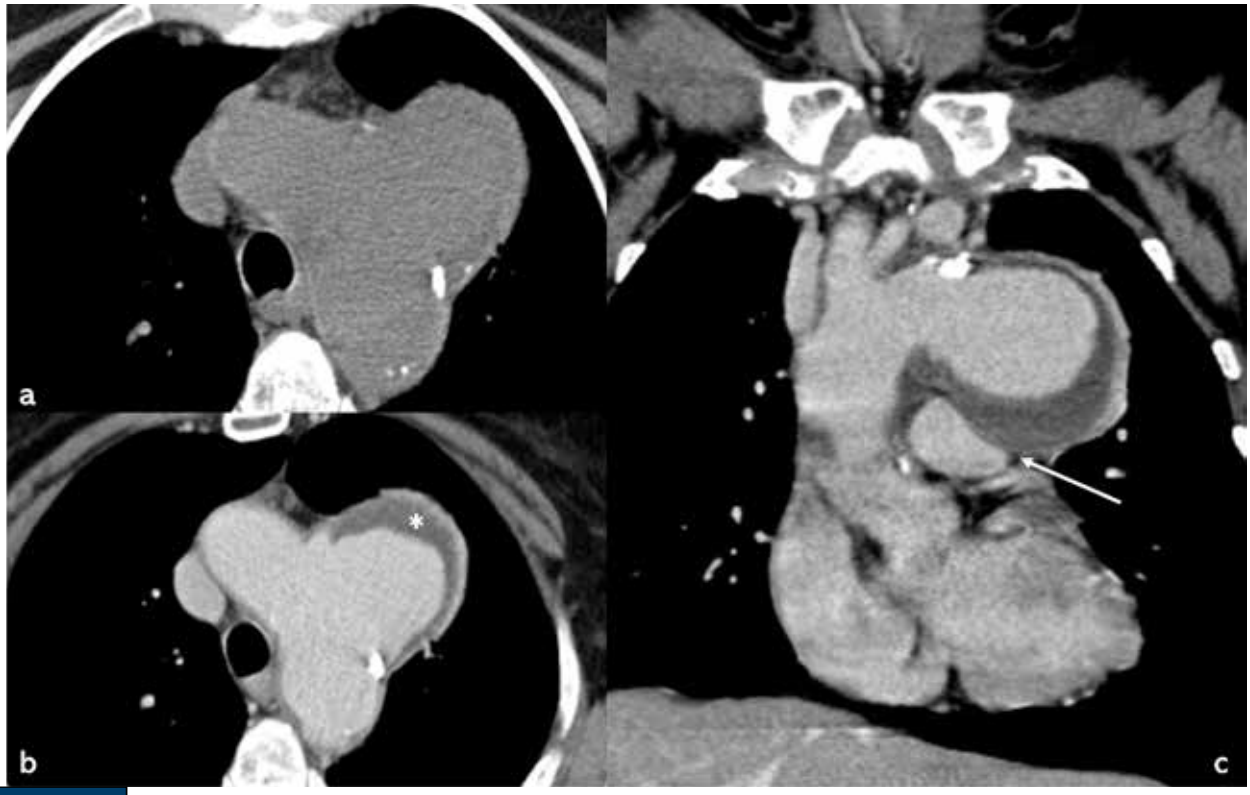

Figure 3

Chest CT – Non-enhanced (a) and post-contrast (b) axial CT show a large saccular aneurysm of the aortic arch, measuring 41 x 47 mm, with a mural thrombus measuring 18 mm of maximum thickness (*). The post-contrast coronal CT slice (c) shows the aneurysm arising at the level of the left subclavian artery, extending into the aortopulmonary window and causing an impression on the left pulmonary artery (arrow).

