Case Report

Cardiovocal Syndrome – a rare cause for hoarseness of voice

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Abstract

Introduction
Hoarseness of voice is a common clinical presentation in ENT practice, which may have different etiological causes. The causes may vary from local pathology of larynx to a neurogenic reason of central nervous system or recurrent laryngeal nerve pathology. Cardiovocal syndrome may cause hoarseness secondary to left recurrent laryngeal nerve palsy when left recurrent laryngeal nerve is mechanically affected due to enlarged cardiovascular structure. In this article we present a case of left recurrent laryngeal nerve palsy due to Cardiovocal syndrome or Ortner’s syndrome in a 75-year-old female.

Conclusions
Ortner's syndrome may be overlooked and unless specifically looked for may be regarded as an idiopathic cause. The diagnostic methods currently used for investigation of recurrent laryngeal nerve palsy specially the CT scan of carina to skull base may have these inherited disadvantages. As the lowest plane of the scan is so proximal to inferior region of the aorto pulmonary window an interpreter may easily miss the aneurism unless specifically looked for. One effect of this would be under diagnosis and when spotted it would be highly inadequate to assess the heart and rest of the thoracic aorta as clearly demonstrated in our patient which needed further studies. It would be a more efficient to include the heart in the CT scans in patients with extra laryngeal causes of left recurrent laryngeal nerve palsy at least among cardiovascular risk factors.

Keywords: recurrent laryngeal nerve palsy, Cardiovocal syndrome, Ortner’s syndrome

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Funding: None

Competing interest: None

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Accepted Date: 22⁰ Feb 2021
Published Date: 12⁰ March 2021
A 75-year-old previously healthy female presented to the district general hospital Trincomalee with hoarseness of voice and cough for five months’ duration in December 2017. A fiberoptic laryngoscopy was performed on the same day revealing a left side vocal cord palsy. Simultaneously upper gastro intestinal endoscopy, a video bronchoscopy, contrast enhanced CT scan of skull base to carina and ESR was arranged. Neither Video bronchoscopy nor Upper gastro intestinal endoscopy managed to add new information, as both were normal. The ESR was age compatible with no evidence of existing inflammatory condition. The CECT scan of the neck and upper thorax was reported as evidence of left vocal cord palsy with prominent piriform fossa, bilateral small thyroid nodules. Ascending aorta and arch showing marked atherosclerotic changes and a 3.8cm by 2.8cm by 1.8cm sacular aneurysm arising from inferior surface of arch of aorta with a neck of 2.5cm without any thrombi formation or dissection.

Figure 1. CT appearance of left vocal cord palsy, with anteromedial displacement of arytenoid cartilage, paramedian vocal cords and enlarged laryngeal ventricle.

Figure 2. Sagittal section of CECT showing aortic aneurism in aorto pulmonary window.
The patient never had any history of chest pain or significant back pain, tuberculosis or childhood heart problems. No history of significant neck pain or other joint pathology. No past history of thoracic trauma or surgery. On examination there was no features of Marfan's syndrome, no papilledema, Examination of central nervous system showed rest of cranial nerves to be normal. Cardiovascular system examination showed mildly elevated blood pressure with thickened peripheral vessel walls and no cardiomegaly with normal heart sounds, there was normal air entry bilaterally and no added sounds on auscultation of the chest and precordium. Mantoux test for tuberculosis was negative, a Diagnosis of Ortner’s disease was made as there was no attributable other cause was visible. A thoracic CT angiogram and echo cardiogram was planned to assess the heart and rest of the mediastinum subsequently but patient went missing in the system.

**Discussion**

Vocal cord palsy may occur due to a local pathology of larynx; it’s surrounding or may be an extra laryngeal cause. A pathology in the vagus nerve or its recurrent laryngeal branch can be a neurological cause for a vocal cord palsy. The symptoms may manifest as a result of pressure on vagus nerve in its origin in the lateral sulcus posterior to the bulb, along its course in the bulbocerebellar cistern, in the jugular foramen as the nerve leaves its intracranial course and in the carotid sheath. The superior laryngeal nerve is given off in the carotid sheet. This nerve divides into two branches, internal laryngeal nerve which is associated with internal laryngeal artery to supply mucosa of piriform fossa and larynx above vocal cord while the external laryngeal nerve travelling with the superior laryngeal artery supplies the cricothyroid muscle. The other main branch of vagus that innervates larynx is the recurrent laryngeal nerve. The right recurrent laryngeal nerve arises from vagus nerve and hooks the right subclavian artery predominately, while at left always hooks the aortic arch the aortopulmonary window prior to its course in the tracheoesophageal groove.\(^1\) Recurrent laryngeal nerve innervates all laryngeal muscles apart from cricothyroid muscle and laryngeal mucosa below the vocal cords.
According to American academy of otolaryngology-head and neck surgery symptoms and signs of vocal cord palsy include the following:

- **Voice changes**, which include hoarseness; breathy voice; extra effort on speaking; excessive air pressure, required in producing usual conversational voice; and diplophonia.
- **Airway problems** include shortness of breath with exertion, noisy breathing, and ineffective cough and also swallowing problems; choking or coughing when swallowing food, drink, or even saliva, and food sticking in throat.

In addition, in the immediate post-operative period recurrent laryngeal nerve palsy can lead to reduction of pulmonary function as a result of loss of natural loss of positive end expiratory pressure [22]. Diagnosis is made via examination of vocal cords, laryngeal electromyography and imaging.

In a computerized tomography (CT) there are some interesting radiological features of recurrent nerve palsy these include the following.

