

## Ortner's Syndrome in Rheumatic Mitral Stenosis: A Case Report

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### ABSTRACT

**Background:** Ortner's syndrome (OS), alternatively known as cardio vocal syndrome, is a disorder characterized by left recurrent laryngeal nerve palsy as a result of underlying cardiopulmonary disease. One of the most important causes is an enlarged left atrium as in case of mitral stenosis. Rheumatic heart disease is still the commonest cause of significant valvular heart disease in the developing world. Lack of health infrastructure leads to late diagnosis of rheumatic heart disease. Most of the patients present with very advanced stages of valvular heart disease.

**Case Presentation:** We present here a case of a 48-year-old female with rheumatic mitral stenosis. Due to late diagnosis, she had developed a rare complication in the form of Ortner's Syndrome.

**Conclusions:** Focused screening for acute rheumatic fever and rheumatic heart disease among school-going children along with adequate secondary prophylaxis would be of great help in early diagnosis and management of valvular and extra-valvular complications of rheumatic heart disease including Ortner's syndrome.

**Keywords:** case report; mitral stenosis; ortner's syndrome; rheumatic heart disease.

### INTRODUCTION

Ortner's syndrome (OS), alternatively known as cardiovocal syndrome, is a disorder characterized by left recurrent laryngeal nerve palsy as a result of underlying cardiopulmonary disease. The most common associated comorbidity is aortic aneurysm, followed by pulmonary hypertension, mitral stenosis, and hypertension. Patients can present with hoarseness of voice, murmur and cough with expectoration for longer durations along with exertional dyspnea.

### CASE PRESENTATION

A 48-year-old female, housewife by occupation presented to medicine outpatient department with complaints of progressive dyspnea on exertion (NYHA Class II → III) for one year, hoarseness of voice for two months, occasional palpitations, fatigue and reduced exercise tolerance. She was apparently well one year ago when she developed gradually progressive exertional dyspnea, initially

NYHA class II, which worsened to Class III over the past three months. She developed hoarseness of voice two months ago which was insidious in onset and progressively worsening. She denied history of sore throat, fever, weight loss or vocal misuse. She also complained of palpitations, intermittent, rapid, and irregular in nature. She did not give history of hemoptysis, chest pain, syncope, or peripheral edema. She also denied recent respiratory tract infection or thyroid disease.

She is a known case of rheumatic heart disease and she had undergone percutaneous trans venous mitral commissurotomy for mitral stenosis five years back. Following the procedure, she was doing fine for next four years. She was being medically managed with Metoprolol 25 mg, Warfarin 3 mg, and was on oral penicillin for secondary prophylaxis. Family history was non-contributory. She did not smoke and did not consume alcohol. On examination, general condition

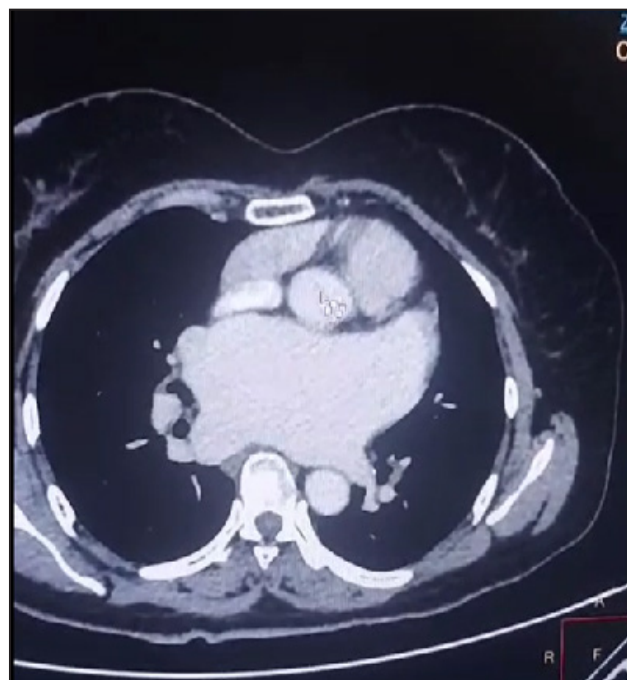
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was fair. She was average built. Pulse was 88 beats per minute, irregularly irregular and low volume. Blood pressure was 90 mmHg systolic and 70 mmHg diastolic. Respiratory rate was 16 breaths per minute. The oxygen saturation was 97% on room air. Jugular venous pressure was not raised. Hoarseness of voice was evident during conversation.

Cardiovascular System examination, on inspection parasternal heave was present, on palpation inspectatory finding was confirmed, apex beat was located in 5<sup>th</sup> ICS, in mid clavicular line. There was diastolic thrill at apex. On auscultation, first heart sound was variable in intensity, there was mid diastolic murmur which was low pitched, rough rumbling of intensity three. Pulmonary component of second heart sound was loud.

