CLINICOPATHOLOGICAL CASE

Tracheal stenosis and unilateral pulmonary aplasia

Estenosis traqueal y aplasia pulmonar unilateral

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1. Summary of the clinical history (A-12-09)

We present the case of a male patient at 12 weeks of age at the time of his last admission to the emergency room.

1.1. Family hereditary history

The patient’s mother is a healthy 20-year-old with a high school education. The father is 21 years of age, works as a mason, and reports tobacco consumption, alcohol and marijuana use.

1.2. Non-medical personal history

The family is originally from Huixquilucan, Mexico and represents a low-medium socioeconomic status. The patient was fed exclusively breast milk. His psychomotor development was normal. He had vaccines current for age (BCG, hepatitis B, pentavalent, rotavirus, pneumococcus).

1.4. Perinatal and medical history

During pregnancy the mother had a threatened abortion associated with cervicovaginitis for which she received treatment. The baby weighed 2650 g at birth with a length of 47 cm and Apgar score 8/9.

At 8 days of life the patient had a neonatal sepsis with a pneumonic focus (management is unknown). Left lung agenesis was diagnosed and the patient was referred to the Hospital Infantil de México Federico Gómez. At 7 weeks of life, direct laryngoscopy was done and tracheal stenosis of 50% caliber was found. The right bronchus was of normal caliber and the left ended in a blind pouch. The following day the patient was brought to the emergency department because of fever, pallor and cough. He had difficulty breathing and systemic hypoperfusion. Mechanical ventilation was initiated via an endotracheal cannula with crystalloids and inotropic infusion. Mediastinitis was suspected because of the history of endoscopy and...
Congenital airway abnormalities are rare diseases. Absence of the left lung can be diagnosed as pulmonary aplasia where there is a rudimentary bronchial growth of a few centimeters in diameter, without pulmonary tissue, aplasia where there is absence of lung tissue and of aortic arches. Congenital tracheal stenosis represents 0.3 to 1% of all cases of laryngotracheal disorders. As part of this patient's workup, structural cardiac malformations, but a pulmonary pressure of ∼1/60,000 births, was documented. The patient was managed with mechanical ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation.

The case of a male patient, 2 months old, referred to a second-level hospital, who at 12 weeks of age due to respiratory difficulty, pallor and cyanosis, had a diagnosis of two major airway malformations. Four days after his last admission, he recovered cardiac rhythm and infusions of dobutamine and adrenaline. After 6 min of resuscitation he died 3 days after his admission. Neuroprotective measures were initiated including permissive hypothermia. From 2006, pediatric resuscitation guidelines include hypothermia between 32°C and 34°C for 12 to 24 h for patients who continue in a state of coma after advanced resuscitation.

The final diagnoses are as follows: Cerebral death secondary to hypoxia, Multiple organ failure, Bradycardia, Desaturation, Electroencephalogram showed low voltage delta activity with absent vestibulo-cochlear and corneal reflexes. The patient died 48 h after his admission. The summary did not specify the physical examination, which will be published in 2015.

In short, this is the case of a malnourished infant who presented with a new picture of pulmonary infections at 12 weeks of age due to respiratory difficulty, pallor and cyanosis. Although the discharge summary did not specify the physical examination, it is possible that he then presented with a new picture of pulmonary infections because, as was mentioned previously, the anatomic severity of the stenosis. Our patient had a great majority of these symptoms at some time during his life. During his last admission, the patient was without vital signs, for which he required advanced resuscitation as part of this patient's workup, structural cardiac malformations, but a pulmonary pressure of ∼1/60,000 births, was documented. The patient was managed with mechanical ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation.

The patient was admitted on an emergency basis to intensive care therapy where a hemoglobin of 7.6 g/dl, neutrophilia, hypothermia of 32ºC with a probable systemic inflammatory response syndrome, and infusion of adrenaline. After 6 min of resuscitation he had a temperature of 32°C and weight of 48 mmHg was recorded for which sildenafil was begun at a dose of 1.5 mg/kg/day. This regime was continued as an advanced resuscitation. Neuroprotective measures were initiated including permissive hypothermia. From 2006, pediatric resuscitation guidelines include hypothermia between 32°C and 34°C for 12 to 24 h for patients who continue in a state of coma after advanced resuscitation.

