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Case Report

Diaphragmatic paralysis after surgery in a patient with scimitar syndrome: A case report

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ARTICLE INFO

Article history:
Received 21 September 2014
Received in revised form
6 December 2014
Accepted 29 April 2015
Available online 7 August 2015

Keywords:
Scimitar syndrome
Congenital heart disease
Phrenic nerve paralysis

ABSTRACT

Introduction: Scimitar syndrome is a complex congenital disorder affecting the right lung. Diaphragma paralysis is one of the important causes of morbidity and mortality in congenital heart surgery especially under 2 years of age in pediatric cardiac surgery. Phrenic nerve injury is a complication which occurs after cardiothoracic surgery and causes increased morbidity and mortality by leading diaphragmatic paralysis.

Aim: We present this rare pulmonary anomaly case of the patient who remained connected for a long time to respiratory equipment as a result of phrenic nerve palsy which developed and was eventually discharged with healing.

Case study: A 6 months of age and 4.5 kg infant was admitted because of recurrent lung infections. It has been seen in posteroanterior chest X-ray and chest tomography that the right lung was hypoplasic and the heart was displaced to the right side of the body. The decision of surgery was taken for the patient who was discussed in the Council. The patient was extubated on 14th postoperative day. Paradoxical diaphragmatic movement was detected as the results of the examination of the patient and fluoroscopy. The patient was discharged from the intensive care unit on 49th postoperative day.

Results and discussion: If the cases require mechanical respiratory support after congenital heart surgery despite intra-cardiac full correction is done and if there was no problem in cardiac function, the paralysis of the diaphragm should be considered. The difference in the recovery time of the phrenic nerve and diaphragm function is thought to be related to the degree of the phrenic nerve injury.

Conclusions: Decrease in action potential amplitude and/or delay or non-response in phrenic nerve latency suggesting or supporting the phrenic nerve lesion are observed in infants and young children mostly in the early period after cardiac surgery. However, this situation is temporary and good care during this period provides restoring the patient's health.

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http://dx.doi.org/10.1016/j.poamed.2015.04.010
1230-8013/© 2015 Published by Elsevier Sp. z o.o. on behalf of Warmińsko-Mazurska Izba Lekarska w Olsztynie.

1. Introduction

Scimitar syndrome is a complex congenital disorder affecting the right lung.[1] It is a rare anomalous pulmonary venous return and characterized by opening of right pulmonary vein to inferior vena cava or right atrium.[2] It was defined by Cooper et al. for the first time in 1836 and published by Neil et al. in 1960.[3] It was given this name because the image in chest X-ray was thought to be similar to the ''Turkish swords.''[4]

Accompanying other pathologies are dextroposition of the heart, secondary to right lung hypoplasia, varying degrees of bronchial abnormalities and providing of the arterial circulation of hypoplasic region by a collateral separated from the ascending aorta.[5] Diaphragm paralysis is one of the important causes of morbidity and mortality especially under 2 years of age in pediatric cardiac surgery.[6] The respiratory distress caused by paralysis of the diaphragm especially in pediatric patients under the age of 2 and consequent reduction in exercise capacity may require long-term treatment of respiratory support, the dependency to respirator leads to a vicious cycle by preparing the ground for respiratory tract infections.

Phrenic nerve injury is a complication which occurs after cardiothoracic surgery and causes increased morbidity and mortality by leading diaphragmatic paralysis.[7] According to the literature, the patient's age has negative correlation with the significance of clinical symptoms.[8,9] Diaphragm paralysis often leads to respiratory failure requiring ventilatory treatment in infants and young children.

2. Aim

We present this rare pulmonary anomaly case of the patient who remained connected for a long time to respiratory equipment as a result of phrenic nerve palsy which developed and was eventually discharged with healing.

3. Case study

The patient, 6-month-old, 4.5 kg infant was admitted because of recurrent lung infection. It has been seen in posteroanterior chest X-ray and chest tomography that the right lung was hypoplasic and the heart was displaced to the right side of the body. In echocardiography, dextrocardia, right pulmonary artery hypoplasia, partial anomalous pulmonary venous return (Scimitar send), ventricular septal defect (midmuscular), persistent foramen ovale, patent ductus arteriosus, tricuspid insufficiency (TI 2–3) were determined. The decision of surgery was taken for the patient who was discussed in the Council. The patient, who was operated, was taken to intensive care under inotropic support. The patient was followed with for the postoperative (PO) first 2 days by anesthetizing. Inotropic support was discontinued completely on the PO 5th day. The patient, who was followed with intubation due to the lung problem, was extubated on the PO 14th day. However, he was intubated again in a short time due to deterioration in hemodynamics and oxygenation. The patient was followed in synchronized intermittent mandatory ventilation (SIMV) and continuous positive airway pressure (CPAP) mode. Paradoxical diaphragmatic movement was detected as the results of the examination of the patient and fluoroscopy. Tracheostomy was opened because of the long duration of intubation on the 18th day. The patient was continued to be followed with SIMV and CPAP mode. In patient who was followed by positive end-expiratory pressure (PEEP) values up to 20 mmHg, O_2 saturation, and blood pressure were 98% and 112/58 mmHg, respectively. In the patient who was followed by reducing the PEEP values, the O_2 saturation, blood pressure and pulse were 97%, 103/62 mmHg and 78 bpm, respectively when the PEEP value was set to 14 mmHg and the patient was taken to T tube on the PO 40th day. The O_2 saturation, blood pressure and pulse were 97%, 101/60 mmHg and 75 bpm, respectively with T tube. Hemodynamically stable patient was taken to room air on the PO 42th day. The O_2 saturation, blood pressure and pulse were 96%, 110/64 mmHg and 72 bpm, respectively, in the room air. The patient without a change in hemodynamics was decannulated on the PO 44th day. When the O_2 saturation, blood pressure and pulse were 95%, 113/62 mmHg, 76 bpm, respectively, the patient, who was extubated and followed in the intensive care, was discharged from the intensive care unit on the PO 49th day. The patient, who was followed in the clinic, was discharged with healing on the PO 57th day.

