Unusual association of diseases/symptoms

Ascending aortic aneurysm causing hoarse voice: a variant of Ortner's syndrome

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Summary

A 68-year-old man with a persistent hoarse voice was found to have a left vocal cord paralysis. Clinical examination revealed signs consistent with aortic regurgitation. Subsequent investigation revealed an ascending aortic aneurysm. He underwent aortic root and ascending aorta replacement and his hoarseness improved. Ortner's syndrome refers to hoarseness due to recurrent laryngeal nerve palsy secondary to a cardiovascular abnormality. Recurrent laryngeal nerve palsy due to aneurysmal dilation of the ascending aorta is extremely rare, with aneurysms of the aortic arch being a more common cause.

BACKGROUND

Hoarse voice is a common symptom that frequently has a benign cause. However, unexplained or persistent hoarseness may indicate serious pathology and warrants further investigation. Ortner's syndrome is a rare cause of hoarseness, and refers to recurrent laryngeal nerve palsy caused by a cardiovascular abnormality. Here we present a case of hoarseness associated with an ascending aortic aneurysm causing left recurrent laryngeal nerve palsy that resolved with surgical repair of the aneurysm. There were clues in the history and clinical examination to point towards a cardiovascular cause for hoarseness, highlighting the importance of looking for such features and investigating appropriately.

CASE PRESENTATION

A 68-year-old man presented with a 3-month history of persistent hoarse voice. He described no other specific symptoms. His medical history included treated hypertension and rheumatic fever as a child. He had a 50 pack-year smoking history and drank little alcohol. Fibre-optic nasoendoscopy revealed a paralysed left vocal cord, with no other abnormality. Cardiovascular examination revealed a blood pressure of 200/80 mm Hg, a collapsing pulse and a diastolic murmur consistent with aortic regurgitation.

INVESTIGATIONS

Contrast-enhanced CT of the chest (figure 1) and subsequent echocardiography revealed aneurysmal dilation of the ascending aorta of 6 cm diameter, associated with moderate aortic regurgitation. The aortic arch appeared normal. No other abnormality was seen on the CT scan or on subsequent flexible bronchoscopy.

DIFFERENTIAL DIAGNOSIS

Vocal cord paralysis can be caused by laryngeal and extralaryngeal pathology. No tumour or ulceration was seen at nasoendoscopy, so an extralaryngeal cause was sought. In

older patients with a heavy smoking history, intrathoracic malignancy, particularly with mediastinal lymphadenopathy, is a common cause. In this case, the history of hypertension and rheumatic fever in combination with the abnormal findings on clinical examination, namely the wide pulse pressure, collapsing pulse and diastolic murmur, made a cardiovascular abnormality likely. Contrast-enhanced CT was the investigation of choice to seek these possible causes.

TREATMENT

The patient underwent surgical repair of the ascending aortic aneurysm with replacement of the aortic root and ascending aorta.

OUTCOME AND FOLLOW-UP

The patient's voice improved within a few weeks of surgery, and his hoarseness had completely resolved.

Figure 1 Contrast-enhanced CT of the chest showing aneurysmal dilation of the ascending aorta (arrowed).

within 4 months. He remained well 2 years following surgery, without any further hoarseness.

DISCUSSION

Ortner's syndrome

Ortner's syndrome, described by Ortner in 1897,¹ originally referred to the left recurrent laryngeal nerve palsy caused by left atrial enlargement due to mitral stenosis. The term is now used to describe recurrent laryngeal nerve palsy due to any cardiovascular abnormality, and is also called 'cardiovascular syndrome'.

Anatomy

The left recurrent laryngeal nerve is susceptible to compression by cardiovascular structures due to its path. After arising from the left vagus nerve, it travels down into the superior mediastinum and loops under the arch of the aorta before ascending to the vocal cords. The small space between the aortic arch and the pulmonary artery through which the left recurrent laryngeal nerve travels is the aorto-pulmonary window. As well as the left recurrent laryngeal nerve, this area also contains lymph nodes and the ligamentum arteriosum. The window is bound anteriorly by the ascending aorta, superiorly by the aortic arch, posteriorly by the descending aorta and inferiorly by the pulmonary artery.

Causes of Ortner's syndrome

Ortner's syndrome is a rare cause of recurrent laryngeal nerve palsy. In previous case series of extralaryngeal vocal cord palsy investigated by CT, only 1/33,² 1/62³ and 1/115⁴ cases were due to a cardiovascular cause. The most common cause of recurrent laryngeal nerve palsy in these series was superior mediastinal lymphadenopathy, most commonly due to malignancy.

A variety of cardiovascular causes of recurrent laryngeal nerve palsy have been described in the literature. Most cases are due to compression of the nerve by structures bounding the aorto-pulmonary window. Causes of Ortner's syndrome due to pulmonary artery enlargement include primary pulmonary hypertension,⁵ recurrent pulmonary emboli⁶ and various congenital heart defects.^{7–11} There is some debate as to whether the original cases that Ortner described were directly due to left atrial enlargement or due to the resulting pulmonary hypertension and enlargement of the pulmonary artery.¹⁰

Enlargement of the aorta is another cause of Ortner's syndrome, most commonly due to aneurysmal dilation of the aortic arch.^{12–13} In our patient, the aortic arch appeared normal, with aneurysmal dilation of the ascending aorta only. There was therefore some doubt as to whether our patient's hoarseness and recurrent laryngeal nerve palsy was due to the aneurysm, which resulted in further investigation in the way of bronchoscopy to ensure a second pathology was not being missed. However, the ascending aorta does abut the aorto-pulmonary window, and therefore aneurysm of the ascending aorta causing recurrent laryngeal nerve palsy is

anatomically plausible. It was felt that the severity of the patient's aortic regurgitation and the size of the aneurysm warranted repair irrespective of his hoarseness on cardiovascular prognostic grounds alone. The recovery of his voice postoperatively supports the diagnosis of cardiovocal hoarseness.

This case highlights the wide differential diagnosis of persistent hoarse voice. While malignancy is the most common extralaryngeal cause, Ortner's syndrome should be considered in patients with a history or clinical examination consistent with cardiovascular disease.

Learning points

- Persistent or unexplained hoarse voice may indicate serious laryngeal or extralaryngeal pathology.
- Ortner's syndrome refers to recurrent laryngeal nerve palsy due to a cardiovascular abnormality, such as enlargement of the aorta or pulmonary artery causing compression of the nerve at the aorto-pulmonary window.
- In rare cases, hoarse voice may be the first symptom of a thoracic aortic aneurysm. The diagnosis should be considered in patients with clinical features suggestive of cardiovascular disease, with contrast-enhanced CT being the investigation of choice.

Competing interests None.

Patient consent Obtained.

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