Ortner’s Syndrome as a Presenting Feature of Congenital Heart Disease in Infants

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ABSTRACT

Ortner’s syndrome is a rare condition characterized by hoarseness of voice due to left recurrent laryngeal nerve (RLN) palsy in association with cardiovascular disease. We report two cases of congenital heart disease in infants presenting with Ortner’s syndrome. We believe that the dilated pulmonary artery in the first case and the left atrial dilatation in the second case caused compression of the left RLN resulting in hoarseness of voice. After the corrective cardiac surgery, the voice of both the infants had normalized. Through this case report, we highlight that Ortner’s syndrome is an important differential diagnosis of hoarseness of voice in infancy.

Key words: Congenital heart disease, infant, Ortner’s syndrome, recurrent laryngeal nerve

INTRODUCTION

Ortner’s syndrome or cardiovocal syndrome refers to hoarseness of voice due to recurrent laryngeal nerve (RLN) palsy secondary to nerve impingement, stretching, or compression at the mediastinum.[1] This syndrome was first described in 1897 by Norbert Ortner, an Austrian physician, in three patients with severe mitral valve stenosis.[2] Subsequently, the syndrome has been described in adult patients with various cardiovascular diseases.[1] However, there is limited literature in children. We herein report two infants with congenital heart disease who presented with Ortner’s syndrome.

CASE PRESENTATIONS

Case 1
A 3-month-old baby presented with complaints of failure to thrive and weak cry for 1 week. On examination, the heart rate was 162/min and respiratory rate was 48/min with mild subcostal retractions. Blood pressure was 86/50 mmHg in the right upper arm. Failure to thrive was present with the weight at the fifth centile for age and sex. The first heart sound was normal, the second heart sound was loud, and an ejection systolic murmur of Grade 3/6 was heard in the left second intercostal space. Chest X-ray showed a cardiothoracic ratio of 0.6 and increased vascular markings in the lungs. A direct laryngoscopy done revealed left vocal cord palsy.

Echocardiography showed 5-mm ventricular septal defect (VSD), moderate-sized patent ductus arteriosus (PDA), and severe pulmonary hypertension. The left atrium, right ventricle, and pulmonary artery were severely dilated. The baby underwent corrective cardiac surgery for VSD and PDA. On follow-up after 2 months, the baby was thriving well and the voice had normalized.

Case 2
A 4-month-old baby presented with complaints of failure to thrive and weak cry by the mother which gradually worsened over 1 month. On examination, the heart rate was 172/min and respiratory rate was 46/min with...
subcostal retractions. Blood pressure was 82/48 mmHg in the right upper arm. Failure to thrive was present with the weight below the fifth centile for age and sex. The first heart sound was normal, the second heart sound was loud, and a pansystolic systolic murmur of Grade 4/6 was heard in the left fourth intercostal space. Chest X-ray showed a cardiothoracic ratio of 0.6 with increased vascular markings in the lungs.

Echocardiography showed three VSDs (5 mm, 4 mm, and 2 mm). Left atrial and right ventricle dilatation was present with normal pulmonary pressures. Laryngoscopy could not be done as the baby went to another hospital for corrective cardiac surgery. On follow-up after 1 month, the baby is thriving well and the voice had improved. Repeat echocardiography showed normal left atrial size.

**DISCUSSION**

Various case series attribute 1%–3% of cases of extralaryngeal hoarseness to Ortner’s syndrome. These include congenital heart diseases (atrial septal defect, VSD, PDA, and Ebstein anomaly), left atrial enlargement (mitral stenosis, mitral regurgitation, and left atrial myxoma), pulmonary artery diseases (pulmonary hypertension and pulmonary embolism), and aortic diseases (thoracic aneurysm).[1,3]

Many explanations have been offered for the pathogenic relationship between cardiovascular disease and left vocal cord paralysis.

Norbert Ortner had initially proposed that the enlarged left atrium impinged on the nerve under the arch of the aorta causing palsy.[2] Subsequent autopsies and radiological studies, however, disputed this hypothesis. Studies done in cadavers by Fetterrolf and Norris showed that the distance between the aorta and the pulmonary artery within the aortic window is only 4 mm and compression of the nerve between the two structures is responsible for palsy.[4]

As more cases of hoarseness of voice with cardiac disorders were reported, more theories were described such as lymphadenitis and scarring in the aortic window causing nerve fixation, pressure from the left bronchus, right ventricular hypertrophy, pulmonary artery atherosclerosis, and anatomical position of the ligamentum arteriosum.[1,5]

The left RLN anatomy will help to understand the cause–effect relationship. The left RLN branches off the vagus nerve just below the aorta. It then loops around the ligamentum arteriosum (between the superior surface of the origin of the left pulmonary artery and the inferior surface of the aortic arch) and ascends to the larynx along the tracheoesophageal groove. The peculiar course of the nerve at the aortopulmonary window makes it susceptible to compression by structures abutting this space.[1] The severe pulmonary artery dilatation in our first case caused the nerve palsy. The left atrial dilatation probably caused the nerve palsy in the second case. The dilated left atrium can elevate the left pulmonary artery into the concavity of the aortic arch obliterating the aortopulmonary window and thus compress the nerve. Although we could not do a laryngoscopy in our second case, the recovery of the voice after corrective surgery supports our hypothesis.

Compression results in insufficient microcirculatory supply to the RLN and causes ischemic neuronal degeneration and subsequent paralysis. The nerve injury gradually progresses from neuropraxia (Grade 1) to neurotmesis (Class V).[6] Thus, early recognition of the nerve palsy is important because reversibility of the nerve damage depends on the duration of injury.

Some studies have even mentioned that hoarseness of voice can be a rare presenting sign of congenital heart disease in infancy, and if a paralyzed left vocal cord is found, a comprehensive cardiovascular diagnostic workup is warranted. The voice usually returns back to normal after definitive surgery for congenital heart disease.

The prognosis of RLN palsy depends on the degree and duration of nerve compression. The recovery time in Grade 1 nerve injury can range from hours to a few months after the cardiac surgery.[6] Since slow or partial injury to the nerve may not always result in hoarseness, routine examination of the vocal fold in all cases of heart disease has also been advocated by some authors.[1]

**CONCLUSION**

Physicians should suspect and look beyond the larynx for a cause of vocal cord palsy in infants presenting with hoarseness of voice. Ortner’s syndrome is an important differential diagnosis of hoarseness of voice in infancy. Comprehensive evaluation and timely intervention can help in restoring vocal cord function and avoiding permanent damage.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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