

CASE REPORT

Ortner's Syndrome (Cardio Vocal Hoarseness): Unique, Infrequent, and Forgotten Entity in the Rural AreaNgurah Agung Reza Satria Nugraha Putra¹, I Ketut Susila²¹ General Practitioner, BaliMed Buleleng Hospital, 81119 Singaraja, Buleleng, Bali, Indonesia² Department of Internal Medicine, Division of Cardiology and Vascular Medicine, Faculty of Medicine, Universitas Pendidikan Ganesha, 81113 Singaraja, Buleleng, Bali, Indonesia**ABSTRACT**

Rheumatic fever is the main cause of mitral stenosis worldwide. Ortner's syndrome (cardio-vocal syndrome) is a rare complication of rheumatic mitral stenosis. It is caused by recurring paralysis of the left laryngeal nerve, mainly caused by mechanosuppression of the nerve from enlarged cardiovascular structures. A 76-year-old woman with chronic rheumatic heart disease (RHD) complained of hoarseness for 17 days, accompanied by shortness of breath, nausea, and vomiting for 1 week. Auscultatory examination revealed a loud first heart sound in the mitral region as well as an irregular rhythm. An electrocardiogram examination revealed right axis deviation and atrial fibrillation. Chest X-ray showed cardiomegaly. Despite normal left and right ventricular function, echocardiography showed severe mitral stenosis with mild mitral regurgitation. Conservative treatment was given with a combination of diuretics, beta-blockers, vitamin K antagonists, and angiotensin receptor blockers. An otolaryngologist was consulted, and the patient was treated conservatively.

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identification and treatment of the cardiovascular disease causing vocal cord paralysis can help restore hoarseness to normal. Furthermore, vocal cord paralysis might be misdiagnosed as other pathological conditions, masking potentially fatal cardiovascular conditions (1).

INTRODUCTION

Rheumatic fever is the most common cause of mitral stenosis worldwide, especially in developing countries. Shortness of breath, chest pain, and hemoptysis are the most common clinical manifestations of rheumatic mitral stenosis. The incidence of symptoms resulting from compression of the enlarged left atrium on adjacent structures has been minimally documented. Hoarseness caused by recurrent laryngeal nerve palsy is a rare complication of mitral stenosis. When a cardiovascular process causes vocal cord paralysis, this condition is referred to as Ortner's Syndrome (1). According to some studies, mitral valve disease is responsible for 0.6%-5% of the incidence of laryngeal recurrent nerve palsy. Prospective studies have also shown that left-sided recurrent laryngeal nerve palsy is more common than right-sided and was reported in all age groups (2,3). The incidence and prevalence of Ortner's Syndrome are rarely documented, especially in Indonesia. Scientific evidence is currently limited to case reports, and no guidelines or recommendations exist on diagnosing and managing Ortner Syndrome (3). Prompt and appropriate

CASE REPORT

A 76-year-old woman with chronic rheumatic heart disease came to the Emergency Department with complaints of shortness of breath, which had worsened since a day before admission. The patient also complained of hoarseness 17 days before admission. The patient had a history of cavities, but treatment was inadequate. The patient currently takes carvedilol 2x3.125 mg, spironolactone 1x25 mg, warfarin 1x2 mg, and furosemide 1x40 mg due to pre-existing hypertension, atrial fibrillation, and chronic heart failure. Vital signs examination revealed a blood pressure of 130/80 mmHg with other normal limit examinations. Auscultation reveals a loud diastolic murmur with an irregular beat in the mitral area (left fifth midclavicular line). Laboratory results were within normal limits. An electrocardiogram examination revealed atrial fibrillation (Figure 1), and cardiomegaly (cardiothoracic ratio 81%) was seen on chest X-ray (Figure 2). Echocardiography confirmed severe mitral stenosis and mild mitral regurgitation due to the rheumatic process,

with normal left and right ventricular function [left ventricular ejection fraction (LVEF) 55.6% and tricuspid annular plane systolic excursion (TAPSE) 27mm] (Figure 3). The patient was not diagnosed with mitral stenosis before the onset of hoarseness. Following an echocardiogram, the patient was diagnosed with mitral stenosis. The author suspects that hoarseness is linked to Ortner's syndrome.