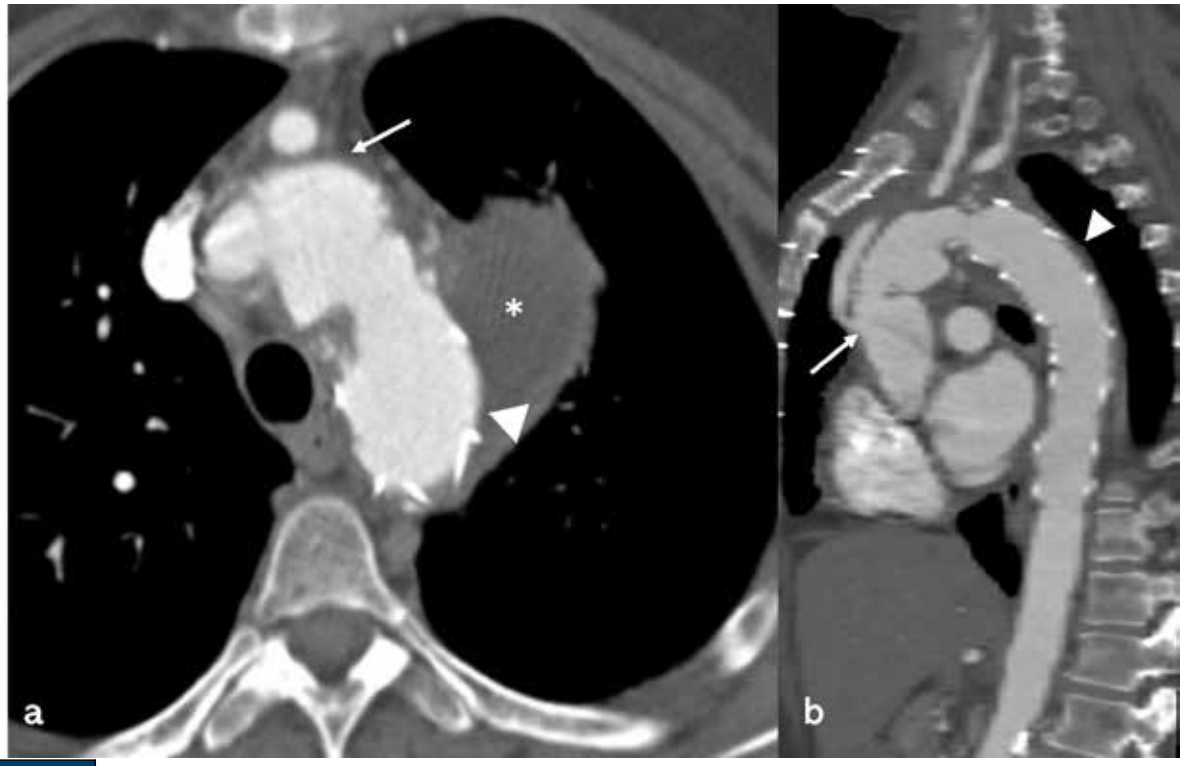

Figure 4

Post-surgery contrast-enhanced chest CT – Axial (a) and parasagital multiplanar reconstruction (b) reveal the presence of a stent on the descending thoracic aorta (arrowhead), distal to the aortic arch graft (arrow). There is complete thrombosis of the remaining aneurysmal sac (*).

cause recurrent laryngeal nerve palsy, which is termed cardiovocal or Ortner's syndrome, named after Norbert Ortner, who described the left vocal cord paralysis secondary to mitral stenosis, in 18975. Cardiovascular causes of vocal cord paralysis can be congenital, iatrogenic or related to several pathologies affecting the heart (mitral disease, atrial hypertrophy, left ventricular aneurysm), pulmonary vessels (primary or secondary pulmonary hypertension) or the aorta (aortic arch aneuryms). This syndrome accounts for less than 11% of the cases of recurrent laryngeal nerve palsy and it is more common in men[2,3,4].

Aortic aneurysms are a rare cause of vocal cord paralysis. They can be mycotic, atherosclerotic, traumatic or pseudoaneurysms, with or without dissection[2,4]. Most of these aneurysms affect the distal portion of the aortic arch, stretching the left recurrent laryngeal nerve at the aortopulmonary window. The aneurysmal sac size is the main determinant for compression. Recovery from hoarseness is variable and depends mostly on the time of diagnosis4.

In a patient presenting with vocal cord paralysis, apart from direct inspection with laryngoscopy, a chest CT should be performed, in order to evaluate the entire path of the recurrent laryngeal nerve.

The treatment of aortic arch aneurysms is indicated when the diameter is above 55mm when tubular aneurysms are considered. For symptomatic secular aneurysms, surgical thresholds are frequently lower. One of the treatment options in these cases is the Frozen Elephant Trunk (FET), consisting of a combination of a distal endovascular stent graft in the descending thoracic aorta and a proximal conventional surgical graft replacing the aortic arch. The usage of an endovascular prothesis limits the surgical exposure, while the replacement of the aortic arch prevents the appearance of late type A dissections. For this reason, the FET technique is the first line option for aortic arch treatment in many centers[7,8].

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