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Case Report

Transient cardiovocal syndrome in neonate: a case report

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Abstract: Background: Cardiovocal syndrome, which refers to left recurrent laryngeal nerve paralysis secondary to cardiovascular disease, is seldom diagnosed in neonates or infancy. Only two case reports were found in the international literature and our report details the youngest patient yet. Additionally, the patient had a distinctive and subtle initial presentation, with isolated weak crying without significant cardiovascular or respiratory symptoms, and a congenital ductus arteriosus aneurysm which made definitive diagnosis challenging. Case presentation: This male neonate presented with weak crying, hoarseness, mild tachypnea, and feeding cyanosis at birth. Laryngoscopy showed left vocal cord paralysis. Heart computed tomography revealed a 0.7-cm ductus arteriosus aneurysm. Because his symptoms were extremely subtle, supportive treatment was provided and he was monitored closely. Six days later, all his symptoms had resolved. Heart ultrasonography and laryngoscopy were repeated and both the ductus arteriosus aneurysm and left vocal cord paralysis had subsided spontaneously. Conclusions: Diagnosis of cardiovocal syndrome in neonates is difficult, especially in neonates whose symptoms are subtle or transient. When cardiovocal syndrome is related to congenital ductus arteriosus aneurysm, a "wait and see" policy is justified because of the benign nature of the entity.

Keywords: Cardiovocal syndrome, neonate, ductus arteriosus aneurysm

Background

Cardiovocal syndrome, also called Ortner syndrome, is defined as left recurrent laryngeal nerve (LRLN) paralysis secondary to cardiovascular disease. It was first described in Nobert Ortner [1] in 1897 when three patients presented with severe mitral stenosis and hoarseness. Subsequent papers reported several adult cases associated with a myriad of clinical situations, but the literature is limited in children and infants [2-4]. Here, a case is presented of a newborn infant with cardiovocal syndrome who recovered spontaneously.

Case presentation

A 2750-g male neonate was delivered at term via vaginal delivery by a 34-year-old mother as her first baby. The infant had a good Apgar score without respiratory distress, meconium stained amniotic fluid, or delayed initiation of crying. However, weak crying, hoarseness, mild tachypnea, and cyanosis during feeding were noted just hours after birth. Therefore, an otolaryngologist was consulted. Fiberoptic bronchoscopy showed left vocal cord paralysis without subglottic stenosis or a tracheal anomaly (Supplementary Video). To determine the cause of left vocal cord paralysis, a series of examinations were conducted including brain ultrasound and cardiac computed tomography (CT) scan. No central nervous system abnormality was noted, but a 0.7-cm outpouching of the thoracic aorta near the aortic isthmus was observed, indicating an aortic ductus arteriosus aneurysm (DAA) (**Figure 1A**, **1B**). Under the impression of cardiovocal syndrome related to DAA, the pediatrician kept the neonate in the nursery for supportive treatment and close observation. His feeding and respiratory status became stable 6 days later. The pediatric cardiologist repeated the heart ultrasound and

