



Ortner's Syndrome: Review of Literature and Report on Four Patients with Hoarseness

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Introduction

Hoarseness, commonly referred to as dysphonia, is one of the most common speech-related complaints presented to otolaryngologists. Although used interchangeably, these terms represent two different diagnoses. Hoarseness refers to a change in voice quality noticed by the patient, while dysphonia refers to a change in voice quality observed by the physician [1]. Unique amongst the myriad of potential causes of vocal dysfunction is Ortner's Syndrome, also known as Cardiovoal Syndrome. This syndrome was first described in 1897 by Dr. Norbert Ortner in three patients with mitral valve disease. Ortner's Syndrome historically refers to left Recurrent Laryngeal Nerve (RLN) palsy due to a cardiac etiology, clinically presenting as hoarseness. This report discusses four patients with hoarseness due to Ortner's Syndrome - three cases caused by aortic aneurysm and one case caused an enlarged Pulmonary Artery (Pa).

Case Series

Case 1

An 85-year-old Caucasian male presented with a one-year history of progressive hoarseness. Medical history included coronary artery disease with stent placement and a two pack-per-day smoking history. Physical exam was unremarkable, however flexible laryngoscopy revealed left True Vocal Cord (TVC) paralysis while in the lateral position. The PET/CT scan without contrast revealed aortic root and ascending thoracic aorta dilation as well as aneurysmal dilation of the aortic arch. The most likely cause of the patients TVC paralysis was determined to be left RLN compression. The patient underwent elective thyroplasty with placement of a size 10 Montgomery implant. One month post-operatively, the patient reported that their opinion, their voice quality had returned to 70% to 80% of what it was before symptom onset.

Case 2

An 82-year-old African-American male was referred by an outside hospital for left TVC paralysis after presenting with a two-month history of dysphonia, breathy voice, globus sensation, and multiple episodes of coughing while eating or swallowing. Chest CT scan with contrast revealed an aortic arch dilation of 6 cm with an intraluminal thrombus. An aortic stent graft was present inferior to the dilation. In-office flexible laryngoscopy revealed left TVC paralysis with fixation in the paramedian position. Subsequent treatment *via* microlaryngoscopy with Cymetra injection significantly improved the patient's voice and maximal phonation time.

Case 3

A 68-year-old Caucasian male presented with complaints of hoarseness and trouble breathing following surgery for an aortic aneurysm. The surgery was further complicated by hemothorax and collapsed lung. He reported a twenty pack-year smoking history and denied complaints of dysphasia, dyspnea, and coughing. Flexible laryngoscopy revealed left TVC paralysis with a widened glottic gap during phonation. The patient underwent microlaryngoscopy with Cymetra injection, suspension microlaryngoscopy with injection medialization, and laryngoscopy with injection augmentation fat graft to the vocal cord. These procedures subjectively improved the patient's voice with only complaints of mild hoarseness and thick mucus production remaining.

Case 4

A 79-year-old Caucasian female presented with a four to five-month history of progressive hoarseness which reportedly worsened throughout the day with prolonged speaking. Notable medical history included scoliosis repair surgery two years prior and a treatment for breast cancer. Atrophic, bowed cords were found on previous exam, for which she was treated with a trial of proton

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pump inhibitors with no reported benefit. She endorsed tonal changes in her voice immediately after her scoliosis surgery in 2015. Chest CT scan revealed bilaterally enlarged atria and PA. The left PA measured 24 mm in diameter and the right PA measured 23 mm in diameter. In healthy adults, the average diameter of the left and right PAs are $18.8 + 2.5$ mm and $19.9 + 2.6$ mm, respectively [2]. The diameter of the pulmonary trunk measured 28 mm. Flexible laryngoscopy with stroboscopy revealed left TVC paresis with increased glottic gap. The patient was treated with dual, bilateral vocal fold injection medializations, which subjectively improved her symptoms one week post-operatively. Of note, no data associating scoliosis surgery with vocal cord dysfunction or voice changes were found in the literature. However, in a retrospective review by Shaw, Skovrlj and Chow, a clear association was seen between age and major complications following scoliosis surgery in adult patients. Their results revealed increasing rates of major short-term complications with each subsequent decade of life [3].

Discussion

Diagnosing left TVC paralysis due to extra-laryngeal nerve palsy remains a challenging task for most otolaryngologists. This becomes increasingly difficult when the underlying cause is seemingly unrelated or potentially an unknown cardiac pathology. A high level of suspicion and a keen knowledge of anatomy are invaluable to the physician when determining if the cause of a patient's hoarseness may require further investigation. In this report four patients presenting with varying degrees of hoarseness due to left RLN compression are discussed [4,5].

