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Case Report

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Scimitar syndrome: case report

Abstract

This clinical case is about a 5-month-old asculin, born full-term, with weight and size suitable for gestational age, APGAR 9/9, no suffering, no resuscitation, and no pathological or surgical history known. Consults the emergency department of the National Children's Hospital after one week of respiratory symptomatology, with frank clinical data of respiratory distress: intercostal retractions, tachypnea, and nasal flutter. Required the ingress use of high oxygen flow cannula to stabilize respiratory distress; in addition to negative response to therapeutic tests with salbutamol. Chest x-ray, among other findings, documents a slight erasure of the cardiac silhouette and dextrocardia; ultrasound can document the presence of pleural effusion, as well as right atelectasis data observed in bronchogram; as part of the initial management plan it is decided to intern the patient for hospital management. During internment, dextrocardia is confirmed by echocardiogram, in addition to right pulmonary vessel hypoplasia, computed axial tomography of cardiac vessels is performed describing it as a result: right pulmonary hypoplasia, abnormal venous drainage of the right lung to the lower vena cava, right intralobar pulmonary abduction, hypoplasia of the right pulmonary artery, thus allowing to document the presence of a Chimitarra syndrome. Management is given by the cardiology service, where the findings are confirmed and the list of problems is reorganized, in this way it is established as problem one to the diagnosis of Chimitarra Syndrome. The case is presented in clinical session to define surgical management of the same.

Keywords: scimitar syndrome, angio-tac, pulmonary hypoplasia, dextrocardia

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Introduction

Scimitar syndrome is a group of unusual congenital anomalies that include total or partial abnormal pulmonary venous drainage between the right lung and inferior vena cava, usually associated with hypoplasia of the right lung and right pulmonary artery.[1] Venous drainage is usually to the inferior vena cava below the hemidiaphragm, although very diverse drains have been described. The bronchial tree may be abnormal and, when it happens on the right, may resemble that of the left lung.[2] The sign was described by Dotter in 1949,[1] but Halasz in 1956 and Neill et al.[3] in 1960, named the syndromeme, however, similar findingswere described in autopsies of 3 children, two of them in 1836 and one in 1912. According to the literature, the incidence is 1 to 5/100 000 live births. Although frequently diagnosed in childhood, the patient may remain asymptomatic into adulthood.[4]

With a prevalence of 0.31 cases per 10000 births, for an equivalent of 2 cases reported during the last year in Costa Rica, this according to the latest annual report of the Specialized Surveillance Unit on congenital defects.[5] The importance of publicizing this case lies in the fact that it is captured early, which allows a timely correction of the structural anomalies described, in addition to the use of computerized axial tomography of cardiac vessels as a diagnostic tool.

Background

This article sets up and exemplifies the execution or start-up of tomographies synchronized with the heart and breathing in minors with low voltage to avoid a low forecast and as a same and the same percentage and in seventy percent of the frequency. Cardia allow us to see and show the real anatomy that cardiac surgeons and cardiologists see themselves every day, since tomography allows us to calculate the left ventricular ejection fraction, right ventricular ejection fraction, atrial volumes, and see the real anatomy as a whole, because many of the pathologies such as patent foramen ovale, ventricular septal defects, anomalous venous drainage, are associated with other malformations, whether in the pulmonary arteries or in the pulmonary veins or the airway. Therefore, it is always important to use a technique that encompasses and involves the anatomical and functional information of the thorax and heart.

Case report

This is a 5-month-old male, born term of mother G2 P2 A0 C0, in vaginal delivery conducted, 39 + 5 weeks, birth weight: 3035 gr, height 47.5 cm, APGAR 9 /9, no suffering, no resuscitation.

Patient consults the emergency department on 2-11-19 with a one-week history of respiratory symptoms. Upon admission, the patient looked pale with significant respiratory distress, intercostal retractions, tachypnea, nasal flutter, rhythmic heart sounds without better audible murmurs in the right hemi thorax, and she did not have hepatomegaly. Peripheral pulses were normal in all extremities. He required a high-flow cannula to stabilize his respiratory failure.

Therapeutic testing with salbutamol and radiography was performed, but patient persisted with respiratory distress and

The chest X-ray showed a right Para cardiac infiltrate that partially erased the cardiac silhouette, the trachea was shifted to the right, there was complete radio-opacity of the right hemithorax, elevation of the right hemidiaphragm, and dextrocardia. Chest ultrasound was performed, where pleural effusion was ruled out, bronchogram and data compatible with right atelectasis were observed. Therefore, it was decided to hospitalize in the medicine service for intravenous oxygen therapy and antibiotic therapy with the diagnosis of right bronchopneumonia.

The initial echocardiogram showed dextrocardia, situs solitus, atrio ventricular and arterial ventricle concordance, normal atrioventricular and semilunar valves, intact interatrial and interventricular septa. There was no dilatation of chambers and contractile function was normal, the right pulmonary branch looked hypoplastic suggesting hypoplasia of the right lung, and scimitar syndrome is suspected however the study was limited by the patient's condition; CT

angiography was suggested when the condition improved in the minor. Finally, chest CT angiography was performed (8/11/19) where right pulmonary hypoplasia, abnormal venous drainage from the right lung to the inferior vena cava, right intralobar pulmonary sequestration, hypoplasia of the right pulmonary artery were documented. It is concluded that the patient is a carrier of Scimitar Syndrome. He is evaluated again by cardiology confirming the findings of total anomalous drainage from the right pulmonary veins to the inferior vena cava without obstruction in its course. There were no data of severe pulmonary arterial hypertension or dilation of right chambers. The case was presented in cardio-surgical session to be taken to surgery for correction. Patient currently awaiting surgery. Cardiac catheterization was not performed because echocardiogram and CT angiography were clear with diagnosis and hemodynamics.

