CASE REPORT

Ortner's Syndrome With Primary Pulmonary Hypertension - A Case Report

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ABSTRACT

Ortner's Syndrome or Cardio-vocal syndrome is a manifestation of hoarseness secondary to left recurrent laryngeal nerve (RLN) palsy with abnormal cardiovascular components. The association of pulmonary hypertension with Ortner's Syndrome has not been widely available in the literature with very few cases reported worldwide.We present a case of a 17 years old lady with underlying Primary Pulmonary Hypertension, who presented with progressive hoarseness over the past 3 years associated with breathlessness and easy fatigue on exertion. Herein we discuss the options of treatment modality for Ortner's Syndrome.
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INTRODUCTION

Ortner Syndrome, also known as Cardio-vocal syndrome, is characterized by hoarseness resulting from left recurrent laryngeal nerve (RLN) palsy along with abnormal cardiovascular manifestations. The syndrome was first described by Nobert Ortner in 1897, in a patient with mitral stenosis (1). Although this syndrome is well-documented in association with certain cardiovascular conditions, such as mitral stenosis, there have been limited reports in the literature regarding its connection with pulmonary hypertension. In this report, we present a case of a patient who presented with hoarseness due to left RLN palsy, which was found to be associated with pulmonary hypertension.

CASE REPORT

A 17-year-old female presented with progressive hoarseness over the past 3 years, accompanied by breathlessness and easy fatigue on exertion. She had no aspiration, dysphagia, fever, chest pain, or hemoptysis history. The patient had no known medical conditions and was not taking any regular medications. There was no prior history of voice abuse, prolonged cough, or significant weight loss. Additionally, there were no head or neck trauma records, and her surgical history was unremarkable.

Clinical examination revealed a medium-built patient without any syndromic features. An auditory-perceptual evaluation using the GRBAS score showed a grade 3 hoarseness, primarily characterized by Breathiness and Asthenia. Her maximum phonation time (MPT) was recorded as 6 seconds. The patient's Voice Handicap Index (VHI-10) revealed a high score of 27, indicating a considerable impact on her voice and overall quality of life. Flexible laryngoscopy demonstrated left vocal cord palsy in a paramedian position with vocal cord bowing [FIG 1]. A cardiovascular examination revealed the presence of a diastolic murmur with a slight displacement of the apex beat. Other systemic examinations were unremarkable.

A comprehensive diagnostic evaluation was conducted to determine the underlying cause of the left vocal cord palsy. Initial investigations, including a Chest X-ray, revealed findings of cardiomegaly. Consequently, a Contrast-enhanced Computed Topography (CT) scan of the neck and thorax was performed, which revealed a dilated pulmonary trunk measuring 39mm (Normal: 30mm) exerting pressure on the left recurrent laryngeal nerve [FIG 2]. No aneurysm or mass was observed in the imaging. Subsequently, a 2D Echocardiography was conducted, showing the presence of pulmonary hypertension, along with pulmonary regurgitation and a normal ejection fraction of 68%.

Due to the long-standing hoarseness, the patient underwent laryngeal electromyography (EMG). The EMG results showed a reduction of the left thyroarytenoid muscle unit action potential compared to

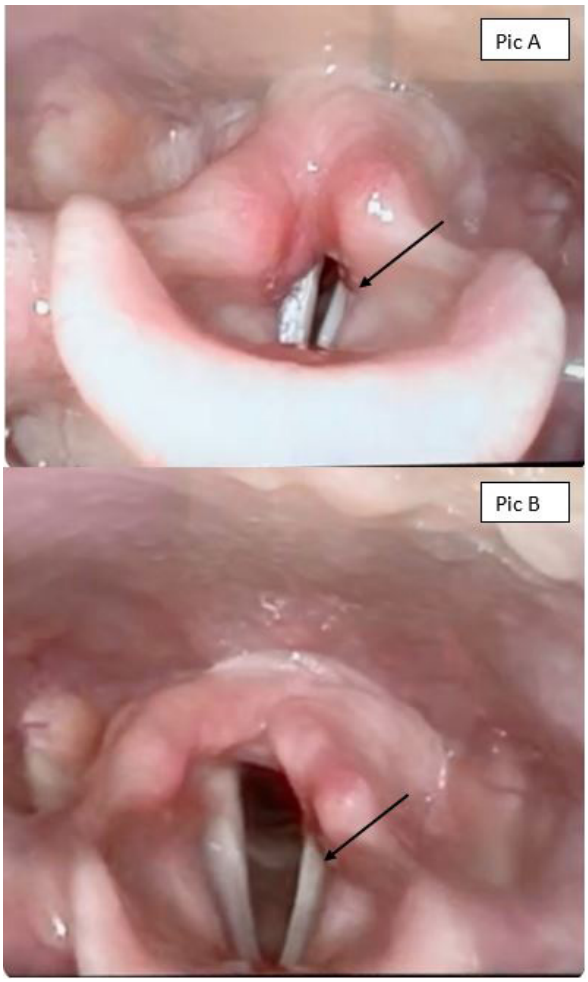

Figure 1: Flexible Laryngoscope showing paralyzed left vocal cord (Black Arrow) with minimal phonatory gap in paramedian position during Adduction (Pic A) and Abduction (Pic B)

