

Case Report

A Rare Cause of Hoarseness - Cardiovocal Syndrome

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Abstract

Vocal cord palsy presented with hoarseness due to cardiovascular pathology, is an extremely rare entity, better known as Ortner's syndrome or cardiovocal syndrome. The common underlying pathology is described as compression of the recurrent laryngeal nerve by the pulmonary artery or enlargement of the left atrium. We reported a case of undiagnosed congenital heart disease in a young woman presented to the ENT clinic for hoarseness and discuss the approach to the diagnosis. Even though hoarseness is often encountered in the ENT outpatient department, cardiovascular related hoarseness is indeed a rare findings. Partial and complete resolution of voice have been reported after treatment of the underlying cardiac pathology.

INTRODUCTION

This case report is unique as it presents a rare cause of unilateral vocal cord palsy which usually may be overlooked by most of the people. This is because when a patient coming with a chief complaint of hoarseness of voice, cardiovascular pathology as a cause of vocal cord palsy has rarely been thought as an attributing cause.

CASE PRESENTATION

20 years old-young woman, who is a non smoker presented with 3 weeks history of hoarseness of voice. Patient was not known to have any underlying medical problem. She also complained of reduced effort tolerance for the past 2 years. Fiberoptic laryngoscopy showed left vocal cord palsy (Figure 1,2). The rest of the otolaryngologic examination was normal. On auscultation, a loud systolic murmur at left sternal edge heard. No carotid bruit. Echocardiography showed pulmonary arterial and right ventricular dilation. Chest X-ray showed enlargement of the left hilum (Figure 3). Further, patient proceeded with computed tomography (CT) of the neck and thorax which showed enlarged pulmonary trunk and artery (Figure 4). A diagnosis of atrial septal defect (ASD) was made. The patient underwent surgical heart repair of the defect and post operatively was uneventful. 3 months follow-up shows improvement of her voice and endoscopic examination showed a partial medialization of the left vocal cord (Figure 5). She also has no complaint of reduced effort tolerance and no swallowing disorder.

DISCUSSION

Atrial septal defect causing Ortner's syndrome is indeed a very rare condition, and it has not many cases reported to the best of our knowledge. Hoarseness due to cardiovascular pathology is an extremely rare entity which is better known as Ortner

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- Ortner

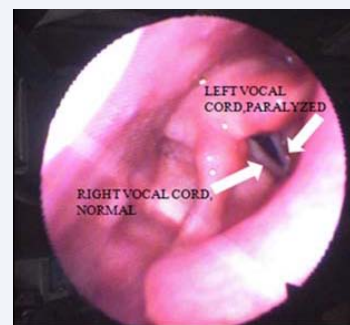


Figure 1 The paralyzed left vocal cord (arrow) in paramedian position during adduction (A) where as.



Figure 2 The paralyzed vocal cord (arrow) during abduction (B).

syndrome or cardiovocal syndrome [1]. It is first described in 1897 by Nobert Ortner, a Viennese physician in a case of mitral stenosis where he explained that hoarseness in that case was due to compression of the left recurrent laryngeal nerve by enlarged left atrium [2,4].

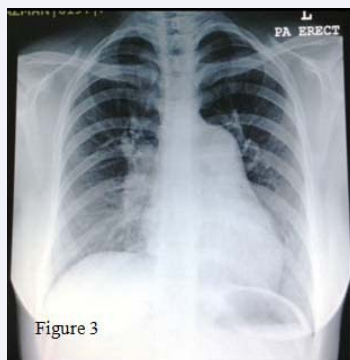


Figure 3 Chest x-ray showed enlargement of left hilum. No area of consolidation. Heart size within normal size. Prominent opacity around the left hilum.