Ortner's Syndrome, has historically been defined as left RLN palsy presenting as hoarseness due to underlying cardiac disease. However, the most common cause of RLN palsy is lung cancer. In one study 42% of cases were attributed to lung cancer and 24% were iatrogenic, with Ortner's Syndrome included in less than 11% of patients labeled as idiopathic [4]. Overall, Ortner's Syndrome comprises only 1% to 3% of extra-laryngeal hoarseness cases [5,6].

While initially described as a consequence of mitral stenosis, numerous forms of cardiac pathology have since been found to cause Ortner's Syndrome. These include congenital abnormalities such as atrial septal defect, ventricular septal defect, and patent ductus arteriosus. Primary pulmonary hypertension, enlarged PAs, and thoracic aortic aneurysms have been found to cause RLN palsy [5,6]. In cases of RLN palsy, mitral stenosis is the underlying cause in 0.6% to 5.0% [7,8].

The pathophysiology of Ortner's Syndrome requires a thorough understanding of the RLN's path in the mediastinum. The left nerve, which is affected 1.75× more often than the right, branches off the vagus nerve as it descends near the arch of the aorta and loops under the arch, to the left of the ligamentum arteriosum. Passing between the aorta and left PA, it then travels superiorly in the tracheoesophageal groove. The nerve enters the larynx deep to the inferior cornu of the thyroid cartilage. Here, the RLN innervates all intrinsic laryngeal structures below the vocal cords, with the exception of the cricothyroid muscle. The nerve is most vulnerable to compression between the aorta and PA. In cases of mitral stenosis, the left RLN is thought to be compressed at this juncture. In contrast to the complex pathway of the left nerve, the right nerve descends and loops around the right subclavian artery [8,9].

However, why mitral stenosis specifically affects the left RLN

more often than the right is easily explained by both the unique path taken by the left RLN around the heart and the physiological sequelae of the chronic mitral valve disease. In mitral stenosis, the stenotic valve decreases the volume of blood the left atria are capable of passing to the ventricles with each contraction. Residual blood in the atria adds to incoming blood from the lungs, increasing the pressure within the pulmonary veins and capillaries. Subsequently, the right heart, normally transmitting blood against significantly lower pressures relative to the left, is now forced to pump against these increased pressures. The right heart compensates by increasing its force of contraction, again increasing the pressure exhibited on the internal wall of the heart. This includes the normally low-pressure trunk and branches of the PA. Over time, such as in chronic mitral stenosis, this increased pressure leads to thickening of the vessel-wall and dilation of the PA.

Diagnosing Ortner's Syndrome in patients with recent-onset hoarseness requires ruling out other potential causes and ordering investigative imaging when any suspicion of cardiovascular involvement is present. In young, healthy patients, hoarseness is often due to benign causes and does not require extensive workup. However, special attention should be paid to elderly patients in whom the disease most often presents. Evaluation begins with obtaining a thorough history and physical exam. Recent recommendations, put forth by the American Academy of Otolaryngology, stress that in patients with new onset hoarseness, evaluation should not be delayed greater than four weeks. Immediate laryngoscopy is recommended if there is any concern for a serious underlying condition. Direct or indirect laryngoscopy to view the vocal cord movement is recommended before pursuing invasive imaging. If no mass is found and the vocal cord is paralyzed without iatrogenic cause, CT and/or MRI scan should be performed [10]. Partial or slow compression of the recurrent nerve may not produce the typical hoarseness seen with Ortner's Syndrome. Routine vocal cord examination should be performed in patients with established heart disease [11]. Interestingly, recent literature suggests that hoarseness may not only precede prominent cardiovascular symptoms but may also be the initial symptom in rapidly expanding aortic aneurysm. These findings have led researchers to hypothesize sudden-onset Ortner's Syndrome may have predictive value in aortic aneurysm rupture [6,12]. One should consider this diagnosis when presented with a patient complaining of hoarseness in combination with a history of cardiovascular disease. Current research recommends any patient with new onset hoarseness, lasting greater than two weeks, should seek immediate evaluation to determine the etiology [5,12].

Conclusion

Ortner's Syndrome represents a unique example of cardiac pathology indirectly leading to vocal cord paresis. The involvement of these distinct anatomical systems reinforces the need for otolaryngologists to understand the pathophysiology of both laryngeal and extra-laryngeal structures. Any persistent hoarseness or voice changes in those patients with cardiovascular history should raise suspicion and be investigated accordingly.

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