

Ortner Syndrome in Infants

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Ortner syndrome or cardiovocal syndrome refers to hoarseness of voice due to recurrent laryngeal nerve paralysis secondary to cardiovascular disease. We present three cases of Ortner syndrome in infants with congenital heart disease. All the three cases had moderate to severe pulmonary hypertension with moderately dilated pulmonary artery. We believe that the dilated pulmonary artery caused compression of the left recurrent laryngeal nerve resulting in hoarseness of voice.

Key words: *Congenital heart disease; Infants; Ortner's syndrome.*

Ortner syndrome or cardiovocal syndrome refers to hoarseness of voice due to recurrent laryngeal nerve paralysis secondary to cardiovascular disease. This syndrome was first described in 1897 in two patients who had mitral stenosis and left recurrent laryngeal nerve paralysis(1). Subsequent papers have described this syndrome in adult patients with cardiovascular disease(1-4). Literature is limited for children(5-6). We herein present three cases of Ortner syndrome in infants with congenital heart disease.

CASE REPORT

Case 1: A 2 months old male child presented with complaints of cough, cold, breathlessness and hoarse cry since two days. On examination, the child was afebrile with heart rate of 180/min, and respiratory rate of 76/min. Blood pressure was 88/50 mm Hg in right upper arm. There was mild central cyanosis. Liver was enlarged and tender with a span of 6 cm in the midclavicular line. Crepitations were heard bilaterally with equal air entry on both sides. First heart sound was normal, second heart sound was loud and an ejection systolic murmur of grade 3/6

was heard in the left 2nd intercostal space without any radiation. Chest X-ray showed a cardiothoracic ratio of 0.6 with enlarged pulmonary artery segment and increased vascular markings in the lungs. A direct laryngoscopy done revealed left vocal cord palsy. Echocardiography revealed non obstructive supracardiac TAPVC to left vertical vein, a moderate sized atrial defect (5 mm) with right to left shunt, two small mid muscular ventricular septal defect 2-3 mm with a bidirectional shunt, severe pulmonary hypertension, and severe dilatation of the pulmonary artery.

Case 2: A 4 months old female child presented with complaints of cough, cold, and breathlessness since five days. There was history of dysphonia since 2 months. On examination the child was afebrile with heart rate of 160/min, and respiratory rate of 66/min. Blood pressure was 84/50 mmHg in right upper arm. There were bilateral crepitations. Pansystolic murmur of grade 3/6 was heard in the left 5th intercostal space. First heart sound was soft with a loud and widely split second heart sound. Liver was enlarged and tender with a span of 7 cm in the midclavicular line. Chest X-ray showed a cardiothoracic ratio of 0.7 with enlarged pulmonary

artery segment and engorged vascular markings in the lungs. A direct laryngoscopy done revealed left vocal cord palsy. Echocardiography revealed mitral atresia, 3mmVSD, moderate sized PDA, with 2 mm ASD, and severe pulmonary hypertension. Left atrium, right ventricle and pulmonary artery were severely dilated.

Case 3: A 3 months old male child presented with complaints of cough, cold, breathlessness since three days. There was history of hoarse cry since 1 month. On examination, the child had tachypnea, tachycardia, normal BP, bilateral crepitations, loud S2, ejection systolic murmur of grade 3/6 in the left 2nd intercostal space, and hepatomegaly. Chest X-ray showed a cardiothoracic ratio of 0.65 with enlarged pulmonary artery segment and engorged vascular markings in the lungs. A direct laryngoscopy done revealed left vocal cord palsy. Echocardiography revealed double outlet right ventricle with mitral atresia with 2 mm ASD. Severe pulmonary hypertension was present with severe dilatation of pulmonary artery.

In all the three cases described, the patients were started on antifailure management and they are on regular follow up and are awaiting surgery. The hoarseness of voice is still persisting in all of them.

DISCUSSION

Many explanations have been offered for the pathogenic relationship between cardiovascular disease and left vocal cord paralysis(2). However, recently the importance of enlarged pulmonary artery as the common and main mechanism of nerve injury has been highlighted. Fetterolf and Norris made a careful study of the anatomic relations of left recurrent laryngeal nerve in cadavers and concluded that the nerve must be squeezed between the left pulmonary artery and the aorta or ligamentum arteriosum(7). Many authors(1,3,8) have concluded that the etiology of left recurrent laryngeal nerve paralysis was compression of the nerve between the enlarged tense pulmonary artery and the aorta at the ligamentum arteriosum. That is the reason why Ortner syndrome also occurs in primary pulmonary hypertension, Eisenmenger syndrome due to atrial septal defect where the atrium is not enlarged, patent ductus arteriosus with pulmonary hypertension(2).

All the three cases which we have described above had moderate to severe pulmonary hypertension with moderately dilated pulmonary artery. We believe that the dilated pulmonary artery caused compression of the left recurrent laryngeal nerve resulting in hoarseness of voice.

Some studies have even mentioned that chronic hoarseness of voice can be a rare presenting sign of congestive cardiac failure in infancy and if a paralyzed left vocal cord is found, a comprehensive cardiovascular diagnostic workup is warranted(6). The voice usually returns back to normal after definitive surgery for the congenital heart disease. There is no correlation between the reported duration of the hoarseness and the severity of the pulmonary hypertension prior to operation and the time to recover function after operation.

We wish to highlight that in a child with congenital heart disease and hoarseness of voice, it is necessary to look for pulmonary hypertension and dilatation of the main pulmonary trunk.

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