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RESEARCH ARTICLE

PHRENIC NERVE PALSY: A RARE CAUSE OF RESPIRATORY DISTRESS IN NEWBORN

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Abstract

Phrenic nerve palsy is a rare cause of respiratory distress in newborns, and often under-diagnosed. It is often accompanied by brachial plexus palsy. The possibility of spontaneous recovery should be considered before choosing surgery. This rare cause of respiratory distress of the newborn can be easily overlooked among the many common etiologies of respiratory distress of the newborn if this entity is not kept in mind and a thorough workup is not performed. We report a newborn who was admitted to the neonatal unit with respiratory distress and paresis of the right upper limb. He had a dystocia labor. Chest x-ray and ultrasound showed an elevation of the right diaphragmatic dome. The diagnosis was phrenic nerve palsy associated with brachial plexus palsy secondary to birth trauma. The newborn was treated with continuous positive airway pressure (CPAP) ventilation without surgical intervention. The outcome was favorable with spontaneous recovery at the age of 2 months. The management of this association is not yet codified. Further investigations will be necessary to develop more definitive guidelines for the treatment of this condition.

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Introduction:-

Phrenic nerve palsy is an injury of the peripheral nervous system that occurs after or during stretching of the C3, C4 and/or C5 cervical spinal cord fibers. Phrenic nerve injury is often unilateral and sometimes associated with brachial plexus paralysis [1].

The clinical manifestation is characterized by early neonatal respiratory distress. The differential diagnosis can be made with neuromuscular diseases. With a mortality rate of 10-15%, the prognosis is generally guarded. Therapeutic recommendations are not yet established [2].

Case Report

Male newborn born by vaginal delivery in cephalic presentation, from a full term pregnancy, of a 41 year old mother followed for poorly balanced type 2 diabetes, fourth gesture, fourth pare. The delivery was dystocic, with a poor adaptation to the extra-uterine life (Apgar at 6 at the 5th minute + delayed cry). The birth weight was 5300 g. The newborn presented from birth a respiratory distress with a decrease of the mobility of the right upper limb. The examination noted a tachypnea at 79 cycles / minute, a tachycardia at 162 beats / minute, an oxygen saturation at 82% in free air and 94% under oxygen, a peribuccal cyanosis during crying and a Silverman score at 4/10. The neurological examination noted a paresis of the right upper limb with hand pronation and supination. Chest

radiography showed an ascension of the right diaphragmatic cupola, which was constant on multiple radiographs (**Figure 1**) and (**Figure2**).

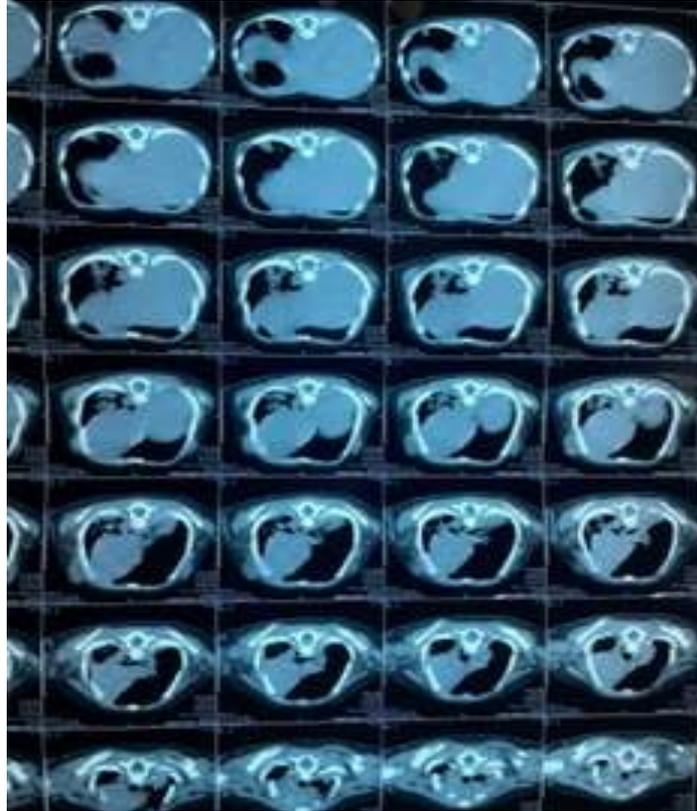
Figure 1:- Chest X-ray showing elevated right side of diaphragm.



Figure 2:- Another Chest X-ray showing elevated right side of diaphragm.



Ultrasound of the domes showed an ascending right dome rising on inspiration, reflecting paradoxical breathing, and a fixed left dome with normal mobility. Chest CT showed bilateral alveolar condensation with abnormal liver perfusion.

Figure 3:- Thoracic angioscanner showing transmediastinal hernia.

The thoracic angioscanner showed asymmetry in the size of the 2 pulmonary hemifields with right anterior transmediastinal hernia without venous return anomaly (figure 3). The diagnosis was right phrenic palsy associated with right brachial plexus palsy secondary to obstetrical trauma. The newborn was treated with oxygen therapy (4 liters/minute) with continuous positive airway pressure for 35 days. The decision was to maintain CPAP without recourse to surgery. In addition, physiotherapy was started at 4 weeks of life. The evolution was good with normal breathing. Follow-up revealed improved movement of the right upper limb.

