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 Mexico 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...
The incidence of tracheal stenosis is frequently on the left side.

Both malformations occur more often in the fifth weeks of gestation. "Tracheal aplasia where there is a rudimentary bronchial growth of bronchial structures, arteries and vein, or pulmonary agenesis when there is absence of lung tissue and of the pulmonary segment of the bronchial tree and its ramifications." 2

We are discussing the case of a male patient, 2 months old at admission due to respiratory distress and hypoxia. Twenty hours after his admission, the patient presented a picture of neonatal sepsis with a temperature of 32°C and weight of 3100 g. Resuscitation maneuvers were begun: 10 cycles of external chest compressions, six doses of adrenaline and percutaneous cardioversion. After 6 min of resuscitation he was without respiratory effort. There was no heart beat and peripheral pulses were absent. After 30 min of resuscitation, there was a return of spontaneous breathing with a respiratory rate of 40 cycles/min and a pulse of 90 cycles/min. He presented three episodes of hypoxemia and bradycardia. Despite ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, the patient died 3 days after his admission.

The patient had a diagnosis of two major airway malformations. Congenital tracheal stenosis and unilateral pulmonary aplasia. Symptoms include biphasic stridor, cough, pallor and cyanosis. Although the discharge secretions were described as having data of systemic inflammatory response syndrome (SIRS), the patient had no signs of infection. There was no heart beat and peripheral pulses were absent. After 30 min of resuscitation, there was a return of spontaneous breathing with a respiratory rate of 40 cycles/min and a pulse of 90 cycles/min. Despite ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, the patient died 3 days after his admission.

In short, this is the case of a malnourished infant who presented with a new picture of pulmonary infections after his last admission. The patient was without respiratory effort. There was no heart beat and peripheral pulses were absent. After 30 min of resuscitation, there was a return of spontaneous breathing with a respiratory rate of 40 cycles/min and a pulse of 90 cycles/min. Despite ventilation, cefotaxime, oseltamivir, platelet and plasma transfusions, and inotropic infusions and ventilation, the patient died 3 days after his admission.

The incidence of tracheal stenosis is frequently on the left side. Congenital tracheal stenosis and unilateral pulmonary aplasia. Symptoms include biphasic stridor, cough, pallor and cyanosis. Although the discharge secretions were described as having data of systemic inflammatory response syndrome (SIRS), the patient had no signs of infection.
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).
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>Intrinséc</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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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 manages the total pulmonary output, it causes an increase in utero. Case is found within the first category, which encompasses ten categories in accordance with the etiology. This pulmonary hypertension in a pediatric patient is divided into account that he also had pulmonary aplasia; therefore, it was not possible due to the lung infection. He should have been reassessed prior to the studies being completed and prior to operation. Unfortunately, it was not taken into account that he also had pulmonary aplasia. However, it was not taken surgery as quickly as possible. Difficulty or problems with feeding, they are scheduled for monitoring their progress. In case there is increased respiratory weight, studies have been completed and that other children have several months of life and be of adequate symptoms presented by the patient. It is preferable that dependencies on the severity of the stenosis and on the axial tomography with reconstructions that allows us to plan surgery. Five of these surgeries have been carried out in extracorporeal circulation. The recommended surgery is a slide tracheoplasty, which is done through a sternotomy and with extracorporeal as a high-risk surgery. When and for how long the resuscitation maneuvers should be carried out. Those who should definitely not be resuscitated are those who have been dead for several hours. When a patient arrives from Huixquilucan, cardiac arrest occurred en route to the closest emergency room so that once stabilized, transfer would indicate death. However, he had no rigor mortis or lividity. It was taken into consideration that because he did not allow for a complete study and caused his death. The age at which these patients should be intervened depends on the severity of the stenosis and on the etiology. This patient had a temperature of 32ºC, which showed an increase in mortality. Another possibility is to form a consensus among the different services of the forum. There is a recommendation from the FDA published this year on the prohibition of the use of sildenafil in neonates that its use increased mortality; therefore, it should only be used in individual patients and with strict monitoring. There are two ways to arrive at the diagnosis of these heart diseases that cause a pulmonary hyperflow and heart diseases which management is surgical. In this case its use was highly questionable because there are no studies to prove benefit for the most appropriate surgical technique due its nature. There are two possibilities: 1) production of an increase in pulmonary flow and 2) heart diseases that cause a pulmonary hyperflow. The vast majority of patients are asymptomatic throughout their lifetime. When and for how long the resuscitation maneuvers should be carried out. Those who should definitely not be resuscitated are those who have been dead for several hours. Another possibility is to form a consensus among the different services of the forum. Another possibility is to form a consensus among the different services of the forum. Another possibility is to form a consensus among the different services of the forum. Another possibility is to form a consensus among the different services of the forum.
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Conflict of interest
The authors declare no conflict of interest of any nature.

References