

Case Report

A rare cause for aphonia - Ortner's syndrome

Punithakumar Ramasamy^{1*}, Suresh Babu Kale¹, Senthilkumar Ramalingam²,
Murugesan Veerappa³

¹Department of Cardiothoracic and Vascular Surgery, ²Department of Cardiothoracic Anaesthesia, Meenakshi Hospital, Thanjavur, Tamil Nadu, India

³Department of Cardiology, Rohini Hospital, Thanjavur, Tamil Nadu, India

Received: 15 June 2018

Accepted: 30 July 2018

*Correspondence:

Dr. Punithakumar Ramasamy,
E-mail: drpunith84@gmail.com

Copyright: © the author(s), publisher and licensee Medip Academy. This is an open-access article distributed under the terms of the Creative Commons Attribution Non-Commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ABSTRACT

Rheumatic mitral stenosis is prevalent in the developing world and presents with a wide array of manifestations. Aphonia and hoarseness of voice secondary to recurrent laryngeal nerve paralysis (Ortner's syndrome) is an uncommon manifestation of mitral stenosis. We present a case severe mitral stenosis with moderate to severe tricuspid valve regurgitation and severe pulmonary artery hypertension that was masquerading as aphonia and hoarseness of voice for eight months. Mitral valve replacement and tricuspid annuloplasty was done and the quality of voice improved post-surgery.

Keywords: Aphonia, Mitral stenosis, Ortner's syndrome, Hoarseness, Left, Recurrent laryngeal nerve palsy

INTRODUCTION

Mitral stenosis is a valvular lesion typically seen in adults as a result of childhood rheumatic carditis. When left undetected, this valve lesion can lead to serious and potentially fatal complications. Left vocal cord paralysis and dysphagia are uncommon complications of mitral stenosis, hardly seen nowadays as intervention is advised early in its course. Mitral valve disease leading to hoarseness of voice due to left recurrent laryngeal nerve paralysis (LRLNP) was first described by Ortner et al and attributed the cause to an enlarged left atrium.¹ Hoarseness can however occur with any cardiovascular pathology and is termed as Cardiovoal Syndrome. Progression of hoarseness to aphonia is a rarity on the backdrop of severe mitral stenosis.

CASE REPORT

A thirty five years old gentleman was referred with a diagnosis of rheumatic heart disease and hoarseness of voice for further management. History of hoarseness of

voice leading to aphonia was present since 8 months for which extensive investigations including computerised tomography of the head and neck region was done. No cause could be found and speech therapy was advised with small improvement from aphonia to hoarseness. Recently, patient developed breathlessness on exertion, class II and on examination, was found to have a loud first heart sound and a mid-diastolic murmur of grade III/VI in the mitral area. Biochemical investigations were within normal limits. Chest-X ray showed dilated left atrium, cardiomegaly and enlarged pulmonary artery. Trans-thoracic echocardiogram revealed severe calcific mitral stenosis (24/14 mmHg) with moderate to severe tricuspid regurgitation, severe pulmonary artery hypertension (RVSP 103 mmHg) and good ventricular function (Figure 1). Direct laryngoscopy showed immobility of the left vocal cord (Figure 2). Intra-operatively, the posterior mitral leaflet was calcified and found plastered to the posterior wall of the left ventricle along with severe sub-valvar thickening (Figure 3). Mitral valve was replaced with number 29 sized TTK Chitra mechanical valve and tricuspid annuloplasty

performed with 32 size Medtronic contour 3D annuloplasty ring (Minneapolis, Minnesota). The perioperative course was uneventful. The patient was discharged on day 7 with normal prosthetic function. The hoarseness of voice had improved significantly on his 3 month follow-up.

