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A Rare Case of Primary Pulmonary Hypertension with Cardiovocal Syndrome

KEYWORDS: primary pulmonary hypertension, hoarseness of voice, cardiovocal syndrome.

Dr. Putta Rajasekhar
Professor of Medicine,Government General Hospital, Kurnool.

Dr. Pothula Ramarao
Assistant Professor of Medicine, Government General Hospital, Kurnool.

Dr. Prathap Bingi
Resident in General Medicine, Government General Hospital, Kurnool.

Dr. Chakra Pani
Resident in general Medicine, Government General hospital ,Kurnol.

ABSTRACT *Pulmonary hypertension, an abnormal elevation in pulmonary arterial pressure, may be the result of left heart failure, pulmonary parenchymal or vascular disease, thromboembolism or combination of these or an idiopathic one which will be termed as primary pulmonary hypertension. This is more common in females presenting mostly as exertional dyspnea. However hoarseness of voice is a rare presentation due to compression of left recurrent laryngeal nerve between the aorta and the dilated left pulmonary artery. Here we are presenting a case of 35 year old female with primary pulmonary hypertension and hoarseness of voice which is called the cardiovocal syndrome.*

Introduction :
Patients with pulmonary hypertension usually presents with exertional dyspnea. Hoarseness of voice is due to recurrent laryngeal nerve palsy, commonly described in patients with severe mitral valve stenosis. Hoarseness caused by damage to the recurrent laryngeal nerve as a result of cardiovascular causes is known as cardiovocal syndrome (Ortner's syndrome). This rare syndrome was first described by Nobert Ortner, an Austrian physician in 1897 in a patient with mitral stenosis and hence called by the name Ortner's syndrome[1]. Hoarseness as a result of cardiovascular causes other than mitral stenosis is called as cardiovocal syndrome.

Prevalence of this syndrome is 0.6 to 5% in patients with mitral stenosis[2] and is much less in other cardiovascular diseases. Here we are describing a patient who presented to us with complaints of hoarseness of voice, dyspnea and was evaluated and subsequently diagnosed as a case of cardiovocal syndrome due to primary pulmonary hypertension.

Case report:
A 35 year old female patient presented to us with complaints of exertional dyspnea, hoarseness of voice, swelling of feet, choking spells for past 9 months , which were increased in severity for the last 20 days. She also had occasional palpitations and denied any history of fever, cough or hemoptysis. There was no significant past history in terms of rheumatic fever or bronchial asthma. She is not a smoker nor an alcoholic. She is neither hypertensive nor diabetic. On examination she was conscious , coherent with hoarse voice ,pulse rate of 90 per minute which is regular and blood pressure in right upper limb is 116/72 mm of Hg. Jugular venous pressure was raised and pedal edema extending upto ankles was present. There are no signs of pallor , icterus, cyanosis, clubbing or lymphadenopathy.

Precordial examination revealed apex beat in left 5th intercostal space 2 cm lateral to the midclavicular line which is tapping in character. Left parasternal heave was present with a palpable second heart sound. On auscultation both S1 and S2 were heard with a loud P2. A pansystolic murmur of grade 4/6 was heard in tricuspid area with a positive Carvello's sign. Initial routine work up of patient revealed normal blood counts, renal and liver function and electrolytes. Chest X-ray showed cardiomegaly with right ventricular apex and a dilated pulmonary artery(figure1). Electrocardiogram showed right axis deviation with a P pulmonale and right ventricular hypertrophy with strain. 2D echocardiogram revealed massively dilated right chambers with severe Tricuspid regurgitation and pulmonary arterial hypertension with a normal biventricular function(figure2). Mitral valve apparatus was normal. Indirect laryngoscopy showed left vocal cord palsy and fibreoptic laryngoscopy confirmed the paralysis of the left vocal cord in paramedian position(figure 3). Investigative work up for the cause of pulmonary hypertension as per the protocol was done and no underlying disorder was found. Computed tompgraphy of chest showed dilated pulmonary trunk and arteries suggestive of pulmonary hypertension. Dilated right atrium and ventricle were noted. CT pulmonary angiography revealed no evidence of thromboembolism(figure 4). Her pulmonary function tests were normal and she was non reactive for retroviral status with a normal thyroid status. ANA was negative in this patient and so autoimmune etiology was ruled out. With all the above work up patient was labeled as a case of pulmonary hypertension with cardiovocal syndrome.

