Atrial Septal Defect and Left Recurrent Laryngeal Nerve Paralysis: A Case of Ortner’s Syndrome and Literature Review

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Received 29 January 2015; accepted 22 February 2015; published 26 February 2015

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Abstract

Introduction: Cardiovocal syndrome, or hoarseness resulting from vocal fold paralysis secondary to cardiovascular pathology, is commonly referred to as Ortner’s syndrome. We present a brief overview of vocal fold paralysis, present an illustrative case of Ortner’s syndrome, and provide a review of the pertinent literature. Here we aim to broaden one’s differential for vocal fold paralysis, discuss its importance as pertains to cardiovascular pathology and outcomes, and highlight the difficulties in therapeutic planning for these unique patients. Methods: A case report and literature review. Results: A 26-year-old female with an atrial septal defect and pulmonary hypertension presented with 5 months of hoarseness. Laryngoscopy revealed left vocal fold paralysis. Imaging from the skull base to chest showed an enlarged pulmonary artery (PA) in the absence of other abnormalities. Literature review suggests that this left laryngeal nerve paralysis results from nerve compression within the aortopulmonary window, a triangle defined by the aortic arch, PA, and ligamentum arteriosum. Imaging in our patient over 8 months demonstrated an increase in PA size from 3.9 to 4.2 cm correlating with the onset of hoarseness. Conclusions: Importantly, hoarseness secondary to laryngeal nerve compression in cardiovascular disease may correlate with a poorer prognosis, i.e., in thoracic aortic aneurysms and mitral valvulostenosis. Awareness of vocal changes in the setting of cardiovascular disease improves diagnostic acumen in vocal fold-paralysis.

Keywords

Ortner’s, Cardiovocal, Vocalfoldparalysis

1. Introduction

Unilateral vocal fold paralysis (UVFP) primarily impacts a patient’s voice, but may result in dysphagia as well as dyspnea. The etiologies for UVFP have evolved over time but continue to include the following leading causes: 1) iatrogenic/postsurgical, commonly with procedures involving the anterior cervical spine, carotid artery, and thyroid; 2) neoplastic, commonly with thyroid and lung neoplasms; 3) others including trauma, systemic disease (e.g. infectious, inflammatory, neurologic, cardiovascular, medication related); and 4) idiopathic, which is a small but persistent group of cases [1]. An uncommon cause of unilateral vocal fold paralysis may result from a cardiovascular disease, Ortner’s syndrome, also known as cardiovocal syndrome. Originally described in 1897, Ortner’s syndrome detailed a left VFP resulting from recurrent laryngeal nerve compression within the aortopulmonary window, a triangle defined by the aortic arch, pulmonary artery, and ligamentum arteriosum [2] [3]. This phenomenon was originally described in the setting of mitral stenosis, but has since been correlated with a broader list of cardiovascular pathology (Table 1). Here we present an illustrative case of a 26-year-old female with an atrial septal defect and hoarseness.

2. Case Report

A 26-year-old Hispanic female was referred to otolaryngology for 5 months of hoarseness. Nearly one-year prior she was in her usual state of health training for Navy boot camp when she developed a progressive productive cough. She was found to have pneumonia, but among her work-up an EKG demonstrated right ventricular hypertrophy prompting referral to a cardiologist. A transthoracic echo was performed which demonstrated pulmonary hypertension with a dilated right atrium and ventricle. Weeks later she developed significant and sudden dyspnea on exertion and her exercise tolerance drastically diminished. Ultimately she required hospital admission, during which extensive cardiovascular workup revealed pulmonary hypertension and a secundum type atrial septal defect. Approximately 4 months after her discharge she began to develop hoarseness. She was then seen and evaluated in the head and neck surgery clinic 5 months later.

In clinic, her voice was hoarse. She had a persistent non-productive cough but denied any respiratory difficulties above her baseline dyspnea secondary to her cardiac disease. She denied any dysphagia. Notably she had no other prior medical or surgical history and denied any tobacco or alcohol use. On exam her neck was flat and without lymphadenopathy. Flexible laryngoscopy demonstrated a paralyzed left vocal fold in the paramedian position. Imaging from the skull base to aortic arch showed an enlarged pulmonary artery (PA) in the absence of other abnormalities consistent with Ortner’s syndrome (Figure 1 [4], Figure 2). A retrospective look at her prior imaging demonstrated an increase in PA size from 3.9 to 4.2 cm over an approximate 8-month period correlating with the onset of hoarseness and the progression of her cardiovascular disease.

Table 1. Cardiovascular pathologies that may result in vocal fold paralysis.