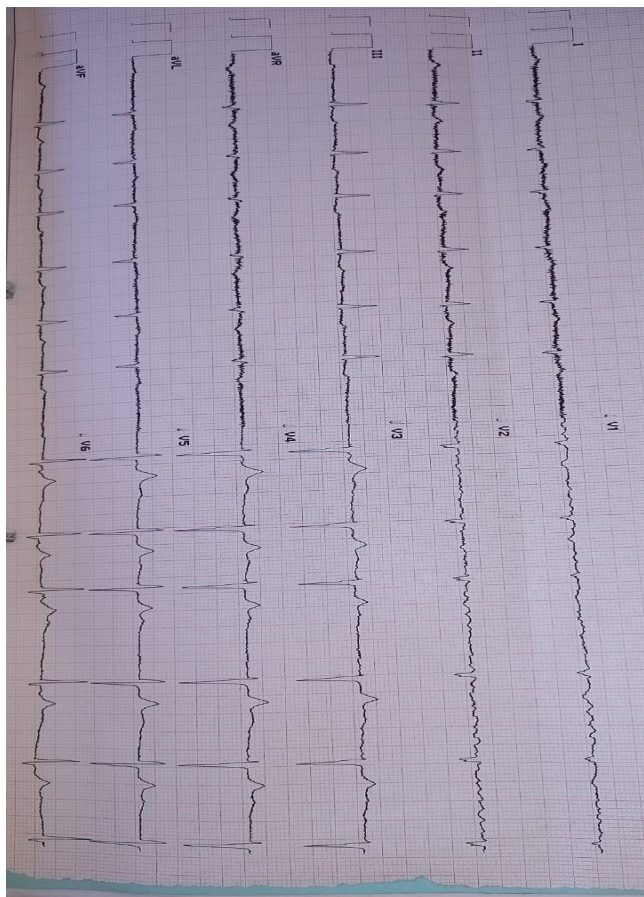


Figure 1: Electrocardiographic results of the patients revealed right axis deviation and atrial fibrillation.

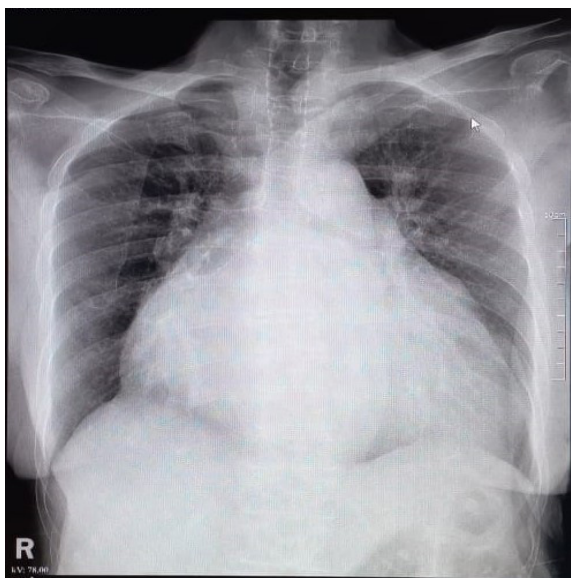


Figure 2: Chest x-ray of the patients revealed cardiomegaly (cardiothoracic ratio 81%) and congestive pulmonum.