Discussion:-

Diaphragmatic dysfunction is a rare cause of respiratory distress in infants. Traumatic lesion of the phrenic nerve is caused by overstretching of the cervical nerves due to hyperextension or twisting and torsion of the head and neck during delivery [2].

There are many risk factors, most commonly traumatic delivery, breech delivery and shoulder dystocia are often associated with perinatal phrenic nerve palsy. Traction and excessive stretching of the 3rd to 5th cervical nerve may result in phrenic nerve disruption or laceration, usually unilateral and on the right side. [4].

In any early neonatal respiratory distress, which is largely due to overactivity of the normal hemi-diaphragm, phrenic paralysis should be suspected. Otherwise, brachial plexus paralysis is manifested by paralysis or paresis of the affected upper limb with pronation-supination of the hand, as in the case of our newborn[6].

The majority of babies with brachial plexus palsy had respiratory complications sufficient to warrant diaphragmatic plication [3].

There is cyanosis and poor respiratory effort requiring mechanical ventilation in cases of bilateral paralysis. Sometimes diaphragmatic paralysis can be missed without a thorough examination [3].

Two mechanisms explain the pathophysiology of this respiratory distress. The first is the impairment of diaphragmatic contraction causing a change in thoracic volume with overuse of the intercostal muscles and the contralateral diaphragm. This leads to paradoxical breathing with respiratory muscle fatigue due to energy failure. The second mechanism is related to the elevation of the diaphragm causing atelectasis with susceptibility to respiratory infections and hypoxia[7-8].

The diagnosis is suggested on chest radiograph when the right hemidiaphragm is two intercostal spaces higher than the left or when the left hemidiaphragm is one intercostal space higher than the right [3].

The thoracic ultrasound allows the qualitative evaluation of the diaphragmatic function. In our case, the thoracic radiography showed an elevation of the right diaphragmatic dome projecting onto the sixth right intercostal space, a constant radiological image on several images [9]. The diagnostic methods of choice are diaphragmatic ultrasound and phrenic nerve conduction studies[3].

There is still no consensus on treatment. The goal is to achieve proper weight gain. Treatment is based on oxygenation, ventilation with continuous positive airway pressure (nasal CPAP), assisted ventilation, or surgical plication of the diaphragm by thoracoscopy or thoracotomy. The indication for each of these methods is not precise. However, the choice and the timing of treatment are controversial [6].

According to Escande et al, non-invasive nasal CPAP should be proposed for the treatment of obstetric phrenic paralysis before more invasive ventilation techniques are introduced [10].

These babies require prolonged mechanical ventilation if both hemi-diaphragms are paralyzed. These babies improve over a period of 2-3 weeks. Further improvement is possible over a period of 2 months. Surgical plication of the diaphragm should be performed if there is no further improvement and the newborn cannot be weaned from the ventilator. In symptomatic patients, surgery should be considered after an adequate trial of expectant management [11].

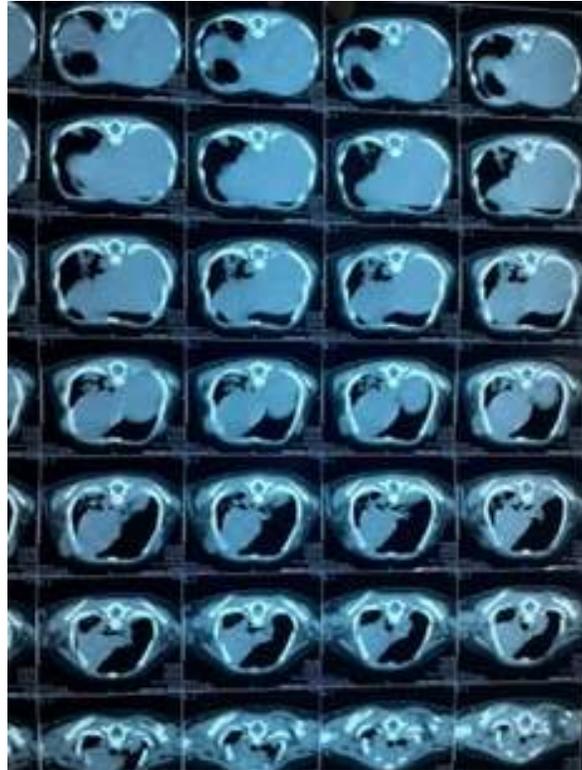
The timing of surgical plication is controversial, before 20 days or at 1 to 2 months of age, as it seems that no spontaneous improvement can be seen after this age[12].

In our case, a treatment based on continuous positive pressure ventilation was chosen. However, the decision of surgical abstention was amply discussed with a multidisciplinary team.

In the literature, it is reported that regression occurs in an average of 1 month in infants followed up without surgery[4].In the study by Bowerson, 2 patients underwent diaphragmatic plication at the age of 1 to 2 months and experienced a significant and rapid improvement in respiratory function postoperatively [9].

Conclusion:-

Phrenic nerve palsy is a rare condition. It is usually associated with brachial plexus paralysis and is unilateral. It should be considered in cases of respiratory distress after traumatic birth. Chest x-ray and ultrasound are usually sufficient for diagnosis. Surgical plication is an option, but it should also be noted that phrenic nerve palsy may resolve without surgery, as in our case.



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