<table>
<thead>
<tr>
<th>Cardiovascular pathology</th>
<th>Ref.</th>
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<tbody>
<tr>
<td>Left ventricular failure secondary to HTN</td>
<td>[3]</td>
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<tr>
<td>Atrial septal defect (ASD)</td>
<td></td>
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<tr>
<td>Eisenmenger’s syndrome</td>
<td></td>
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<tr>
<td>Patent ductus arteriosus (PDA)</td>
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<tr>
<td>Primary pulmonary HTN (PPH)</td>
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<tr>
<td>Pulmonary embolism (PE)</td>
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<tr>
<td>Aortic arch pseudoaneurysm</td>
<td>[10]</td>
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<tr>
<td>Mitral stenosis (MS)</td>
<td></td>
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<td>Mitral regurgitation (MR)</td>
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<tr>
<td>Atrial myxoma</td>
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<tr>
<td>Left ventricle aneurysm</td>
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<td>Corpulmonale</td>
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<tr>
<td>Rheumatic heart disease</td>
<td>[11]</td>
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Figure 1. Axial CT with contrast at the level of the hypopharynx/supraglottis. Note the expansion of the left piriform sinus with air and medialization of the left aryepiglottic fold/false vocal fold (white arrow). CT evidence of VFP must not be confused with cervical neoplasms, arytenoid cartilage subluxation or fracture, or oblique imaging findings, all of which can mimic VFP.

Figure 2. Cross sectional imaging of aortopulmonary window. White arrow denotes aortopulmonary window, the borders of which are defined by the aortic arch (Ao), pulmonary artery (PA), and ligamentumarteriosum, shown in coronal (left), axial (middle), and sagittal (right) sections.

The patient was deemed a high operative risk from a cardiovascular standpoint. Operative intervention including medializationthyroplasty and/or arytenoid adduction would likely have improved her dysphonia. In addition, we discussed the temporary benefits she could obtain from an injection laryngoplasty but she elected against this option. Given her mild symptoms, the possibility of recovering nerve function by addressing her cardiovascular disease, and her high operative risk, the risks of proceeding with a more definitive laryngeal intervention on aggregate outweighed the benefits. As such we elected to await her cardiovascular treatment and recovery.

3. Discussion

Evaluation of dysphonia from UVFP begins with an auditory-perceptual evaluation, in which one may appreciate a hoarse or breathy voice. Laryngoscopy then provides direct visualization to confirm vocal fold paralysis or paresis. Imaging provides a means of determining the possible etiology. Imaging often begins with a chest X-ray but may include a CT or MRI typically from the skull base to aortic arch if the chest X-ray is unrevealing. High-resolution imaging allows for the assessment of any potential pathology along the course of the recurrent laryngeal nerve [1].
In this report we provide an example of a unique cardiovascular explanation for UVFP. Cardiovocal or Ort-
ner’s syndrome has been described in a wide array of cardiovascular pathologies that all result in encroachment
upon the aortopulmonary window; Table 1. A report from Japan described two cases of pulmonary hypertension,
one resultant from a longstanding PDA similar to our case, with new onset hoarseness. Here, cross sectional CT
imaging was shown to be an easy and noninvasive method for confirming the cause of hoarseness [3]. In Ort-
ner’s syndrome secondary to pulmonary hypertension, CT imaging demonstrates an enlarged pulmonary truncus
that exhibits cephalad displacement to be adjacent to the lower border of the aortic arch; namely, lying within
the same tomographic plane. Autopsy studies by Fetterrolf and Norris showed that the distance between the aor-
ta and the pulmonary artery within the aortopulmonary window is only 4 mm [5]. Hence, hoarseness in our pa-
tient corresponding to the dilation of her PA by 3 mm (3.9 to 4.2 cm) is a very reasonable and perhaps expected
finding.

The presence of VFP in the setting of cardiovascular disease (e.g. aortic aneurysms, mitral stenosis) is r e-
garded as a poor prognostic indicator. In the setting of VFP secondary to thoracic aortic aneurysms, some have
used the development of VFP to influence the need for surgical intervention [6] [7]. Some have even suggested
that all patients with select forms of cardiovascular disease be referred for laryngoscopy as part of their overall
clinical evaluation [3] [8]. Cardiovocal syndrome provides the head and neck surgeon with a unique dilemma.
Namely, these patients are high-risk surgical candidates with potentially reversible disease through correction of
their underlying cardiovascular pathology. 2 of 4 patients with thoracic aortic aneurysms and 3 of 8 patients with
distal aortic arch aneurysms had improvement or complete recovery in vocal fold mobility following cardiovas-
cular treatment [6] [9]. Our patient was still undergoing cardiovascular evaluation in the setting of her progres-
sive disease and undoubtedly was a high-risk surgical candidate currently compensating and functioning quite
well despite her VFP. As such, we collectively decided to await possible cardiovascular intervention and recov-
ery while clinically following her VFP.

4. Conclusion

Overall, it is important for the clinician to be aware that hoarseness in the setting of cardiovascular disease may
be due to cardiovocal syndrome. CT imaging provides an efficient means for diagnosis and the therapeutic
course must be well thought out in a multidisciplinary setting.

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