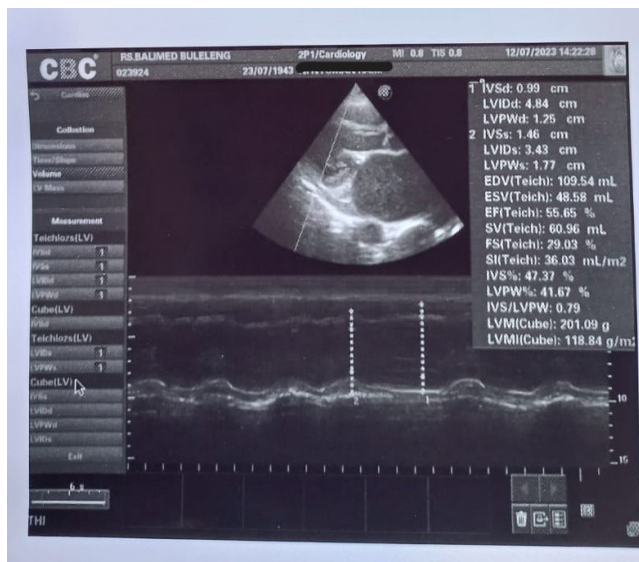


Figure 3: Echocardiographic studies confirmed severe mitral stenosis and mild mitral regurgitation due to the rheumatic process, with normal left ventricular and right ventricular function. Left ventricle ejection fraction (LVEF) is 55.6%, and tricuspid annular plane systolic excursion (TAPSE) is 27mm.

The patient was treated conservatively with spironolactone 1x25 mg intraoral, furosemide 2x20 mg intravenous (IV), carvedilol 2x6.125 mg intraoral (IO), warfarin 2x2 mg IO, and candesartan 1x8 mg IO. The patient was also referred to an otolaryngologist and conservatively treated, however, the patient's hoarseness persisted. Due to restricted facilities and infrastructure, direct endoscopic assessment of the underlying laryngotracheobronchial condition was impossible. Furthermore, mitral valve surgery was not considered due to a lack of cardiothoracic surgeons and suitable surgical facilities. As a result, the patient was planned to be referred to a hospital with more comprehensive capabilities.

DISCUSSION

Cardio-vocal syndrome (Ortner syndrome) was first reported by Nobert Ortner in three patients with severe mitral stenosis in 1897, but Stocker and Enterline published the first comprehensive description of cardio-vocal syndrome in an English-language journal in 1958. According to the NCBI, Ortner syndrome has only been described in a few cases, and it is considered rare. A 2021 NCBI literature search revealed just 76 recorded cases, despite recent papers identifying additional causes of the illness. The incidence of this syndrome linked with left atrial enlargement is estimated to be 0.6-5% (4).

Hoarseness due to this syndrome is caused by compression of the left recurrent laryngeal nerve (LRLN) by the enlarged left atrium due to mitral stenosis. The recurrent laryngeal nerve, which originates from the vagus nerve, provides an ipsilateral motor supply to the intrinsic muscles of the larynx. This nerve enters the larynx through the cricopharyngeal muscle and

innervates the intrinsic laryngeal muscles that control phonation. Hoarseness may result from dysfunction of this nerve and cause more severe symptoms when both left- and right-sided nerves are affected (1,2). While hoarseness can have many benign causes, it's a critical warning sign that should prompt further investigation. It's often the only initial symptom, making early recognition vital (3).

Although mitral stenosis was once the most prevalent cause, other reasons, such as aortic aneurysm and pulmonary hypertension, are now thought to be more common. Chronic and uncontrolled hypertension is a major risk factor for the development of aortic aneurysms (dilatations of the aorta) and aortic dissections (tears in the aortic wall). The aorta is close to the left recurrent laryngeal nerve. Aneurysms, particularly those of the aortic arch, can directly compress or stretch the nerve. Aortic dissections, which are medical emergencies, can also cause nerve compression (4). Chronic heart failure can also produce congestion and persistent lung inflammation, resulting in enlarged tracheobronchial lymph nodes in the mediastinum. This swelling can increase pressure on the LRLN. Acute decompensated heart failure can also cause the heart chamber size and pulmonary artery pressure to rise dramatically. This "dynamic dilatation" might compress the nerve abruptly, causing hoarseness (5).