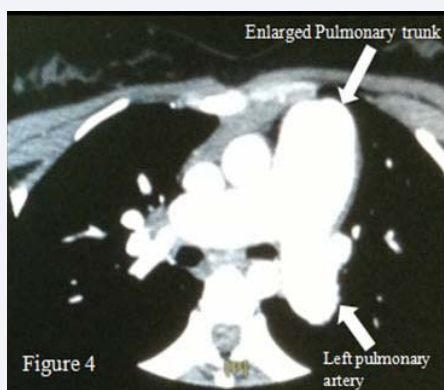


Figure 4 CT neck and thorax showed enlargement of pulmonary trunk and artery.

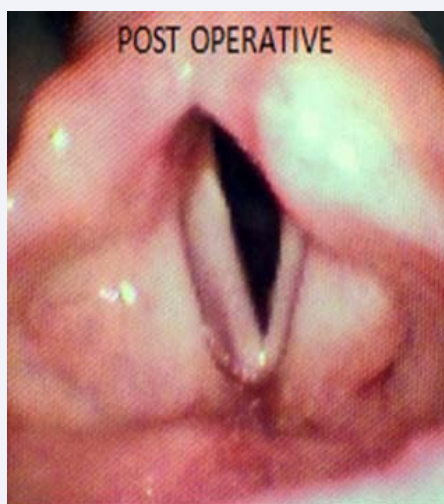


Figure 5 The partial medialization of the left vocal cord 3 months post operatively.

However, as a subject of controversy that, a variety of cardiac problem such as ischaemic heart disease, primary pulmonary hypertension, aortic aneurysm, and various congenital heart disorders can all lead to paralysis of the left recurrent laryngeal nerve [5].

Based on its anatomical pathway, the vagus nerve (the tenth cranial nerve) which further divided into the recurrent laryngeal nerves are located at its own levels for the left and right based on their anatomy course. Recurrent laryngeal neck will ascend up between esophagus and trachea to reach the neck. The right recurrent laryngeal branch hooks around the right subclavian artery. However, the left recurrent laryngeal nerve is longer in its course. Immediately when it branches out from the vagus nerve, the nerve hooks under the aortic arch and came to lie deep to the ligamentum arteriosum before ascending up to the neck [1-4]. Left side of the recurrent laryngeal nerve is more vulnerable to damage due to its anatomical course.

We have identified only eighteen cases being reported between the 1962 until 2015 by using a Pub Med and MEDLINE with keywords of 'Ortner syndrome, vocal cord palsy'. All the presenting symptoms are hoarseness and highest incidence are seen in women. Age at presentation, gender, and presenting symptoms delivered to these patients are summarized in Table 1. However, only 9 cases were included in the table because the other 9 cases were describing on the disease per se and no cases were discussed on the symptoms and signs.

This syndrome is postulated to be due to dilated pulmonary artery. Another theory that is stipulated is due to the longer course of the left recurrent laryngeal nerve, where it can be easily compressed in between the aortic arch or the ligamentum arteriosum [4] and this can be well seen from the CT images. This pathophysiology was documented in the anatomic studies of Fettroff and Norris [6] in 1911, where he contradicted Ortner's hypothesis [4]. However, there is still various theory debated about the pathogenesis of vocal cord palsy but the exact mechanism is still unknown for certain.

Hoarseness of voice may occur as an initial presenting symptom which is an early symptom of cardiac decompensation [7]. Although Ortner syndrome caused by Cardiovascular syndrome caused by idiopathic pulmonary artery hypertension and dilated pulmonary trunk has also been described in the literature.

In our case, ASD had not been diagnosed during childhood. The patient did not have any clinical symptoms related to the left recurrent laryngeal nerve for more than 20 years. This is due to a gradual development of left atrial pressure and only a passive rise in the pulmonary artery pressure. As the disease progresses, cardiac output may become fixed and eventually fall. Thus, the patient may remain asymptomatic for many years and even with the presence of shortness of breath, fatigue or reduced effort tolerance, they may lead comfortable lives, just as presented by our patient in this case. This patient gradually developed hoarseness in just three weeks time which further history and examination led to the diagnosis of ASD only her age of 20 years. This explains why patients who are diagnosed with Ortner's syndrome do not develop symptoms of heart failure despite the presence of significant enlargement of the left atrium and pulmonary artery.