Figure 1: Chest X-ray showed cardiomegaly with right

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ventricular apex and a dilated pulmonary artery

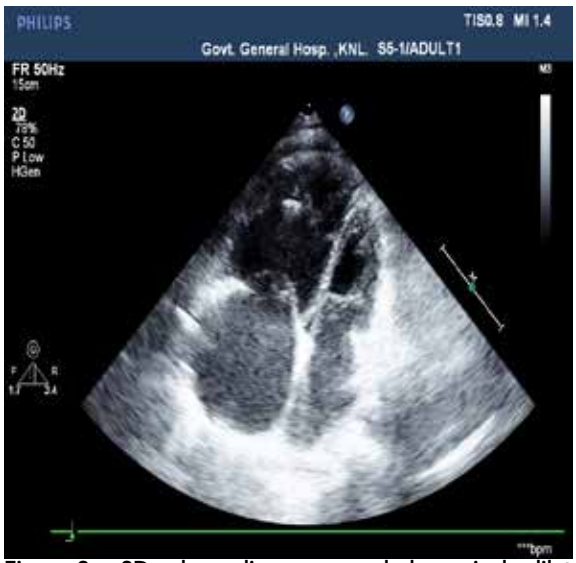

Figure 2: . 2D echocardiogram revealed massively dilated right chambers

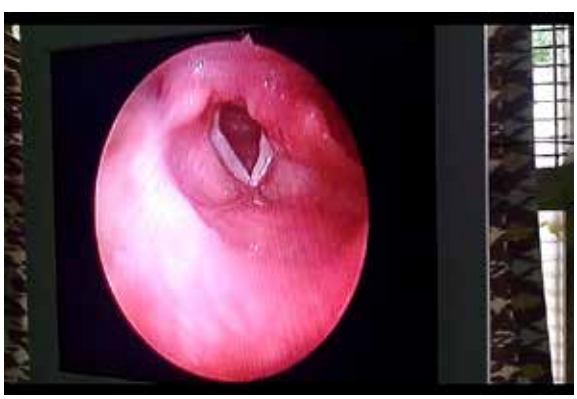

Figure 3: fibreoptic laryngoscopy showing paralysis of left vocal cord in paramedian position

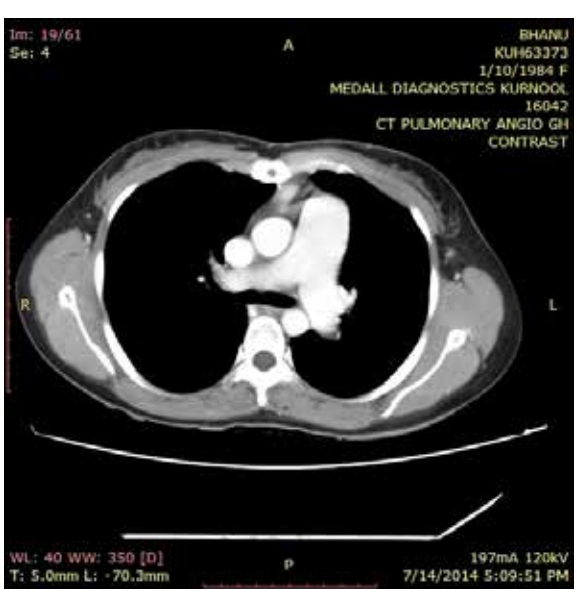

Figure 4: CT pulmonary angiogram showing dilated pulmonary vessels and no evidence of thromboembolism.