4. Results and discussion

Scimitar syndrome has wide range of symptomatology depending on the accompanying anomalies. While the clinic is more complicated in cases diagnosed in infancy due to congestive heart failure and pulmonary hypertension, patients may be asymptomatic in the older age group.[3] Among the anomalies accompanying to the syndrome, hemivertebra and scoliosis, a variety of congenital heart disease can be considered besides venous return and diaphragmatic abnormalities. Congenital heart disease may accompany the syndrome in approximately 25% of cases, and the most common of them is septal defect.[10] In our case, ventricular septal defect, persistent foramen ovale, and patent ductus arteriosus were detected in echocardiography. The most common form of the syndrome is the association of hypogenetic lung and partial anomalous pulmonary venous return.[11] Two forms including infantile and adult were identified. In general, if the patient diagnosed is an infant; it may be observed with in the presence with larger tables such as heart failure and pulmonary hypertension occurring due to the broad systemic and pulmonary collateral arteries.[12] Our case was evaluated as infantile type.

Symptoms include dyspnea, tachypnea and hemoptysis.[13] Third part of the cases is diagnosed in childhood with chest X-ray taken due to recurrent pneumonia.[14] In our case, the problem of recurrent pulmonary infections was seen. CT, CT angiography, digital subtraction angiography (DSA) can be used besides radiography for diagnosis in these patients magnetic resonance angiography (MRA) emerges as a new method that allows the imaging of pulmonary vascular structures and pathologies in a non-invasive way.[15] Surgical treatment was applied for fixing the shunt occurring due to

venous return anomaly, infected bronchiectasis and recurrent pneumonia after the definitive diagnosis.

In various studies, the rate of diaphragmatic paralysis developing after congenital heart surgery was found as 0.3%–12.8%.[6] Age of the patients with paralysis of the diaphragm is important in the clinical follow-up after surgery. According to the literature, the patient's age has negative correlation with the significance of clinical symptoms.[8,12] If the cases require mechanical respiratory support after congenital heart surgery despite intra-cardiac full correction is done, and if there was no problem in cardiac function, the paralysis of the diaphragm should be considered. Especially in the early PO period, diagnosis may be delayed in intubated cases, who were ventilated with positive pressure, due to elevation of the diaphragm is not obvious. It has been reported in the literature that the diaphragm functions were completely recovered within 6–12 months after the recovery in phrenic nerve injury; however the paralysis has also been reported to be in excess of 3 years.[6]

In the literature, paralysis of the phrenic nerve was found to be transient in some patients with electrophysiological assessments repeated PO follow-up. The time required for the recovery of phrenic nerve and diaphragm functions and prognosis indicate variability. In studies in adult patients with diaphragm paralysis, Markand et al. have reported that recovery was seen in 3 of the 4 patients with diaphragm paralysis that they have followed in the period after cardiac surgery within 3–6 months. Diaphragmatic paralysis of our patient recovered 40 days after the operation. The exact time of the irreversible paralysis of the phrenic nerve should be identified in order to prevent an unnecessary surgery in patients with possible temporary paralysis of the phrenic nerve. The difference in the recovery time of the phrenic nerve and diaphragm function is thought to be related to the degree of the phrenic nerve injury. The presence of a temporary paralysis suggests that multiple factors such as focal ischemia or edema caused a temporary conduction block. Faster healing and a good prognosis are expected in cases without axonal loss and with nerve damage causing mild demyelination. In contrast, serious phrenic nerve injury leading to demyelination and axonal loss should come to mind in cases showing slow recovery time.

5. Conclusion

Decrease in action potential amplitude and/or delay or non-response in phrenic nerve latency suggesting or supporting the phrenic nerve lesion are observed in infants and young children mostly in the early period after cardiac surgery. However, this situation is temporary and good care during this period provides restoring the patient's health.

Conflict of interest

None declared.

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