Hoarseness of voice as presenting complaint of idiopathic pulmonary arterial hypertension

Sir,

Pulmonary arterial hypertension (PAH) has a varied way of presentations, the most common of which is dyspnea that is initially exertional. Hoarseness of voice as initial and sole presenting feature of a patient of PAH is very rare and until now not reported widely. Patients with idiopathic PAH usually present with nonspecific complaints of fatigue, weakness, breathlessness, or chest pain. Rarely, these patients may present with hoarseness of voice as their initial complaint. This condition is described as Ortner’s syndrome or the cardiovocal syndrome. It occurs due to the anatomical disadvantage that the left recurrent laryngeal nerve (RLN) has, by lying between the pulmonary artery and aorta, predisposing it to stretch or trapping upon enlargement of these structures. We are presenting a case of a 23-year-old female with hoarseness of voice as the initial presentation of idiopathic PAH due to compression of the left RLN between the enlarged pulmonary artery and aorta. This case highlights the fact that PAH should be kept in the differential diagnosis of hoarseness of voice, even in the absence of the usual signs and symptoms characteristic of the disease.

A 23-year-old homemaker presented to us with hoarseness of voice for 2 months. There were no other respiratory complaints, no history of upper respiratory tract infection, smoking, vocal abuse, trauma, or any previous surgery.

General physical examination was not significant. Bilateral vesicular breath sounds were heard over the chest. On cardiovascular system examination, right parasternal heave was present, and a pansystolic murmur was heard at the tricuspid area.

The routine blood investigations, including blood counts, liver, and kidney functions were within normal limits. Chest radiograph [Figure 1] was suggestive of cardiomegaly with prominent pulmonary conus. The electrocardiograph showed right axis deviation, right ventricular hypertrophy, and poor progression of R waves. This was followed by a two-dimensional echocardiography, which revealed dilatation of the right atria and ventricle, right ventricular wall motion abnormality with severe PAH (mean pulmonary artery pressure (PAP) ~50 mmHg, pulmonary artery systolic pressure = 74 mmHg and tricuspid regurgitant jet
velocity = 4.0 m/s. There was no evidence of atrial septal defect on contrast study.

To search for a cause for pulmonary hypertension (PH), a computed tomography (CT) thorax [Figure 2] was performed which showed no lung parenchymal abnormality and dilated pulmonary trunk. Ultrasonography of abdomen was normal. Blood investigations for human immunodeficiency virus (HIV), thyroid profile, and extractable nuclear antigen (ENA) for connective tissue disorders were also within normal limits.

Hence, the patient was diagnosed to have idiopathic PAH, and the hoarseness of voice was explained by Ortner's syndrome.

Baseline functional status was established by the 6 min walk test. Walk distance was 322 m, and the patient did not desaturate during the test. She was started on tadalafil 40 mg once daily with potassium-sparing loop diuretics. She is in monthly follow-up at our institute, and there has been an improvement in voice.

PH is defined as an increase in mean PAP ≥25 mmHg at rest as assessed by the right heart catheterization.\(^1\) PAH (Group 1 PH) includes heterogeneous conditions that have similar clinical, hemodynamic profiles, and pathological changes. According to the Dana Point (2008) classification of PH,\(^2\) PAH includes idiopathic PAH, heritable, drug-induced, and associated with PAH groups.

PAH is a rare disease. Although worldwide prevalence rates are not known, overall prevalence in European countries has been reported as 15–50 cases per million population.\(^3\)

PAH usually presents with easy fatigability, dyspnea, chest pain, or syncope. Hoarseness of voice is rare in patients with PAH. The left RLN passes under the arch of the aorta to reach the larynx. This anatomical location puts it at the disadvantage of being compressed by enlargement of the nearby cardiac chambers or great vessels. Left RLN palsy due to the cardiac cause is known as Ortner's syndrome or the cardio-vocal syndrome.\(^4\) The common causes are left atrial enlargement, usually secondary to mitral stenosis, or thoracic aortic aneurysm. In rare cases of PAH, there is trapping of the left RLN between the aorta and the dilated left pulmonary artery, leading to Ortner's syndrome.\(^5,6\)

The diagnosis of PH in our case was established by chest radiography, electrocardiography, and echocardiography. Further investigations were performed to rule out known causes of PH, such as CT thorax for class 3 (secondary to lung diseases), contrast echo for congenital valvular heart diseases, ultrasonogram of the abdomen for portal hypertension, ENA profile for connective tissue disorders such as scleroderma, HIV testing, and fasting thyroid profile. After ruling out known causes of PH, we reached to the diagnosis of idiopathic PAH. The patient was started on vasodilator and diuretic treatment and is doing well since then.

Like every other disease in medical science, PAH can also present in varied ways. Other than the tell-tale signs and symptoms of the disease, hoarseness of voice as a symptom should be kept in mind as it could be the initial manifestation, as in this case. Treatment of idiopathic PAH is feasible nowadays, and good response can be expected if diagnosed early and managed properly.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

Rajiv Garg, Anubhuti Singh, Kamal Kumar Sawlani\(^1\), Ashwini Kumar Mishra

Departments of Pulmonary Medicine and \(^1\)Medicine, King George Medical University, Lucknow, Uttar Pradesh, India
E-mail: rajivkgmc@gmail.com

Figure 1: Chest radiograph suggestive of cardiomegaly

Figure 2: Computed tomography thorax revealing normal lung parenchyma
REFERENCES

1. D’Alonzo GE, Barst RJ, Ayres SM, Bergofsky EH, Brundage BH, Detre KM, et al. Survival in patients with primary pulmonary hypertension. Results from a national prospective registry. Ann Intern Med 1991;115:343-9.

2. Simonneau G, Robbins IM, Beghetti M, Channick RN, Delcroix M, Denton CP, et al. Updated clinical classification of pulmonary hypertension. J Am Coll Cardiol 2009;54 1 Suppl: S43-54.

3. Peacock AJ, Murphy NF, McMurray JJ, Caballero L, Stewart S. An epidemiological study of pulmonary arterial hypertension. Eur Respir J 2007;30:104-9.

4. Ari R, Harvey WP, Hufnagel CA. Etiology of hoarseness associated with mitral stenosis: Improvement following mitral surgery. Am Heart J 1955;50:153-60.

5. Rosenberg SA. A study of the etiological basis of primary pulmonary hypertension. Am Heart J 1964;68:484-9.

6. Shah KD, Ayyar KH, Shah UK. Hoarseness of voice a presenting manifestation of primary pulmonary hypertension. Indian J Otolaryngol 1980;32:35-6.