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Ortner's Syndrome Associated with Secondary Pulmonary Hypertension due to Chronic Obstructive Pulmonary Disease

(Running title:** Ortner's syndrome due to secondary pulmonary hypertension**)

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Abstract

Ortner's syndrome, also known as the cardiovocal syndrome, is characterized by hoarseness due to recurrent laryngeal nerve compression leading to paralysis by a dilated left atrium in mitral stenosis. Chronic pulmonary hypertension rarely may cause this syndrome and is often overlooked in the context of secondary pulmonary hypertension. We present a case of a 52-year-old man with hoarseness secondary to the paralysis of the left recurrent laryngeal nerve with severe pulmonary hypertension. The patient was a lifelong smoker with a FEV1 of 29% and a FEV1/FVC of %32 of the predicted. The mean pulmonary artery pressure was 96 mm Hg confirmed by cardiac echocardiography. The greatly dilated pulmonary artery (main pulmonary artery diameter: 3.5 cm) in this patient resulted in compression of the left recurrent laryngeal nerve and produced the syndrome.
Our patient is unique sample for Ortner's syndrome due to severe pulmonary hypertension associated with long-standing chronic obstructive lung disease. The case will contribute significantly to the literature in elucidating the etiology of Ortner's syndrome as a noteworthy paradigm highlighting the need for the awareness of this rare association. Our findings will also be a extremely useful in terms of revealing the effects of the treatment on hoarseness which is the hallmark of this syndrome.

***Keywords:** Ortner's syndrome, hoarseness, COPD, pulmonary hypertension.*

Introduction

Ortner's syndrome, also known as **cardiovocal syndrome,** is a rare condition characterized by hoarseness of voice due to compression of the left recurrent laryngeal nerve by cardiovascular structures or mediastinal pathology (1,2). This nerve runs a long course around the aortic arch, making it vulnerable to compression in cases of cardiovascular enlargement or abnormalities. While it is classically associated with left atrial enlargement in mitral valve disease, secondary pulmonary hypertension may also contribute to laryngeal nerve involvement. One of the most noteworthy but exclusively rare causes of Ortner's syndrome is pulmonary artery dilatation (3-6). This report describes a patient with Ortner's syndrome caused by enlargement of the main pulmonary artery secondary to pulmonary hypertension due to long-standing chronic obstructive lung disease introducing the mechanisms involved in such cases. Our patient will also be an important data source to reveal the etiologic causes and the possible effects of treatment in cases of Ortner syndrome due to different etiologies.

Case report

A 56-year-old man was admitted for hoarseness of three weeks and dyspnea in exertion of six weeks. He was a smoker of 40 p-y with a history of chronic obstructive pulmonary disease of eight years. Family history did not reveal any disease of significant concern. Physical examination showed signs of chronic obstructive pulmonary disease and cor pulmonale revealing a respiratory rate of 24/min, bilateral pretibial edema, prolonged expiration, rhonchus over the lower lung zones with wide splitting of second heart sound. His peripheral blood count and serum biochemistry were within normal limits. ECG displayed sinus tachycardia with a normal cardiac axis. Chest x-ray revealed signs of chronic obstructive lung disease with an enlarged heart and pulmonary arteries (Figure 1).