Examination of respiratory, gastrointestinal and nervous systems did not reveal any abnormalities. Neck inspection and palpation documented a normal thyroid gland, no masses, or enlarged lymph nodes. Otorhinolaryngology consultation was done for hoarseness of voice. Investigations were done. The patient underwent nasopharyngolaryngoscopy. Left pyriform sinus appeared slightly enlarged as compared to the right, with relative medialization and thickening of the aryepiglottic fold, along with slightly enlarged left laryngeal ventricle; features suggestive of left vocal cord paralysis with no signs of intrinsic laryngeal pathology or tumor (Figure 1). Non contrast and contrast enhanced CT of neck and chest revealed moderate cardiomegaly with left atrium dilatation. In view of left atrial enlargement and vocal cord palsy, mediastinal compression of left vagus and left recurrent laryngeal nerve palsy was suggested (Figure 2-4).

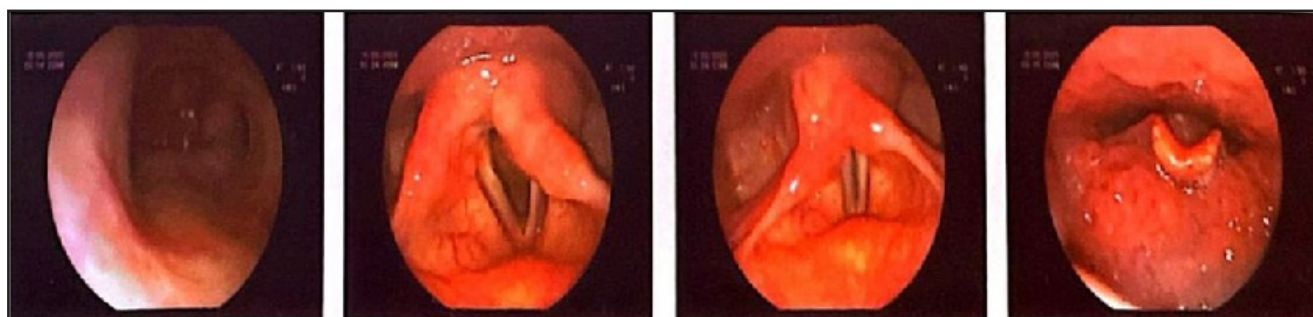
ECG demonstrated atrial fibrillation with controlled ventricular response. Chest X-ray revealed



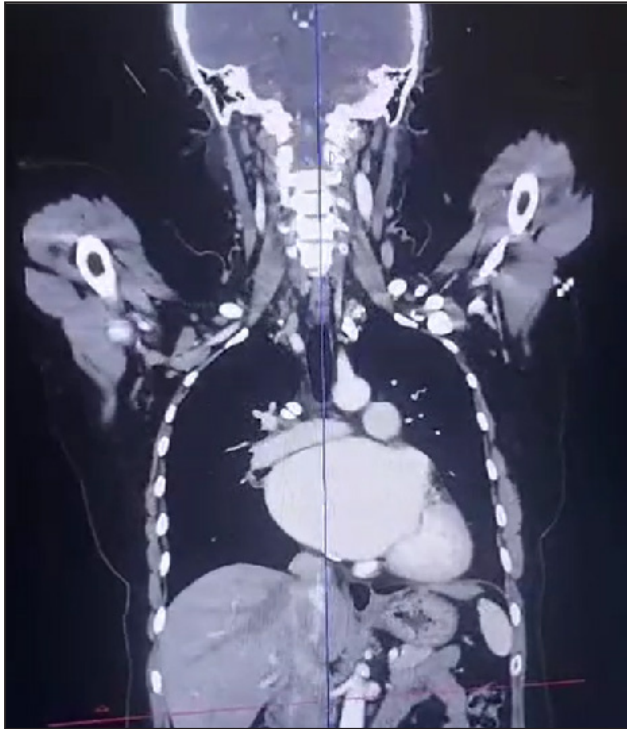
**Figure 2. Dilated Left atrium seen in axial view of CT chest.**

straightening of left heart border, double density sign suggestive of Left atrial enlargement and elevated left main bronchus. Echocardiography report showed rheumatic heart disease with status post PTMC Mitral valve area of 1.1 cm<sup>2</sup>, Moderate MR, Mild TR (TRPG=20 mmHg), Dilated LA (Left atrial dimension- 5.5 cm), Mild LV Systolic Dysfunction, LVEF=46%. She was diagnosed as a case of rheumatic heart disease with mitral stenosis with atrial fibrillation presenting with Ortner's Syndrome.