In this case, chest X-ray and echocardiography examination helped establish the diagnosis in this patient. A chest X-ray examination is taken as the initial imaging examination. Furthermore, echocardiographic examination has been shown to assist in identifying any structural abnormalities of the heart, such as dilation of the left atrium and pulmonary artery. Echocardiographic examination revealed severe mitral stenosis and mild mitral regurgitation, which proved as the most important examination modality to help confirm the diagnosis of Ortner's Syndrome in addition to chest X-ray examination. If echocardiography shows significant dilatation of the left atrium and pulmonary artery, a computed tomography (CT) or magnetic resonance imaging (MRI) scan is not required. But if echocardiography results are normal, CT or MRI of the neck and chest should be performed. Regarding hoarseness, direct endoscopic examination remains the gold standard for evaluating pathological disorders in the laryngotracheobronchial area. Laryngoscopic examination in Ortner's syndrome could reveal left vocal cord paralysis with normal superficial and submucosal appearances.

When determining the differential diagnosis for Ortner syndrome, looking for other medical conditions that can produce hoarseness is important. Several conditions may cause hoarseness, such as (1):

- Local laryngeal causes: vocal polyps, laryngeal tuberculosis, smokers' nodules, and carcinoma larynx.
- Vascular or cardiac causes: aortic dissection,

pseudoaneurysm, large left atrial myxoma obstruction, congenital heart diseases, pulmonary artery enlargement, and pulmonary embolism.

- Neoplastic causes: bronchogenic carcinoma, lymphoma, esophageal carcinoma, neurogenic tumors, thyroid carcinoma, malignant thymus disease, and lymph node metastasis.
- Surgical/iatrogenic causes: heart surgery, median sternotomy, patent ductus arteriosus ligation or embolization, and more.

However, the most frequent cause that clinicians should consider when diagnosing is mitral stenosis, which can result in left atrial enlargement and pulmonary hypertension (3).

The prognosis of Ortner's syndrome strongly depends on the underlying etiology and disease duration. Symptoms can be managed with diuretics, beta-blockers, digoxin, non-dihydropyridine calcium channel blockers, and anticoagulation using vitamin K antagonists with a target INR of 2 to 3, which is in line with the guidelines of the European Society of Cardiologists (ESC) and the European Association for Cardio-Thoracic Surgery (EACTS) 2021. If the symptoms are severe and treatment is non-compliant, surgical management to tighten the vocal cords should be considered. There are case reports of significant improvement in hoarseness following mitral valve repair (MVR) surgery. If the patient's cardiovascular condition does not allow for causal treatment and the symptoms persist, voice therapy and even tightening of the affected vocal cords may be considered. In some cases, correcting the lesion with MVR helps improve voice after 3 months and should be monitored continuously (2).

The lack of comprehensive supporting examinations, such as laryngeal endoscopy and neck and chest CT scans, is a weakness in this case study, which is the exact etiology remains unknown and the definitive diagnosis of Ortner's Syndrome cannot be confirmed. Furthermore, the lack of skilled resources capable of performing surgery is a concern in this case, so the patient will be referred to a more advanced facility. We also must be aware that RHD is endemic in Indonesia, which has the world's fourth highest frequency (after China, India, and Pakistan), with 1.18 million cases reported each year. In Indonesia, the mortality rate for RHD is predicted to be 4.8 per 100,000 people. To avoid long-term problems such as Ortner's syndrome, it is critical to notice signs and symptoms of RHD as early as possible. If Ortner's syndrome is suspected, the most crucial aspect is early diagnosis of the source of laryngeal nerve palsy, because nerve damage is reversible depending on the duration of the injury.

CONCLUSION

Ortner syndrome is a rare condition that can be caused by multiple cardiopulmonary disorders. Early identification

of Ortner's syndrome allows early treatment, which can prevent vocal cord paralysis. To evaluate vocal cord paralysis, it's important to look beyond the larynx to find the cause of vocal cord paralysis in patients with hoarseness. It's also important to have regular cardiac check-ups to rule out causes of vocal cord paralysis.

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