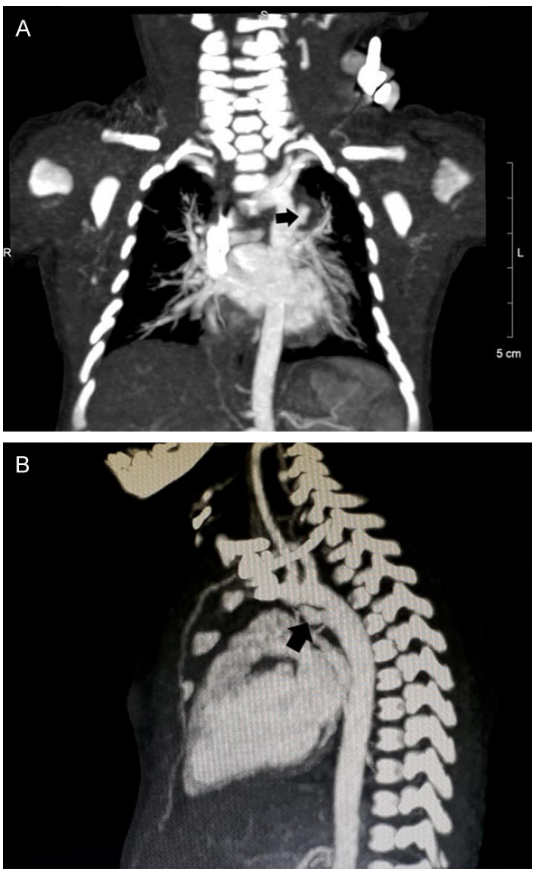

Figure 1. CT of the heart and vessels showed a ductus arteriosus aneurysm, 0.7 cm in diameter (arrow) (A: AP view, B: Lateral view).

found the DAA had disappeared (**Figure 2**). Repeated laryngoscopy at 21 days of age showed that the vocal cord paralysis had resolved. Finally, the infant was able to cry normally and had growth by the following clinic visit.

Discussion

Because the recurrent laryngeal nerve supplies all of the muscles of the larynx except the cricothyroid muscle, searching along the route of the nerve is done to identify possible causes of vocal cord paralysis. The right recurrent laryngeal nerve crosses and hooks around the right subclavian artery and the LRLN crosses the aortic arch and hooks around the ligamentum arteriosum. Both recurrent laryngeal nerves finally ascend in the tracheoesophageal groove. Due to its lengthy and vulnerable course, the LRLN is more prone to injury than the right recurrent laryngeal nerve [5, 6].

Isolated left vocal cord paralysis at birth can be congenital or acquired. In addition to birth trauma, a cardiopulmonary disorder must also be kept in mind [3]. Although Ortner initially postulated left vocal cord palsy as the result of compression between an enlarged left atrium and the aortic arch, many recent autopsies and radiologic studies refuted his hypothesis. Fetterrolf and Norris reported the distance between the aortic arch and pulmonary artery was only 4 mm, so they favored the narrow aortopulmonary window and compression between the two structures as the likely cause of LRLN palsy [7]. Various congenital heart diseases are reportedly related to cardiovocal disease, such as primary pulmonary hypertension, Eisenmenger syndrome due to atrial septal defect without enlarged atrium, and patent ductus arteriosus with pulmonary hypertension [8].

DAA, a rare but potentially fatal abnormality [9-12], also resides in the aortopulmonary window. It can be congenital or acquired. The mechanism of congenital DAA is uncertain, but it may be a normal variant of an elongated ductal bump or part of a normal process of spontaneous ductal closure [13]. Acquired DAA is commonly found as a complication of surgical closure of patent ductus arteriosus or after a ductal infection [14, 15]. This case was ruled to be one of congenital DAA, which is usually diagnosed before 2 months of age. The risk factors include large-for-gestation-age size at birth, maternal gestational diabetes mellitus, [16] and mothers with blood type A [13]. The true incidence of cardiovocal syndrome may be underestimated, as only 16% of patients had symptomatic related congenital DAA [10]. Until recently, the incidence of congenital DAA was reported as 0.8% based on neonatal autopsy reports [17], 1.5% based on fetal ultrasonography at more than 30 weeks' gestation [10], and 8.8% based on echocardiography of full-term neonates by echocardiography [13].