## DISCUSSION



**Figure 1. Nasopharyngolaryngoscopy revealed left vocal cord palsy.**



**Figure 3. Dilated Left Atrium seen in Coronal View in CECT of Neck and Chest.**

Ortner's syndrome (OS), alternatively known as cardiovocal syndrome, is a disorder characterized by left recurrent laryngeal nerve palsy as a result of underlying cardiopulmonary disease.<sup>1</sup> It accounts for almost 11% cases of recurrent nerve palsy.<sup>2</sup> This syndrome is named after Nobert Ortner, who initially reported the phenomenon in 1897, and the then most common cause was thought to be an enlarged left atrium due to disease such as mitral stenosis.<sup>3</sup> Nevertheless, recent cases indicate that nerve palsy may result because of the compression between the enlarged pulmonary artery and the aorta as in thoracic aortic aneurysms or pulmonary hypertension which have become the leading causes.<sup>3</sup> The most common associated comorbidity is aortic aneurysm (41%), followed by pulmonary hypertension (35%), mitral stenosis (17%), and hypertension (12%).<sup>1</sup>

The left recurrent laryngeal nerve which supplies the intrinsic laryngeal muscles originates with the ipsilateral vagus nerve at the aortic arch level, and then passes around the aorta and up the tracheoesophageal groove.<sup>4</sup> As it curves beneath the aortic arch, it traverses the aortopulmonary window



**Figure 4. CECT of Neck and Chest: Mid Sagittal view showing possibility of left recurrent laryngeal nerve compression due to left Atrium dilatation.**

and passes behind the ligamentum arteriosum.<sup>4</sup> This long course makes it vulnerable to injury from lesions affecting surrounding structures. As a result, the nerve can be compressed by cardio vascular pathologies like dilated pulmonary artery or aneurysm of the aortic arch resulting in the manifestation of Ortner's syndrome.<sup>4</sup> Patients can present with hoarseness of voice, murmur and cough with expectoration for longer durations along with exertional dyspnea.<sup>5</sup> Patients also report symptoms such as dysphagia, dysphonia and aspiration.<sup>6</sup> Least common symptoms include chest pain, bipedal edema and hemoptysis.<sup>1</sup> Examination findings often consist of murmurs associated with underlying cardiac condition, raised jugular venous pressure, left parasternal heave indicating right ventricular hypertrophy.<sup>5</sup> Patient could be malnourished and have a low body mass index.<sup>6</sup>

It is more pertinent to first eliminate other malignant and non-malignant pathologies, e.g., cervical, thoracic, and mediastinal and intrinsic pathology of



the larynx by laryngoscopy, X-ray of the chest, CT neck and chest and endoscopy before determining Ortner syndrome.<sup>7</sup> Chest X-ray Posteroanterior (PA) view may show cardiomegaly or well-defined, lobulated mediastinal lesion on the left side indicating aortic aneurysm.<sup>5</sup> Electrocardiogram may show atrial fibrillation and features of left atrial enlargement.<sup>8</sup> A nasofibrolaryngoscopy would reveal left vocal cord paralysis without any signs of intrinsic laryngeal pathology or tumor.<sup>7</sup> An assessment of structural integrity of cardiovascular system should be made using echocardiography since cardiovascular abnormalities to be the most frequent aetiology.<sup>1</sup> Multi detector CT angiography is currently the imaging modality of choice for diagnosis.<sup>9</sup> CT angiography should be done to rule out aortic dissection.<sup>10</sup>

Treatment depends on the underlying cause and may include thoracic surgery, radiation therapy, endovascular aortic repair, or appropriate guideline-directed medical therapy (GDMT) for heart failure. Surgical interventions vary based on comorbidities. Aneurysm-related OS is treated with open aortic repair or Thoracic Endovascular Aortic Repair. Congenital defects like Atrial septal defect or Ventricular septal defect are usually surgically closed. Other procedures included vocal cord medialization, thyroplasty, and laryngoplasty.<sup>11</sup> Laryngeal reinnervation is

another interesting approach to unilateral vocal fold paralysis.<sup>10</sup> The majority of cases reported on Ortner's syndrome had a conservative approach with focus on improvement of quality of voice.<sup>10</sup> Our patient is on regular follow up and is improving satisfactorily.

## CONCLUSION

Rheumatic heart disease remains the most common cause of valvular heart disease in the developing world. Mitral stenosis is the dominant valvular lesion in the majority of cases. Lack of proper screening and prompt treatment leads to very late identification of the disease, and by then, most of the patients develop complications. Hoarseness of voice in this subset of patients' needs to raise concern about the possibility of Ortner's Syndrome.

## Author's contributions:

Dr. Asraf Hussain conceptualized the study and supervised the case report. Dr. Prashant and Dr. Moniska wrote the original draft. All authors have read and approved the final manuscript.

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Patient and the patient party.

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