Discussion:

Cardiovocal syndrome is a clinical entity manifested by hoarseness of voice caused by an impaired ability of left recurrent laryngeal nerve to transmit impulses to laryngeal muscles because of stretching or impingement of nerve from disease induced changes in cardiac or great vessel anatomy. Ortner first described this syndrome in a patient with mitral stenosis and postulated that left atrial dilation was the cause of nerve palsy by compression against the arch of aorta[3]. On the basis of autopsy studies Fetterrolf and Norris showed that the distance between the aorta and pulmonary artery within the aortic window is only 4 mm and suggested that compression of the nerve between the two structures is responsible for palsy[4]. A variety of cardiovascular causes apart from mitral stenosis like thoracic aortic aneurysms, patent ductus arteriosus, atrial septal defect, ventricular septal defect, pulmonary hypertension and recurrent pulmonary embolism can also cause this syndrome.

Recently many theories were described such as lymphadenitis and scarring in aortic window causing nerve fixation[6], pressure from left bronchus, right ventricular hypertrophy, pulmonary artery atherosclerosis, anatomical position of ligamentum arteriosum[7] or dilated pulmonary artery [8] as seen in our case may cause this syndrome.

Anatomically vagus nerve gives off the left recurrent laryngeal nerve at the aortic arch. This nerve hooks around the lateral aspect of ligamentum arteriosum, passes up the right side of aortic arch and ascends in the groove between esophagus and trachea. This lengthy course of the nerve in the thorax makes it vulnerable to compression, traction and erosion by enlarged or displaced cardiac chambers and dilated great vessels [9,10].

Most recently it is suggested that compression of nerve between pulmonary artery and the aorta is a constant factor in almost all cases of cardiovocal syndrome. Pulmonary artery pressure was raised and the palsy may be relieved by mitral valve surgery or an intervention that decreases pulmonary artery pressure.

Pulmonary hypertension is a well known entity presenting commonly with dyspnea(80%), fatigue(19%), and syncope(13%). If the cause is unexplained then it is primary PAH. The right ventricle responds to an increased right ventricular systolic pressure to preserve cardiac output. In chronic cases progressive remodeling of pulmonary vasculature takes place which can sustain or promote pulmonary hypertension even if the initiating factor is removed. Physical examination of the patient typically reveals raised JVP, decreased carotid pulse and a palpable right ventricular impulse and there is a loud pulmonary component of second heart sound with a right sided fourth heart sound and features of tricuspid regurgitation in late stages. Whenever there is a suspicion of pulmonary hypertension the workup of the patient should be as follows.

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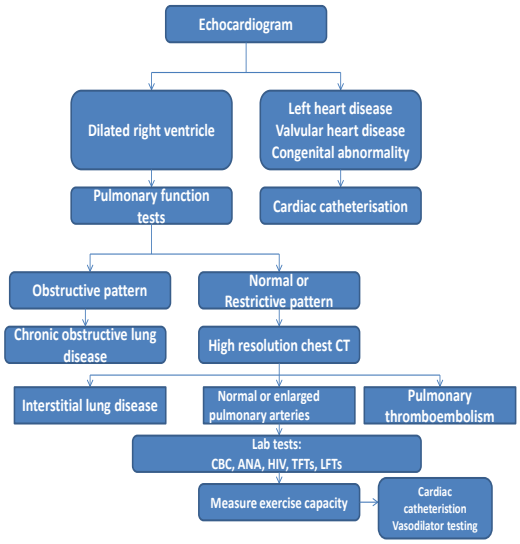

In our patient all the necessary investigations as the above flowchart revealed no significant underlying cause for pulmonary hypertension. The hoarseness of voice is attributed to the compression of recurrent laryngeal nerve between the aorta and dilated pulmonary artery. Our patient was treated with diuretics, calcium channel blockers and warfarin. She was under follow up with symptomatic improvement.

Conclusion:

Hoarseness of voice due to pulmonary hypertension is very rarely reported. Measures to decrease pulmonary artery pressure when initiated early are prompt in improving the symptoms. A high index of suspicion is needed to make a early diagnosis which can improve the overall prognosis of symptoms and the underlying cardiovascular disease.

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