Case Report

Ortner’s Syndrome as an uncommon presentation of Idiopathic Pulmonary Arterial Hypertension (IPAH)

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Abstract: More than a century ago a cardiovocal syndrome was described by Ortner where he attributed a case of left vocal fold immobility to compression of the recurrent laryngeal nerve by a dilated left atrium in a patient with mitral valve stenosis. Currently the term Ortner’s syndrome has come to encompass any nonmalignant, cardiac, intrathoracic process that results in embarrassment of either recurrent laryngeal nerve-usually by stretching, pulling, or compression; thereby causing vocal fold paralysis. Not surprisingly, the left recurrent laryngeal nerve with its longer course around the aortic arch is more frequently involved than the right nerve, which passes around the subclavian artery. Here we discuss Ortner’s syndrome as an uncommon presentation of idiopathic pulmonary arterial hypertension.

Introduction

Hoarseness of voice due to left recurrent laryngeal nerve paralysis was first described in 1897 by Norbert Ortner, an Austrian physician, in a patient with mitral valve disease (mitral stenosis and left atrial enlargement). Various cardiopulmonary conditions associated with left recurrent laryngeal nerve palsy have been described, over the last 100 years. Thus, the syndrome is now also termed as cardiovocal syndrome.

Here we report a case of Ortner’s syndrome occurring due to idiopathic pulmonary arterial hypertension (IPAH). The findings of our review of literature relating to cardiovascular disorders affecting the recurrent laryngeal nerve and the pathogenesis of hoarseness are also discussed.

Case report

A 24 year old unmarried female was admitted with a 12 month history of progressive dyspnoea, cough, fatigue, and weight loss. She had noticed progressive hoarseness of voice 3 months duration. She had not had chest pain, haemoptysis, or ankle swelling. She had no previous history of rheumatic valvulopathy, connective tissue disorders or chronic lung disease. She had no past history to suggest any provoked or unprovoked deep vein thrombosis or pulmonary embolism features. She denied any past or current use of recreational drugs or weight reduction medications.

On examination, she had a regular pulse. Blood pressure was 100/80 mmHg. JVP was 10cm, with prominent CV waves. On palpation there was a prominent right ventricular heave and palpable pulmonary artery impulse. On auscultation there was loud P₂ and a pulmonary flow murmur. Signs of tricuspid regurgitation were present, with a pulsating liver and shifting dullness indicating ascites. Respiratory examination was not suggestive of apical lung pathology. The neurological examination revealed no features of Horner’s syndrome.

Her pulse oximetry was low at 83 % saturation on room air with arterial hypoxaemia confirmed by blood gas analysis. Her Chest x-ray showed enlargement of the heart and main pulmonary arteries with peripheral oligaemia of both lung fields but had no signs of left atrial enlargement (Figure 01). The electrocardiogram (ECG) showed right axis deviation, right bundle branch block and right atrial and right ventricular hypertrophy (Figure 02). The left heart was normal on echocardiography with normal mitral and aortic valves. Her right ventricle was dilated, with septal paradox and evidence of RV volume overload (Figure 03).

She had severe tricuspid regurgitation & pulmonary regurgitation with increased right ventricular pressures and pulmonary arterial pressures. There were no cardiac shunts. Direct laryngoscopy confirmed left vocal cord palsy.
Bronchoscopy studies were negative. Routine haematology, autoantibody profile and inflammatory markers were unremarkable. Liver function tests were slightly abnormal with moderate elevation of transaminases. Abdominal ultrasound confirmed moderate ascites and dilated hepatic veins. There were no space occurring lesions.

Right heart catheterization confirmed severe pulmonary arterial hypertension without any intra cardiac shunts. CT Pulmonary angiography showed dilated central pulmonary arteries, with peripheral pruning (Figure 04). Further contrast CT chest excluded any tumoral compression or aortic arch pathology and clearly demonstrated the compression nature of the left recurrent laryngeal nerve by the grossly dilated pulmonary trunk.

**Figure 1-** Chest X ray peripheral oligemia/ reduced vascular marking of underline pulmonary hypertension.

**Figure 2-** ECG showing tall P waves of underline pulmonary hypertension.

**Figure 3-** 2D echo showing severe RA, RV volume overload.

**Figure 4-** CT Pulmonary Angiography (CTPA) showing grossly dilated Main Pulmonary Artery without any obvious paranchymal/interstitial lung disease.

**Discussion**

The causes of recurrent laryngeal nerve paralysis have been classified as non-surgical paralysis, surgical paralysis (thyroid/oesophageal operations and intubation) or a combination of the two (1).

In 1897, Ortner described a series of 3 cases of mitral stenosis with concomitant hoarseness of voice because of left recurrent laryngeal nerve palsy. The cause was attributed to compression of the left recurrent laryngeal nerve by an enlarged left atrium (2).

Since then various authors have recorded their experiences of recurrent laryngeal nerve involvement in various cardiac disorders such as Eisenmenger complex (3), left ventricular failure (4), atrial septal defect (5), patent ductus arteriosus (PDA) (6), primary pulmonary hypertension
ventricular aneurysm (11), mitral regurgitation (12), atrial myxoma (13), left ventricular aneurysm (14), cor pulmonale (15) and various types of aortic aneurysms (16,17,18,19,20,21).

Left recurrent laryngeal nerve palsy has occasionally been reported in patients with primary pulmonary hypertension (22,23). Patients with primary pulmonary hypertension and chronic pulmonary embolization, unlike those with mitral valve disease, do not have enlarged left atria. Compression of the recurrent laryngeal nerve by this structure cannot be implicated in the etiology of the palsy.

In our patient compression of the nerve between two high pressure vessels, aorta and pulmonary artery, seems more likely, and the time course of the onset of the hoarseness of voice supports such a causal relation. These case reports also support the view that the nerve palsy in Ortner’s syndrome is caused by compression of the nerve by the pulmonary artery and not merely by the left atrium. Recognition that an association may occur between recurrent laryngeal nerve palsy and pulmonary hypertension, for reasons other than those of mitral disease, may save such patients from unnecessary investigations.

Conclusion

Ortner’s syndrome is a cardio vocal syndrome which is a rare condition. It may be secondary to many cardiopulmonary disorders. Pulmonary hypertension or any other cause leading to dilatation and increased tension of the pulmonary artery result in left recurrent laryngeal nerve being compressed between the aorta and the tense pulmonary artery. This case highlights an uncommon presentation of Ortner’s syndrome due to Idiopathic pulmonary arterial hypertension in the absence of mitral stenosis (which is the originally described etiology of ortners syndrome).

Consent

Informed written consent was obtained from the patient for publication of this case report and any accompanying images.

References

have increasingly used duct occluders[5] for small sized defects, whilst large defects have been closed with Amplatzer septal occluder [3] and Amplatzer VSD devices[6].

Conclusion
Our case highlights the usefulness of Amplatzer type septal occluder when a device with a larger retention skirt for a given waist is required.