- Atrophy of thyro-arytenoid muscle
- Anteromedial deviation of the arytenoid cartilage
- Paramedian vocal cords
- Enlarged laryngeal ventricle
- Atrophy of the posterior cricoarytenoid muscle

Out of the features mentioned above we managed to identify anteromedial deviation of arytenoid cartilage, paramedian vocal cords and enlarged laryngeal ventricle. (Ref figure 2.)

Recurrent laryngeal nerve palsy of left side leading to vocal cord palsy may be due to various reasons.

Causes of recurrent Laryngeal nerve palsy [2]

<table>
<thead>
<tr>
<th>Vascular/cardiac</th>
<th>Aortic dissection or pseudo aneurism, left atrium enlargement, congenital heart diseases, pulmonary artery enlargement, pulmonary embolism</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neoplastic</td>
<td>Bronchogenic carcinoma, lymphoma, oesophageal carcinoma, neurogenic tumours (paraganglioma, schwannoma), thyroid carcinoma, malignant thymus disease, lymph node metastasis; retrosternal goitre</td>
</tr>
<tr>
<td>Surgical/iatrogenic</td>
<td>Heart surgery, median sternotomy, patent ductus arteriosus ligation or embolization, left lobectomy/pneumonectomy, mediastinoscopy, radical esophagectomy, tracheal resection, thymectomy, thyroidectomy, anterior approach in spine surgeries, carotid endarterectomy, external radiotherapy</td>
</tr>
<tr>
<td>Inflammatory</td>
<td>Sarcoidosis, silicosis, fibrosing mediastinitis</td>
</tr>
<tr>
<td>Infiltrative</td>
<td>Amyloidosis</td>
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<tr>
<td>Infectious</td>
<td>Tuberculosis, histoplasmosis, coccidioidomycosis, bacterial abscess, mycotic aortic pseudo aneurysm</td>
</tr>
<tr>
<td>Traumatic</td>
<td>Deceleration injuries, penetrating chest injuries</td>
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</tbody>
</table>
Ortner’s syndrome or cardiovaocal syndrome occurs as a result of compression of recurrent laryngeal nerve secondary to a benign cardiovascular cause. Ortner’s syndrome / cardiovaocal syndrome was first described in 1897 among three patients with severe mitral stenosis, where in it was postulated that left atrial dilatation can cause left recurrent laryngeal nerve palsy due to the compression of the nerve against the aorta. [3] With more cases of cardiovaocal syndrome surfacing many have doubted the explanation offered by Ortner as left recurrent laryngeal nerve been compressed between aorta and left atrium. Ortner calculated the cause to be compression of the left recurrent laryngeal nerve by an enlarged left atrium [4]. Since then various authors have recorded their experiences of recurrent laryngeal nerve involvement in various cardiac disorders such as Eisenmenger complex [5], left ventricular failure [6], atrial septal defect [7], patent ductus arteriosus [8,9], primary pulmonary hypertension [10,11,12], recurrent pulmonary artery embolism [13], mitral regurgitation [14], atrial myxoma [15], left ventricular aneurysm [16], cor pulmonale [17] and various types of aortic aneurysms [18,9,21]. The type of aortic aneurysms that are associated with Left recurrent laryngeal nerve palsy includes Saccular, Atherosclerotic, Pseudo aneurysms, Dissections, Traumatic and Mycotic aneurysms. [22]

Following Ortner’s explanation and encounter of vocal cord palsy, existence of other cardiovascular causes leading to the neural compression between a dilated pulmonary artery and the aorta or aortic ligament was suggested based on X-ray and autopsy findings [8]. With further exposure of similar cases with time the explanation evolved further in to lymphadenitis and scarring in the aortic window causing nerve fixation. Among patients with tuberculosis hoarseness can occur either due to the tuberculous inflammation of the vocal cord (i.e. tuberculous laryngitis) or recurrent laryngeal nerve palsy as a result of acute lymphadenopathy, caseating granulomatous inflammation or late complications of chronic fibrosis [19] which may or may not due to Ortner’s syndrome.

Later more explanations were entertained such as pressure from the left bronchus, right ventricular hypertrophy, pulmonary artery atherosclerosis, anatomical position of the ligamentum arteriosum as a cause of cardiovaocal syndrome [7]. Another explanation was postulated as dilated pulmonary artery as the cause [10]. In a phenomena termed dynamic dilatation some suggested that patients with arteriosclerotic heart diseases suddenly suffered left recurrent laryngeal nerve paralysis because rapid onset of left ventricular failure produced sudden pulmonary hypertension with acute dilatation of the pulmonary vessels.

Causes of Ortner's syndrome due to pulmonary artery enlargement include primary pulmonary hypertension, recurrent pulmonary emboli [19] and various congenital heart defects. [20,12] Lung cancer has been attributed as the leading cause of recurrent laryngeal nerve palsy followed by surgical manipulation. Idiopathic causes contribute to less than 11%, Various case series attribute 1–3% of cases of extra laryngeal hoarseness to Ortner's syndrome [22].

CONCLUSION

Ortner’s syndrome may be overlooked and unless specifically looked for may be regarded as an idiopathic cause. The diagnostic methods currently used for investigation of recurrent laryngeal nerve palsy specially the CT scan of carina to skull base may have these inherited disadvantages. As the lowest plane of the scan is so proximal to inferior region of the aorto pulmonary window an interpreter may easily miss the aneurism unless specifically looked for. One effect of this would be under diagnosis and when spotted it would be highly inadequate to assess the heart and rest of the thoracic aorta as clearly demonstrated in our patient which needed further studies.

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