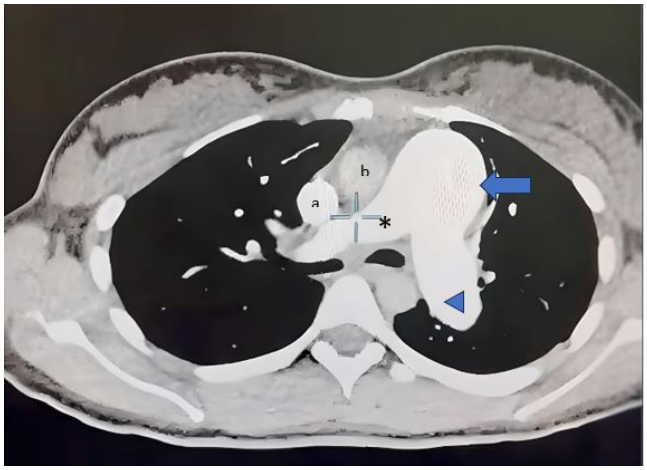

Figure 2: Contrast Enhanced Pulmonary Angiography (CTPA) showing enlarged pulmonary trunks (blue arrow), with dilated right pulmonary artery (asterisk) and left pulmonary artery (arrow head). Normal Superior Vena cava (a) and Ascending Aorta (b), appears normal.

the right thyroarytenoid, suggesting electrophysiological evidence of left recurrent laryngeal neuropathy likely secondary to her pulmonary hypertension.

The patient is currently under the care of the Cardiovascular team and has been prescribed dual anti-hypertensive therapy to improve her medical condition. Considering the significant decline in her voice quality, a planned treatment approach involving non-selective re-innervation of the recurrent laryngeal nerve is being considered once her condition is optimized. Additionally, a collaborative effort between medical and speech therapy teams will be employed to manage the patient's voice therapy, aiming to improve her overall quality of life.

DISCUSSION

Cardio Vocal Syndrome, also known as Ortner Syndrome, represents a clinical diagnosis characterized by unilateral vocal cord paralysis that arises as a secondary manifestation of cardiovascular diseases. The identifiable cardiovascular pathologies contributing to this condition encompass a range of issues, such as valvular lesions, shunt lesions, vascular causes, structural anatomical abnormalities, and pulmonary associations, such as pulmonary hypertension and thoracic aneurysm (2).

The pathophysiology of this syndrome is hypothesized to result from the compression of the left recurrent laryngeal nerve caused by the enlargement of the left atrium, which, in turn, is due to underlying cardiac conditions. In our patient, we have documented a rare case of Ortner's syndrome secondary to primary pulmonary hypertension. The postulation of this correlation between primary pulmonary hypertension and unilateral vocal cord paralysis arises from the compression of the left recurrent laryngeal nerve between the aorta and dilated pulmonary artery (3).

The management of Ortner's Syndrome is centered around addressing the underlying pathology, and it involves considering non-surgical or surgical interventions based on the clinician's assessment of the patient's voice quality. This assessment encompasses various clinical investigations, including imaging modalities, such as chest radiography, CT scan, Echocardiography (ECHO), Electrocardiography (ECG), and also assessment through auditory-perceptual evaluation using GRBAS scoring, patient self-assessment of the impact on their quality of life with tools like the Voice-Handicap Index (VHI), and direct imaging using flexible laryngoscopy, possibly complemented by laryngeal electromyography (LEMG) to aid in diagnosis and decision-making. These comprehensive evaluation methods empower the clinician to customize the most appropriate and effective treatment plan for each individual case.

The impact on the patient's quality of life plays a significant role in the decision-making process between opting for non-surgical or surgical intervention in Ortner's Syndrome. This is due to the possibility of spontaneous recovery of the recurrent laryngeal nerve (RLN), which has been observed in some cases. As a result, a period of 12-24 months of surveillance is often recommended for patients with a less severe impact on their quality of life (3) However, surgery can be considered for those individuals experiencing a higher impact on their quality of life, and show no improvement despite the observation period, surgical intervention becomes a viable option to address the condition more promptly and effectively.

There are several surgical approaches for managing Ortner's Syndrome such as vocal cord augmentation, whereby materials of augmentation such as fat or hyaluronic acid are injected into the deep portion of the thyroarytenoid until a medialization of the vocal cord is seen surgically. Another emerging technique that has garnered increasing interest is non-selective surgical reinnervation. This procedure involves the anastomosis of nearby functioning nerves, such as ansa cervicalis with the recurrent laryngeal nerve (RLN) to restore muscle bulk and tone to the affected vocal fold. The primary goal of this technique is to provide tone and bulk to the laryngeal muscle, which is the main concern in cases of unilateral vocal cord paralysis. While the restoration of vocal motion is of secondary importance, it is a beneficial outcome as well (4).

One significant drawback of non-selective reinnervation is the time required for the increased muscle tone to improve voice quality, which is typically reported to take around 4.5 months (5). Despite this drawback, non-selective reinnervation remains a viable treatment option that can be further explored and utilized.

In the case of our patient, given the considerable impact on her overall quality of life, our plan is to aim for and proceed with non-selective reinnervation once her primary pulmonary hypertension has been optimized. This approach holds the potential to improve her voice and enhance her overall well-being in the long term.

CONCLUSION

Ortner's Syndrome secondary to pulmonary hypertension is a condition where the management approach needs to be customized based on the patient's quality of life. In the context of modern medicine, exploring and implementing treatment options involving non-selective reinnervation becomes essential for addressing this syndrome effectively.

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