Discussion

The scimitar syndrome or also called congenital venolobar syndrome , where there is a congenital anomaly of the right pulmonary veins to the inferior vena cava or the right atrium[6] consists of three findings: right pulmonary hypoplasia, partial pulmonary venous return anomaly, and unilateral pulmonary sequestration. Sometimes not all of these features are present, and other heart abnormalities, such as hypoplasia of the left heart or aorta, may also be detected.[7]

With a very varied clinical presentation, with a large number of signs and symptoms that can make diagnosis difficult in children and young adults with congenital heart disease.[8] As in the case presented, which is given an initial management, oriented to respiratory pathology.

From the radiological point of view, the following characteristics are presented: a small ipsilateral lung with an ipsilateral mediastinal displacement. The abnormal pulmonary vein drains and enlarges into the diaphragm in a "scimitar" shape.[9] However, not all the characteristics tend to be present, the presence of dextrocardia together with the partial effacement of the cardiac silhouette were suggestive to raise the presence of scimitar syndrome in the case presented.

Normally, surgery for scimitar syndrome can be postponed beyond the neonatal period or even beyond childhood. In addition to the possibility of developing pulmonary hypoplasia, arrhythmias and infections, most affected infants do well.[10]

Scimitar syndrome may remain undiagnosed into late adulthood and present withpulmonary hypertensiveness and RV insufficiency. Careful evaluation of cardiopulmonary structures and the presence of a left-to-right short circuit should be part of the evaluation of all adult patients diagnosed with PAH and RV failure.[11]

It is important to recognize the radiological features and anatomical associations of scimitar syndrome, as features of the primary condition and associated abnormalities may have implications for surgical treatment.[12] Therefore, the use of diagnostic tools such as computerized axial tomography of cardiac vessels plays a fundamental role in the characterization of defects and, together with the echocardiogram, was sufficient for diagnosis, without the need to submit the patient to cardiac catheterizations.

Cardiac catheterization is the invasive method par excellence for the accurate diagnosis of the syndrome, since it is possible to clearly assess abnormal venous drainage. In addition to performing relevant measurements with pressures (mainly of the pulmonary artery) and gradients, it is useful to rule out other associated malformations.[13]

Definitive treatment remains surgical, redirecting the anomalous pulmonary vein or veins into the left atrium. Alternatively, palliation by percutaneous closure of aortopulmonary collateral flow in infants with scimitar syndrome may markedly improve congestive heart failure, reducing pulmonary overcirculation, dilation of right cavities, and delaying cardiac surgery for at least the first few years of life.[14]

Surgical treatment is reserved for hemodynamically significant cases[15] (Figure 1 & 2).

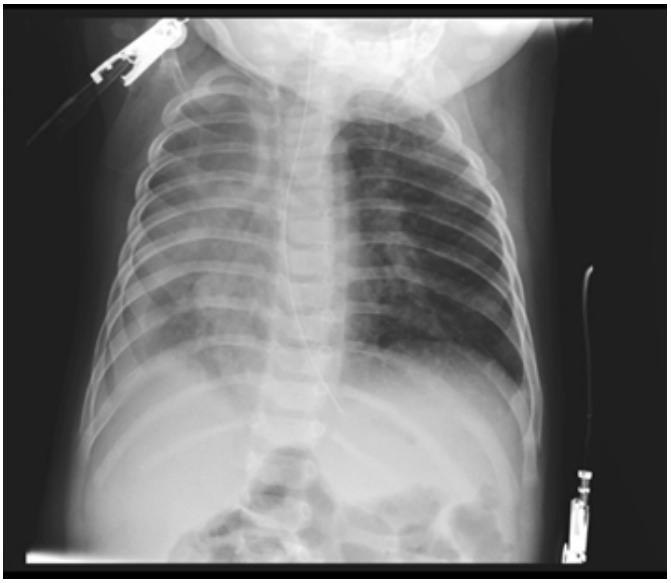

Figure 1 Chest X-ray showing radio opacity of the right hemi thorax of confluent appearance, suggesting an atelectasis process, versus right pulmonary hypoplasia, showed left contralateral lung with increased radius lucidity in relation to compensatory emphysema with contralateral deviation of the cardiac silhouette.

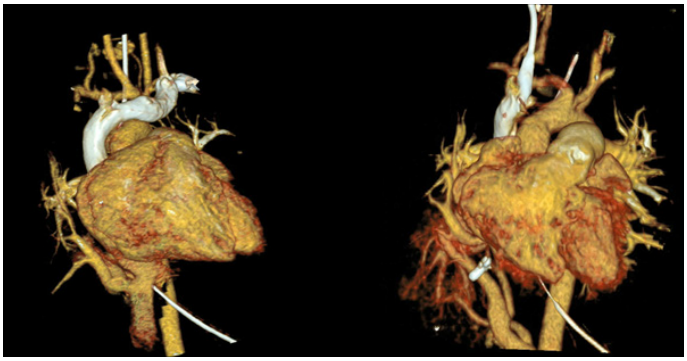

Figure 2 Volumetric tomography of cardiac silhouette and large vessels, where there is evidence of right superior vena cava without malformations, pulmonary artery of usual arrangement, the emergence of the right pulmonary vein of the inferior vena cava is observed, findings in relation to Scimitar Syndrome.

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Conflicts of interest

The authors deny conflicts of interest.

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