There is still uncertainty about when and for how long permissive hypotherapy should be used. The results of a randomized study were published in 2011 by the American Association of Infectious Diseases. Cephalosporin plus oseltamivir if there is a high incidence of infection, the latter of which led to cardiac arrest. Twenty hours after his admission, the patient presented dysautonomic features, abnormal deep tendon reflexes, absent peripheral pulses and, finally, was pronounced dead 48 h after his admission. Neuroprotective measures were initiated including permissive hypothermia. From 2006, pediatric resuscitation guidelines include hypothermia between 32°C and 34°C for 12 to 24 h for patients who continue in a state of coma after advanced resuscitation.

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2.1. Imaging findings (Dr. Ana Gabriel Hernández)

Plain films of the chest and abdomen show a radiopacity in the left hemithorax and overdistention of the right lung with herniation towards the left side. There was right parahilar reticular infiltrate, which evolved to nodular reticulum. A decrease in the intercostal spaces is appreciated with elevation of the diaphragm in the area of the opacity (Fig. 1).

2.2. Bronchoscopy findings (Dr. Gustavo Teyssier Morales)

There was a decrease in the caliber of the trachea two rings below the subglottic region until just before the carina, which only allowed the passage of a 2.9 mm fiberoptic bronchoscope. The tracheal rings were complete and the left bronchus ended in a blind cul-de-sac; therefore, diagnosis of congenital tracheal stenosis and pulmonary aplasia was made.

2.3. Anatomic pathological findings (Dr. María de Lourdes Cabrera Muñoz)

Pulmonary aplasia is a rare congenital malformation and may be associated with malformations of the airways and other organs, among which are tracheal and renal malformations. The autopsy showed a patient of apparent age equal to chronological age with right cryptorchidism and without external malformations. Upon opening the chest cavity, there was absence of the left lung. The right lung was increased in size and displaced the heart towards the left, and there was a small left pleural cavity. The trachea showed a permeable lumen and reduction of the diameter of the lumen below the glottis. Two main bronchi originated from the carina, the left one which ended in a cul de sac; the right was elongated and morphologically straight. The right lung showed two lobules (Fig. 2).

The terms aplasia, agenesis and pulmonary dysplasia are confusing and have been used in the medical literature in an inconsistent and interchangeable manner. However, the term aplasia is preferred when there is absence of pulmonary tissue and there exists a rudimentary main bronchus, and agenesis when there is absence of pulmonary tissue and of the main bronchus.

In this case, the findings correspond to a left pulmonary aplasia with left lung pseudoisomerism. It has been reported that 70% of pulmonary aplasias are left-sided and with a prevalence in males. In up to 50% of the cases there are other associated malformations, which include tracheoesophageal fistulas, esophageal atresia, horseshoe kidney, cardiac malformations and anal atresia, among others.

Serial cross-sections of the larynx, trachea and main bronchi showed larynx with edematous mucosa and no other alterations. Reduction of 50% of the tracheal lumen in a long segment was confirmed, from the infraglottic region to the region of the carina. This was in accord with the presence of complete tracheal rings secondary to their posterior fusion in "napkin ring" instead of normal rings seen on the posterior face of the membranous part (Fig. 3).

In a microscopic study of the rings, hyaline cartilage with normal histology was seen, which confirms the diagnosis of posterior diffuse congenital tracheal stenosis in accordance with the classification proposed by Landing and Wells (Table 1).

The rare combination of tracheal stenosis and unilateral pulmonary aplasia has been recognized to be lethal and patient survival is related with the extent of stenosis.
Tracheal stenosis and unilateral pulmonary aplasia and the possibility of having corrective surgery.

The heart presented dilation of the right cavities. Pulmonary artery trunk was dilated and gave origin to the right pulmonary artery. The left pulmonary artery was not identified and a persistent left superior vena cava was noted. There was atrioventricular and arterial ventricle concordance without other alterations.

The following congenital malformations were also found in this patient: renal fusion with sole ureter and anomalous origin of the renal arteries of the terminal portion of the aorta (Fig. 4).

Well-developed parenchyma with areas of infiltrate of multifocal intraalveolar by neutrophils and recent hemorrhage is observed in the histopathological study of the right lung. The arteries and arterioles are of medium caliber. Data from pulmonary vascular disease are characterized by increase of the connective tissue of the adventitia, hyperplasia and hypertrophy of the muscle layer with reduction of the lumen (Fig. 5).