The first standard imaging is usually chest radiography as it gives possible causes of an underlying condition such as mediastinal masses and if there's any presence of cardiomegaly. CT of neck and chest should be an investigation of choice for all

Table 1: This table summarized 9 cases being reported with the keyword of 'Ortner syndrome, vocal cord palsy' in PubMed/Medline search.

Cases	Age at Presentation Clinical Features Definitive Treatment	Gender	History at presentation	Underlying Syndrome
1	41	Female	Sudden onset of hoarseness	Pulmonary hypertension and Eisenmenger's syndrome
2	69	Female	Progressive hoarseness	Ductus Arteriosus Aneurysm
3	Not mentioned	Not mentioned	Not mentioned	Eisenmenger patient with severe pulmonary hypertension and dilated pulmonary artery
4	66	Female	2years history progressive hoarseness	Pulmonary hypertension caused by severe COPD
5	48	Female	Sudden onset of hoarseness for 1 month	Aortic pseudoaneurysm with underlying hypertension
6	38	Male	hoarseness and cough with expectoration of 2 months with exertional dyspnoea for the last 2 years.	Aortic stenosis with dilated left atrium and pulmonary artery
7	17	Male	hoarseness of voice of 7 months	Rheumatic heart disease (mitral stenosis with mitral regurgitation)
8	66	Male	Smoker, hoarseness for 8 months, precipitated by an attack of common cold	Saccular aneurysm arising from the arch of aorta with peripheral thrombus underlying hypertension
9	28	Female	persistent hoarseness and dyspnea on effort	pulmonary artery dilatation with underlying primary pulmonary hypertension (PPH)

patients with hoarseness, suspected vocal cord paralysis as it is important for the evaluation of the aorta, pulmonary region and mediastinum which can be difficult on plain radiographs [9]. Furthermore, it is also for determination of the site and extent of the pathology to arrive at the diagnosis and finally, guiding the possible treatment options.

The CT images in this case report showed enlarged pulmonary trunk and artery. As there is no other explanation possible for the left recurrent laryngeal nerve palsy, it is thought to be mechanically compressed. When we combined clinical symptoms as well as the radiological findings, they were consistent with the cardiovocal syndrome. Cardiothoracic surgeon was consulted, and surgical heart repair of the defect was done. Postoperatively, the patient showed improvement of voice.

Although, there were several reports by few authors that CT or MRI imaging may depict features of congenital heart disease as well as dilated pulmonary artery [1,9], the enlargement of the pulmonary trunk on CT scan was seen after the onset of left vocal cord paralysis in our patient. In keeping with the hypothesis of Stocker et al [10] in 1958, we think that these findings of enlarged pulmonary trunk and artery cause the compression of the nerve in our patient.

Therefore, arriving at its diagnosis is crucial for the treatment related. This is because the duration of injury will decide reversibility of the nerve damage.

Successful recovery will depend on the cardiovascular causes and its severity. In cases where the cardiac-related causes in a patient who are not fit for surgical treatment, other possible treatment such as surgical tightening of the affected vocal cord or voice therapy may be considered.

Considering our patient's age and good medical condition, surgical correction of the atrial septal defect was done. Subsequent follow-up of the patient revealed much improvement of the voice.

CONCLUSION

Although hoarseness of voice is often encountered in the Otolaryngology outpatient department, cardiovascular-related hoarseness or better known as Ortner syndrome is indeed an unusual presentation. Complete resolution of voice is correctable with the treatment of the underlying disease. It is of paramount importance for clinicians to consider this possibility in patients whose chief complaints are hoarseness and cardiovocal syndrome.

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