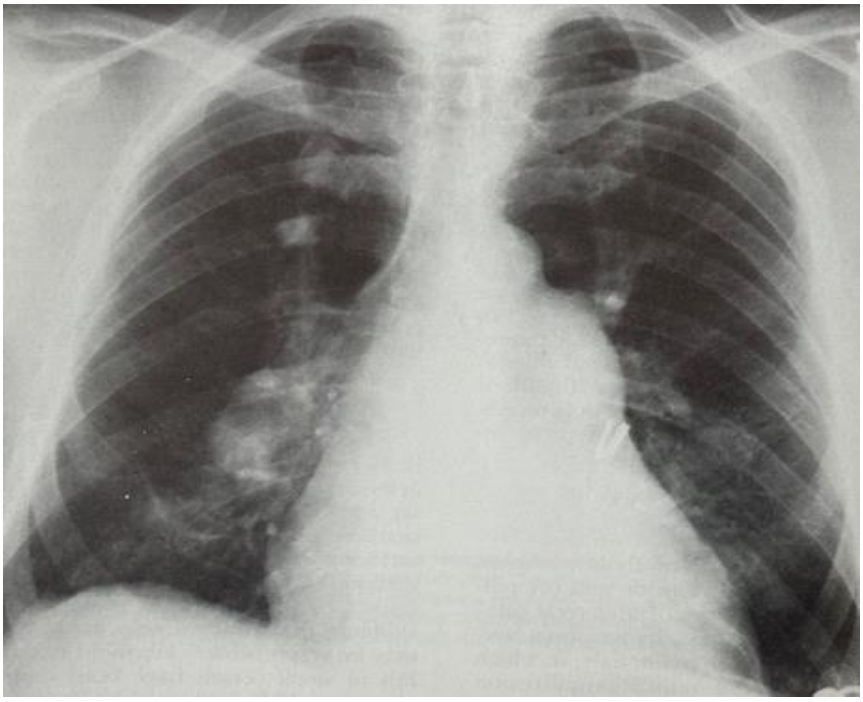

Figure 1: Chronic obstructive pulmonary disease and enlarge pulmonary arteries

Pulmonary function tests displayed an obstructive pattern with a 32% FEV_1/FVC and a 29% FEV_1 value of the predicted. ABG analysis showed a pH: 7.32, a pO_2: 48.2, and a pCO_2 value of 50.2 mm Hg revealing type II respiratory failure. Thorax CT revealed a dilated main pulmonary artery of 4.0 cm diameter compressing the left laryngeal nerve (Figure 2).

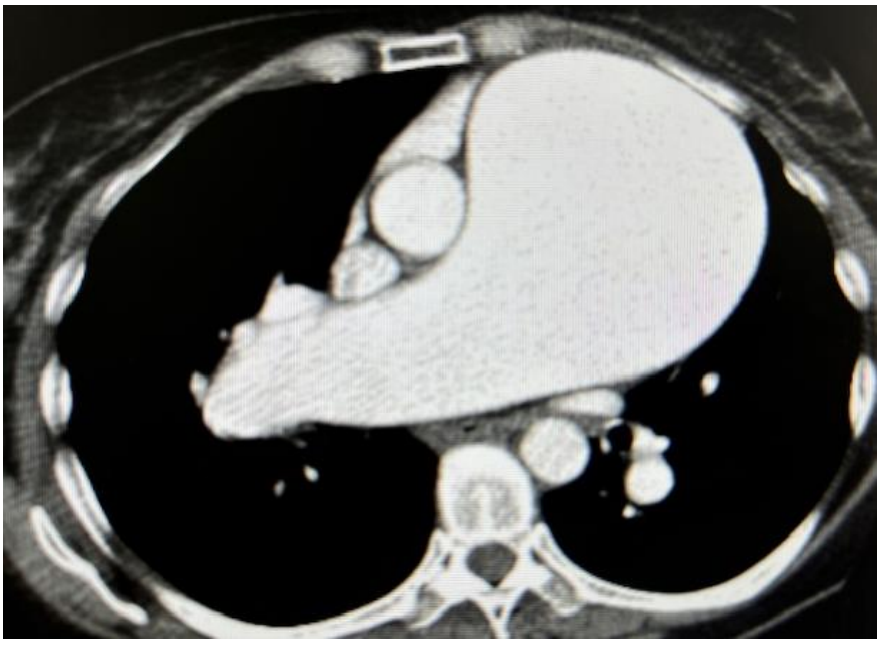

Figure 2: Chest CT revealing enlarged main and right pulmonary artery.

Echocardiography demonstrated a mean pulmonary hypertension of 46 mm Hg. Laryngosgopic examination showed left vocal cord paralysis. Right heart catheterization confirmed severe pulmonary hypertension with a mean pulmonary artery pressure of 52 mm Hg.

The patient was started on oral methylprednisolone, nasal oxygen, nebulized salbutamol, ipratropium bromide, IV furosemide, and sildenafil. Following a treatment of two weeks, the patient had a significant resolution of symptoms with a marked improvement of hoarseness while the arterial blood gases showed pO_2: 64.6 mm Hg, pCO_2: 42.8 mm Hg, and pH of 7.36. Echocardiography revealed a post treatment mean pulmonary artery pressure of 36 mm Hg. Given the history of long-standing chronic obstructive lung disease, type II respiratory failure, and the associated high pulmonary hypertension, the findings were attributed to Ortner's syndrome due to the compression of left recurrent laryngeal nerve by the enlarged main pulmonary artery caused by secondary pulmonary hypertension.