Secondary to the outpatient event of prolonged cardiac arrest, the patient developed ischemic hypoxic encephalopathy with cerebral edema and encephalomalacia (Fig. 6), morphological data of liver shock, myocardium, intestine and acute tubular necrosis. Sepsis diagnosis was made by the presence of acute pneumonia, acute laryngo tracheitis and postmortem cultures positive in blood and intestines for Klebsiella pneumoniae.

3. Final diagnoses

Left pulmonary aplasia with ipsilateral pulmonary artery agenesis associated with diffuse posterior intrinsic tracheal stenosis (in “napkin ring”).

Associated malformations

• Left bronchus in blind pouch
• Left lung pseudoisomerism
  — Long right main bronchus
  — Right lung with two lobes
• Persistent left superior vena cava
• Anomalous origin of renal arteries of terminal aorta
• Renal fusion with sole ureter

Concomitant alterations

• Grade B pulmonary vascular disease
• Dilatation of the trunk of the pulmonary artery and right cavities

Table 1

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<th>Classification of congenital tracheal stenosis.</th>
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<td>Generalized or diffuse (30%)</td>
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<td>Diffuse posterior in “napkin ring”</td>
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<td>“Funnel”-shaped stenosis (20%)</td>
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<td>Segmented stenosis (50%)</td>
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<td>Tracheal cartilaginous sleeve (rare)</td>
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<td>Extrinsic</td>
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<td>Arterial and aortic malformations</td>
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Figure 4  Anterior and posterior aspects of the fused kidneys. Ureter originates from the collecting system. The renal arteries originate from the terminal aorta.

Figure 5  (A) Well-developed lung parenchyma (HE ×20). (B) Area of pneumonia with multiple intra-alveolar polymorphic nuclei. (C) Hyperplasia of the media and increased connective tissue in the adventitia of the arteries secondary to pulmonary vascular disease.
in pressure. Due to having only one pulmonary artery that the agenesis or pulmonary malformations originating case is found within the first category, which encompasses into ten categories in accordance with the etiology. This pulmonary hypertension in a pediatric patient is divided according to the last consensus done in Panama (2011), Orellana)

In analyzing this case, it is believed that the patient did not allow for a complete study and caused his death. The youngest patient operated post-unwitnessed cardiac arrest 15 min. In this case the resuscitation lasted 20 min so the Association recommends carrying out resuscitation for those who should definitely not be resuscitated. As to the time, the American Heart to the hospital. Therefore, there were indications that he had no rigor mortis or body temperature is one of the important pieces of evidence that showed an increase in mortality. Another possibility is to form a consensus among the different services of the hospital who use the drug in order to determine whether or not it should be used and in which cases. The vast majority of patients are asymptomatic throughout their lifetime. The most common is the presence of heart diseases that cause a pulmonary hyperflow and heart diseases with elevated pulmonary pressure; however, it should not be indicated in congenital malformations. The most common is the presence of congenital tracheal stenosis, limited his ventilation allowed him to live a normal life with certain restrictions. However, the second, a diffuse congenital tracheal stenosis, limited his ventilation and favored pulmonary infections for which some type of surgical intervention would have been required to increase his ventilation but due to the lung infection he should have been re-assessed prior to the studies being done through a sternotomy and with extracorporeal axial tomography with reconstructions that allows us to plan the most appropriate surgical technique due its nature. There are two ways to arrive at the diagnosis of these heart diseases that cause a pulmonary hyperflow and heart diseases with elevated pulmonary pressure. Diagnosis of pulmonary hypertension is made using echocardiogram. Normal pressure is 25 mmHg at rest, between 25 to 40 mmHg is considered to be mild hypertension, and 40 to 60 mmHg as moderate hypertension, and above 60 mmHg as severe hypertension. Sildenafil is a selective pulmonary dilator and has been used for heart diseases with elevated pulmonary pressure; however, it should not be indicated in congenital malformations. The morphological data of multivisceral shock presented by the patient. It is preferable that the patient died due to sepsis. The youngest patient operated was 1 month of age. These children are seen weekly to monitor their progress. In case there is increased respiratory difficulty or problems with feeding, they are scheduled for surgical intervention would have been required to increase his ventilation but due to the lung infection he should have been re-assessed prior to the studies being done through a sternotomy and with extracorporeal axial tomography with reconstructions that allows us to plan the most appropriate surgical technique due its nature. Patients should have computerized tomography, magnetic resonance, color Doppler, and echocardiograms.
Tracheal stenosis and unilateral pulmonary aplasia

Conflict of interest
The authors declare no conflict of interest of any nature.

References