Hoarseness of voice as a primary presentation of pulmonary hypertension: a rare case of Ortner’s (Cardio-vocal) syndrome

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Abstract

Ortner’s (cardio-vocal) syndrome is a rare disease entity that involves compression of the left recurrent laryngeal nerve due to enlargement of cardiovascular structures. The syndrome was first described by Norbert Ortner (an Austrian physician) in 1897 and is usually reported to be associated with mitral stenosis. However, we report a rare case when a 20-year-old girl presented with hoarseness of voice and was found to have an enlarged main pulmonary trunk due to pulmonary hypertension that was compressing the left recurrent laryngeal nerve. The case highlights a rare presentation of a relatively common disease and stresses the importance of detailed cardiac workup in a patient with hoarseness of voice. We present the case and review the literature.

Key words: Ortner’s syndrome, Pulmonary hypertension, Vocal cord palsy.

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Case description

Enlargement of cardiovascular structures can frequently affect the left (unlike right) recurrent laryngeal nerve and cause left vocal cord paralysis as patients present with hoarseness of voice. This disease entity has been named as Ortner’s syndrome after the name of the physician who first described it. It presents a diagnostic challenge in the absence of significant cardiac symptoms. We report a young girl who is diagnosed with this condition and present the related literature.

A 20-year-old lady was referred from primary health center due to complaints of hoarseness of voice for the last four months. She reported only mild exertional palpitations and shortness of breath. She did not report any previous cardiac problem in childhood. Examination revealed a loud pulmonary component of 2nd heart sound, a pan-systolic murmur at the tricuspid area, and a palpable thrill. There was no evidence of right heart failure or any stigmata of connective tissue disease. Electrocardiography showed sinus rhythm with dominant R-wave in lead V1 indicating right ventricular hypertrophy. Laboratory tests did not reveal any abnormality.

Figure 1:
2-D trans-thoracic echocardiography, parasternal short axis view, showing dilated main pulmonary artery.

Figure 2:
Computerised tomography of the chest indicating severely dilated main pulmonary trunk (42 mm) with no evidence of pulmonary embolism.
2-D transthoracic echocardiography revealed normal left ventricle in size and function, right ventricular hypertrophy (6 mm thickness of free wall), mild to moderate tricuspid regurgitation, pulmonary hypertension (pulmonary pressure 60-65 mmHg) and an enlarged main pulmonary trunk (figure 1).

Computed tomography (CT) of the chest showed dilated main pulmonary trunk (max diameter 42 mm, figure 2). An angiogram excluded pulmonary emboli.

The patient was also seen by the Otolaryngology team in view of the main complaint of hoarseness of voice. Indirect laryngoscopy showed left vocal cord palsy. CT scan of the neck excluded a mass lesion or enlarged lymph nodes. We concluded that the enlarged pulmonary trunk secondary to pulmonary hypertension has compressed the left recurrent laryngeal nerve as it passes between the aorta and the pulmonary artery, thus causing left vocal cord paralysis. The case has been referred to the tertiary center where she will be reviewed by the pulmonary hypertension team and the thoracic surgeons, and will be followed in our clinic.

Discussion and literature review

Hoarseness of voice is a frequent presentation in the otolaryngology department. The causes include neoplastic (32%), surgical (30%), idiopathic (16%), traumatic (11%), central (8%) or infectious (3%)1. However, cardio-vascular related hoarseness is unusual. Norbert Ortner (an Austrian physician) first described the condition in 1897, when he reported three patients with mitral stenosis and left atrial enlargement, and postulated that the dilated left atrium compressed the left recurrent laryngeal nerve against the aortic arch, and caused left vocal cord paralysis2. However, a series of careful autopsies and radiological studies in the in the early 20th century disputed his hypothesis. On the basis of the autopsy studies, Fetterroff and Norris showed that the distance between the aorta and pulmonary artery within the aortic window is only 4 mm and suggested that compression of the nerve between the two structures is responsible for the nerve palsy3. Understanding the anatomy is crucial. The left recurrent laryngeal nerve arises from the left vagus nerve at the level of the aortic arch curve. It then curves around the aorta on the outer side of the ligamentum arteriosum and ascends in the groove between the esophagus and the trachea. The nerve then continues along the tracheoesophageal groove to supply all the muscles of the left vocal cord except the cricothyroid muscle. This prolonged course makes it vulnerable to injury due to pathology in the surrounding structures. Hoarseness of voice might be the only symptom in these patients. Shi-Min Yuan 4 noted that 35% of the patients with Ortner’s syndrome present with hoarseness only. Dyspnea with or without cough (32%) and dysphagia (11%) are the other major symptoms. Aortic aneurysm (atherosclerotic or degenerative) is the most common risk factor noted followed by congenital heart defects (patent ductus arteriosus, atrial septal defect, ventricular septal defect), left atrial disorders and pulmonary artery dilatation4. Indirect laryngoscopy is needed to diagnose vocal cord palsy. Further investigations include echocardiography, computed tomography (with angiogram) and magnetic resonance imaging to conclude in most cases. The presented case had enlarged pulmonary artery secondary to pulmonary hypertension. The pulmonary artery aneurysm had compressed the left recurrent laryngeal nerve as it passes between the aortic arch and pulmonary artery. Early diagnosis of Ortner’s syndrome may be helpful in starting prompt treatment, restore the vocal cord function and avoid permanent damage5. Clinical recognition of hoarseness in patients with cardiovascular disease should lead to immediate referral for laryngoscopy to confirm nerve palsy. A prompt assessment for aspiration, increased vocal effort, altered voice quality, dyspnea on exertion and decreased quality of life should be made. Absolute indications for vocal cord surgery include aspiration pneumonia and patient’s desire to improve voice-related quality of life (like singers). The surgery involves medialisation of the paralysed vocal cord so that glottic space closes during phonation and the normal vocal cord can make contact with the paraled one. In a study of 90 patients with Ortner’s syndrome, 45% had surgery, 13% had interventional treatment and 42% were treated conservatively6. The recovery in hoarseness is highly variable. Out of 58 patients in the same study who were followed-up, the hoarseness resolved in 44%, improved in 29%, persisted in 22% and exacerbated in 3% patients. Sometimes treating the causative factor may help symptoms. Stobø K7 reported resolution of hoarseness after endovascular repair of thoracic aortic aneurysm.

Ortner’s syndrome is rare. Although aortic aneurysms are the most common risk factor, pulmonary hypertension with dilated pulmonary artery can compress the left recurrent laryngeal nerve and cause hoarseness of voice. The case clearly identifies the importance of detailed cardiac workup in patients presenting with hoarseness. Early diagnosis helps in guiding immediate treatment and may alleviate the distressing symptoms.

Statement of ethical publishing

The authors state that they abide by the statement of ethical publishing of the International Cardiovascular Forum Journal8.

Conflict of interest:

The authors declares that there is no conflict of interest.

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