Discussion

Ortner's syndrome is a rare cardiovocal syndrome that refers to recurrent laryngeal nerve palsy from cardiovascular disease that was first described by Norbert Ortner (1). It is a rare but clinically significant condition that can result from various cardiac disorders (2,3). In this case, severe pulmonary hypertension secondary to sustained chronic obstructive pulmonary disease led to compression of the left recurrent laryngeal nerve. Management of Ortner's syndrome typically involves addressing the underlying disorders as the syndrome may be associated with many disorders that necessitate a thorough differential diagnosis (7-10). Assessment includes many diseases including left atrial myxoma, aort trauma or dissection, and haemophilia (11-14). In this patient, optimization of chronic obstructive pulmonary disease, pulmonary hypertension, and cor pulmonale treatment was initiated with bronchodilators, a diuretic, and a pulmonary vasodilator. Regular follow-up with cardiology and otolaryngology was established to monitor both cardiac status and laryngeal function.

Following treatment of the underlying chronic obstructive lung disease, pulmonary hypertension, and cor pulmonale, almost a complete improvement was achieved in all symptoms of the patient, including almost complete regression of hoarseness. As our findings suggest, hoarseness of Ortner's syndrome may reveal complete resolution following the medical treatment of the underlying disorder. If pulmonary hypertension is contributing to Ortner's syndrome, medical treatment to lower pulmonary pressures (e.g., with endothelin receptor antagonists, phosphodiesterase-5 inhibitors, or prostacyclins) can reduce the compression on the recurrent laryngeal nerve and improve hoarseness due to the elastic recoil of the main pulmonary artery. In cases where pulmonary hypertension is moderate to severe, reducing the heart's workload with medications like diuretics or pulmonary vasodilators can reduce the size of the pulmonary arteries and thereby leading to improvement of hoarseness. However, if the nerve damage is extensive or irreversible, the improvement may be limited or completely irreversible and hoarseness may persist. In patients with a long-standing **solid compression or infiltration by malignant tumor cells permenant hoarseness is inevitable due to disruption or complete interruption of nerve conduction** caused by structural abnormalities. Currently, there is not any laboratory or imaging modality available to determine the degree of damage and destruction caused by laryngeal nerve compression in Ortner's syndrome. Improvement of hoarseness can only be interpreted by observing the effect of medical treatment that may be possible in patients with vascular compression. In cases with solid compression of the laryngeal nerve, the clinical course may evolve into a more complicated outcome and may require a significant delay for remission

In this respect, the only way to determine whether hoarseness is partially or completely reversible is to observe the clinical profile of the patient that comes out after the medical or surgical treatment of the underlying disease. This evolving understanding underscores the need for high clinical suspicion of Ortner's syndrome in patients with cardiovascular disease and new-onset hoarseness, guiding timely diagnosis and management for improved outcomes. The only determining factor in the resolution of hoarseness with treatment is the pathological extent of the laryngeal nerve damage and the nerve conduction process. In cases with malignancy, reversibility of hoarseness in Ortner's syndrome appears to be much lower or unfeasible since disruption nerve conduction is usually irretrievable due to permanent structural changes that have taken place.

Conclusion

This case highlights the association between the Ortner's syndrome and secondary pulmonary hypertension associated with chronic obstructive pulmonary disease. Clinicians should be aware of this rare complication in patients with severe pulmonary hypertension, as early recognition can lead to appropriate management and potential improvement in of the laryngeal nerve function before irreversible damage occurs. The hallmark for partial or complete relief of hoarseness is the depth and extent of laryngeal nerve damage. Exact effect of the medical or surgical treatment can only be evaluated according to the patient response during the follow-up. Partial or complete reversal of hoarseness in patients with Ortner syndrome with surgical treatment is less likely due to the solid compression or malignant cellular infiltration of the nerve. On the other hand, as it is the case in our patient, medical treatment may yield complete or partial resolution response in hoarseness by reducing the pulmonary artery pressure which may may decrease the vessel compression as a results its elastic structure, thereby alleviating the compression over the laryngeal nerve.

Author's contributions

Cuneyt Tetikkurt contemplated and wrote the case report.
Muammer Bilir prepared the laboratory findings of the patient.
Halil Yanardag analyzed the imaging findings.
Seza Tetikkurt wrote the pathologic mechanism of the syndrome.

Conflicts of interest

All authors declare that they do not have any conflicts of interest associated with this study. Authors confirm that there does not exist any supporting or funding agencies for this research

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