Chest plain-film radiography was initially used to diagnose congenital DAA, but a left upper mediastinal mass is easily neglected if the DAA is small [18]. Transthoracic echocardiography is a better way to diagnose and monitor these cases. Three important signs, including an unusual ductal shunt jet, "triple star sign," and "rabbit ear sign" are diagnostic imaging signs of DAA [13]. Magnetic resonance imaging or

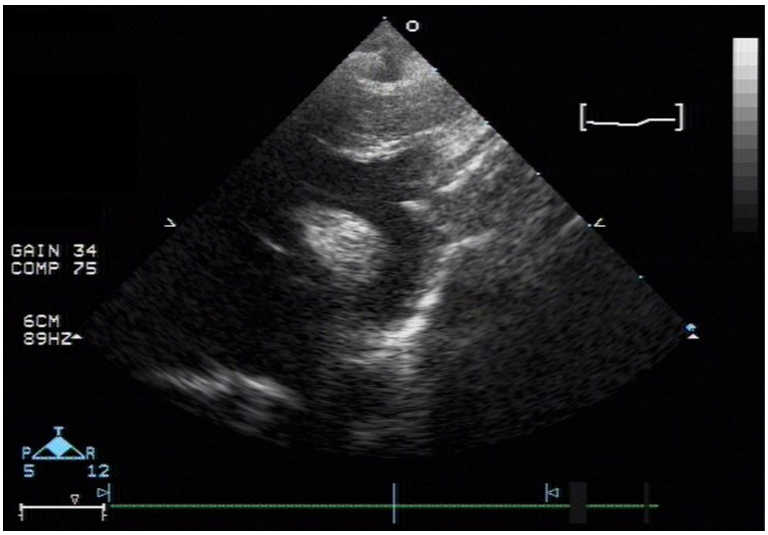

Figure 2. Follow-up heart ultrasound showed the absence of ductus arteriosus aneurysm.

computed tomography is also helpful, especially when a possible thrombus extends into the adjacent vascular structures or in the presence of compression of the extravascular structures [13].

Although oral or intravenous indomethacin can be given to close a patent ductus arteriosus in preterm infants [19], its effect in full-term infants or DAA is uncertain. Surgical resection of the DAA is indicated in the following conditions: persistent DAA beyond the neonatal period, DAA associated with connective tissue disease, the presence of thromboembolism, or significant compression of adjacent structures [10]. Fortunately, some DAAs spontaneously disappear after ductal closure, or they become organized after progressive thrombus formation [13]. Therefore, a "wait and see" strategy is a reasonable and safe policy for coping with congenital DAAs [20].

There are two important lessons to learn from this case. First, abnormal crying at birth can be an important clue to an underlying disease. An alert and curious pediatrician can make an accurate and prompt diagnosis by identifying the qualities of neonatal crying and their associated symptoms, such as stridor, cyanosis, or feeding difficulties. Differential diagnosis of weak crying or hoarseness in an infant includes congenital hypothyroidism, low muscle tone, infection, or vocal cord lesions, which can be easily found by an otolaryngologist performing fiberoptic laryngoscopy.

Second, whether or not cardiovocal syndrome resolves depends on the underlying pathology and the degree and duration of injury to the recurrent laryngeal nerves. According to Sunderland's classification, only class I (neuropraxia) and class II (axonotmesis) nerve injury can completely recover when the offending agent is removed [21]. Although laryngeal electromyography can provide prognostic information, it is difficult to perform on infants, and only the positive predictive value is reliable [22]. Although the precise association between the duration of hoarseness and recovery time remains unknown, most cases of cardiovocal syndrome in infancy are reported to have favorable prognoses, possibly due to the short term of nerve compression or traction [3, 20, 23]. Therefore, if immediate identification of the underlying heart disease is achieved, proper treatment can be provided through either observation or operation, and full recovery of vocal cord function can be expected in infancy.

Conclusions

Cardiovocal syndrome in infancy usually has a good prognosis. An otolaryngologist must be familiar with the disease and cooperate with pediatricians to avoid complications such as choking, aspiration, or respiratory distress. Repeated laryngoscopy and heart ultrasonography are also necessary for follow-up.

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Disclosure of conflict of interest

None.

Abbreviations

DAA, ductus arteriosus aneurysm; CT, computed tomography; LRLN, left recurrent laryngeal nerve.

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Supplementary Video 1. Fiberoptic laryngoscopy at birth showed left vocal cord paralysis.