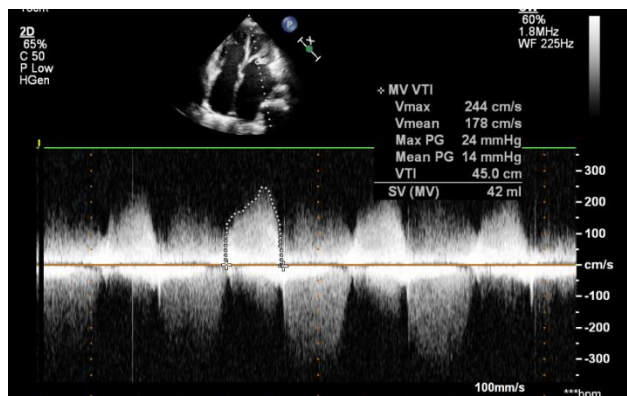


Figure 1: Transthoracic echocardiogram showing significant mitral stenosis.

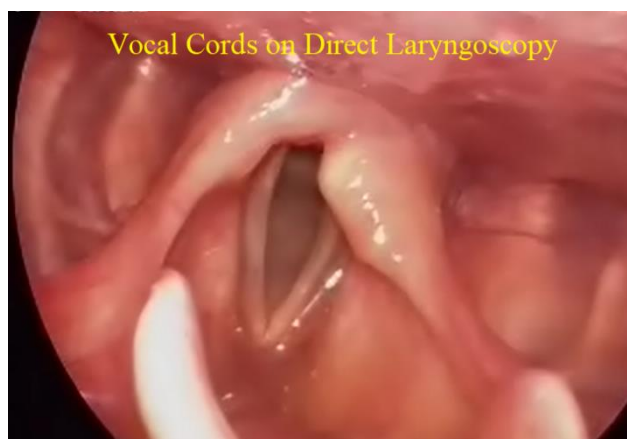


Figure 2: Direct laryngoscopy for vocal cord assessment.

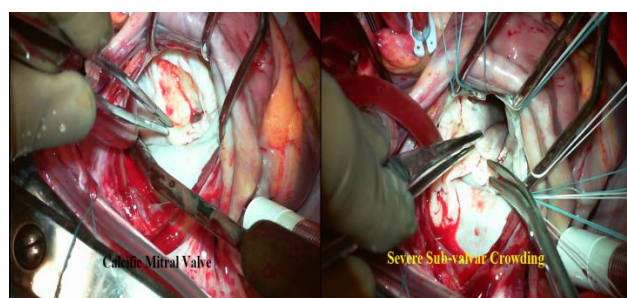


Figure 3: Operative image showing calcific posterior mitral leaflet and severe sub-valvar thickening.

DISCUSSION

Hoarseness of voice can occur in a variety of conditions including inflammatory or irritant causes, neoplasia,

neuromuscular and systemic diseases and psychiatric disorders.² Mitral valve disease leading to hoarseness and aphonia is rare and seen in 0.25 per cent to 0.5 per cent of total cases of LRLNP.³ Recurrent laryngeal nerve (RLN) is a terminal branch of the vagus nerve that innervates the larynx with a different course on each side. On the right side, it crosses the first part of the subclavian artery and hooks around to travel between the trachea and oesophagus. On the left, it arises from the left vagal trunk in the thorax when it crosses the arch of aorta, hooks around the ligamentum arteriosum and ascends in the groove between the trachea and the oesophagus. The RLN supplies all the muscles of the larynx except cricothyroid, as well as sensory supply to the larynx below the vocal cords and the upper part of trachea.

Vocal cord paralysis appears to be caused by compression of the LRLN between the enlarged hypertensive pulmonary artery, the aorta, and the ligamentum arteriosum and not by dilatation of the left atrium, as some observers have thought.⁴ The compression of the nerve and subsequent palsy is now said to occur between the aorta and pulmonary artery with the aorta playing an important role in the biomechanical environment of the RLN.^{5,6} The onset of hoarseness is usually insidious, may be intermittent and in due course of time may progress to complete aphonia. LRLNP is more common than the right RLN palsy and causes the left vocal cord to be in the paramedian position. At times the position can be variable on laryngoscopy. Symptoms include hoarseness, dysphagia, and shortness of breath during speech. The shortness of breath may lead to reduction in pulmonary function secondary to loss of natural positive end expiratory pressure that occurs with normal glottic closure. At times, effective cough cannot be mounted and the patient is at significant risk for aspiration as the paralysed vocal cord cannot protect from aspiration, especially liquids. The degree of symptoms depends on the extent of paresis and the compensation by the other vocal cord.

Most of the patients with mitral stenosis have gradual development of left atrial hypertension, a large, compliant left atrium, and only a passive rise in pulmonary-artery-pressure, a rise commensurate with the increase in left atrial pressure. As the disease progresses their cardiac output may become fixed and eventually fall. The large, compliant left atrium and the low cardiac output may serve to prevent a sudden rise in the pulmonary venous pressure and protect them from paroxysmal dyspnoea and pulmonary oedema. Such patients may remain asymptomatic for many years and even after the appearance of symptoms, generally shortness of breath and fatigue, may lead comfortable lives. Their degree of disability may be slight. This also explains patients with Ortner's syndrome not developing symptoms of heart failure despite significant enlargement of the left atrium or pulmonary artery. Although cardiovocal syndrome was initially described in the setting of mitral stenosis it has since been associated with

a broader list of cardiovascular pathology in adults including: atrial septal defect, patent ductus arteriosus, Eisenmenger's syndrome, aortic aneurysm, primary pulmonary hypertension, mitral regurgitation and congenital heart disease.^{4,7-10}

To conclude, clinical recognition of hoarseness in patients with cardiovascular disease is important and prompt referral be made for laryngoscopy and confirmation of LRLN palsy. Cardiac screening is important in patients presenting with hoarseness to rule out treatable causes and avoid psychological trauma.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

REFERENCES

1. Ortner N. Recurrenslahmung bei mitral stenose. Wien Klin Wochenschr. 1897;10:753-5.
2. Feierabend RH, Shahram MN. Hoarseness in adults. Am Fam Physician. 2009;80:363-70.
3. Schiefley CM, Smith HL. Mitral stenosis and paralysis of recurrent laryngeal nerve. Minnesota Med. 1942;25:362.
4. Vahabzadeh-Hagh AM, Yim C, Fitter J, Chhetri DK. Atrial Septal Defect and Left Recurrent Laryngeal Nerve Paralysis: A Case of Ortner's Syndrome and Literature Review. Int J Otolaryngol Head Neck Surg. 2015;4:61-5.
5. Mulpuru SK, Vasavada BC, Punukollu GK, Patel AG. Cardiovascular syndrome: a systematic review. Heart Lung Circ. 2008;17:1-4.
6. Williams MJ, Ayyalasomayajula A, Behkam R, Bierhals AJ, Jacobs ME, et al. A computational study of the role of the aortic arch in idiopathic unilateral vocal-fold paralysis. J Appl Physiol. 2014;118:465-474.
7. Nakao M, Sawayama T, Samukawa M, Mitake H, Nezu S, et al. Left recurrent laryngeal nerve palsy associated with primary pulmonary hypertension and patent ductus arteriosus. J Am Coll Cardiol. 1985;5:788-92.
8. Verbeke X, Vliebergh J, Sauer M, Leys M. Hoarseness revealing Ortner's syndrome. Acta Clin Belg. 2015;70:230.
9. Pathirana U, Kularatne S, Handagala S, Ranasinghe G, Samarasinghe R. Ortner's syndrome presenting as thoracic aortic aneurysm mimicking thoracic malignancy: a case report. J Med Case Rep. 2015;9:147.
10. Kumar Das S, Ghosh I, Das A, Kumar Dey S. Mitral regurgitation: A rare cause of Ortner's syndrome. J Clin Diagn Res. 2012;6:99-100.

Cite this article as: Ramasamy P, Kale SB, Ramalingam S, Veerappa M. A rare cause for aphonia - Ortner's syndrome. Int J Otorhinolaryngol Head Neck Surg 2018;4:1321-3.