June 2020, Volume 5, Issue 1. e131488

Scimitar Syndrome Issued from a Consanguineous Marriage: A Rare Congenital Cardiomyopathy

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DOI: 10.5812/intjcardiovascpract-131488

Submitted: 29-12-2019
Accepted: 27-02-2020

Keywords:
Scimitar Syndrome
Consanguinity
Cardiomyopathies

Abstract

We present here a rare case of Scimitar syndrome on a full-term newborn female issued from consanguineous marriage and diagnosed at the age of two months who was suffering from polypnea since the age of two weeks. She was admitted at the age of two months for polypnea, non-dysmorphic facies, healthy systemic arterial tension, regular pulse, but the family history raises that before her birth, two newborns respectively boy and girl died both at an early age with dilated cardiomyopathy. After hospitalization, we realized chest radiography, which has shown opaque right lung and suspicion of scimitar syndrome. After that, an echocardiogram was performed, showing severe hypoplasia of the right pulmonary artery with pulmonary hypertension. The Thoracic computed tomography confirmed the diagnosis by revealing hypoplasia of the right pulmonary artery with pulmonary arterial hypertension, right pulmonary hypoplasia (bilobar), and abnormal partial right pulmonary venous return toward the vena cava under the diaphragm.

INTRODUCTION

Scimitar syndrome is rare congenital cardiomyopathy mostly diagnosed at the childhood rarely till adulthood. It is defined as congenital pulmonary venolobar syndrome unusual the venous return from the right lung, which is abnormal to one of the systemic venous rather than to the left atrium [1]. In our case, the anomaly of the pulmonary venous return is partial as the common Scimitar syndrome. In other instances, the defect interests the total pulmonary venous returns. The diagnosis was suspected on a chest X-ray, which showed the Scimitar shape confirmed by echocardiogram and computed tomography (CT) scan.

CASE PRESENTATION

Here we present a two months female issued from consanguineous marriage, with the antecedent of two neonatal deaths of females and males respectively by dilated cardiomyopathy. The first sign was polypnea; the blood pressure and pulse were normal. The chest X-ray evocated the diagnosis of Scimitar syndrome (Fig 1). The echocardiogram demonstrated a right ventricle moderately dilated and enlarged with good function, tricuspid insufficiency estimating the systolic pressure of the right ventricle at 54 mmHg. Slightly dilated pulmonary artery trunk with normal sized left pulmonary artery and severe hypoplasia of the right pulmonary artery, which measures 4 mm (Fig 2A, B). Thoracic CT scan showed dilation of the trunk of the pulmonary artery measured in 12.6 mm, hypoplasia of the right pulmonary artery which is small and measures 4.4 mm but permeable giving birth to 2 lobar branches for the middle and lower lobe with agenesis of the upper lobar bronchus, left pulmonary artery of normal size permeable, partial abnormal right venous return towards the inferior vena cava, with scimitar sign, dilation of the right heart chambers with ratio right ventricle/ left ventricle > 1, paradoxical septum and

small right lung with agenesis of the upper lobe with the attraction of the elements of the mediastinum on the right (Fig 3A-E). Repairing the anomaly can be performed by several methods: reorienting the blood flow, managing cardiac anomalies, and ligating anomalous blood supply [2]. In our patient, the condition was fully explained to the family, and at present conservative approach was considered.

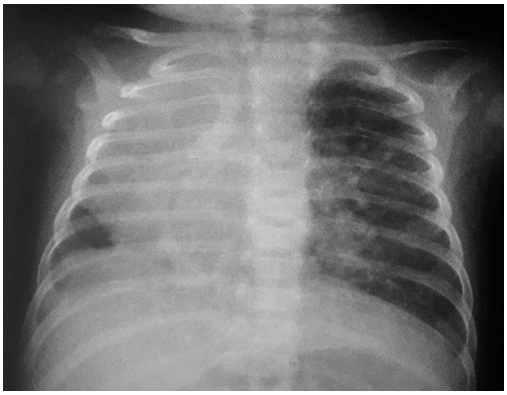

Figure 1. Chest X-ray; The heart is moved into the right chest; the right lung is hypoplasic.

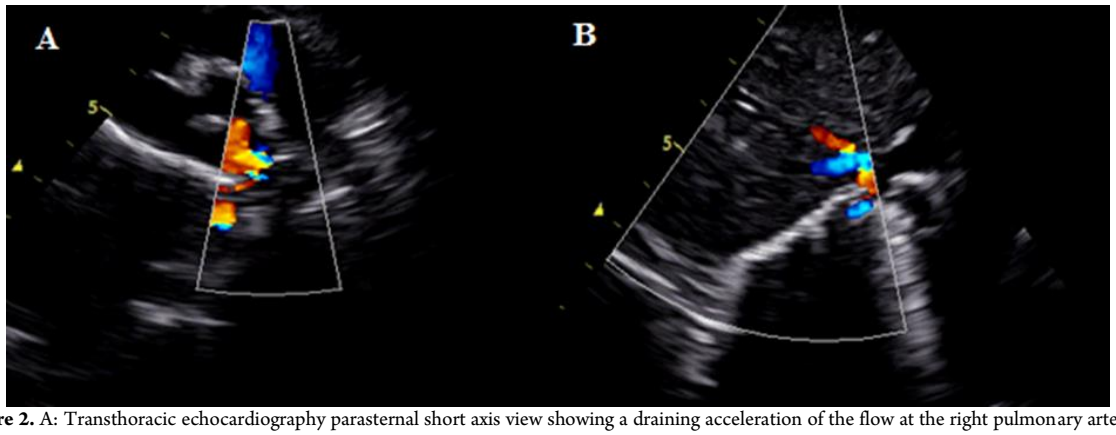

Figure 2. A: Transthoracic echocardiography parasternal short axis view showing a draining acceleration of the flow at the right pulmonary artery. B: Transthoracic echocardiography showing the scimitar vein into the inferior vena cava on a 2-dimensional image

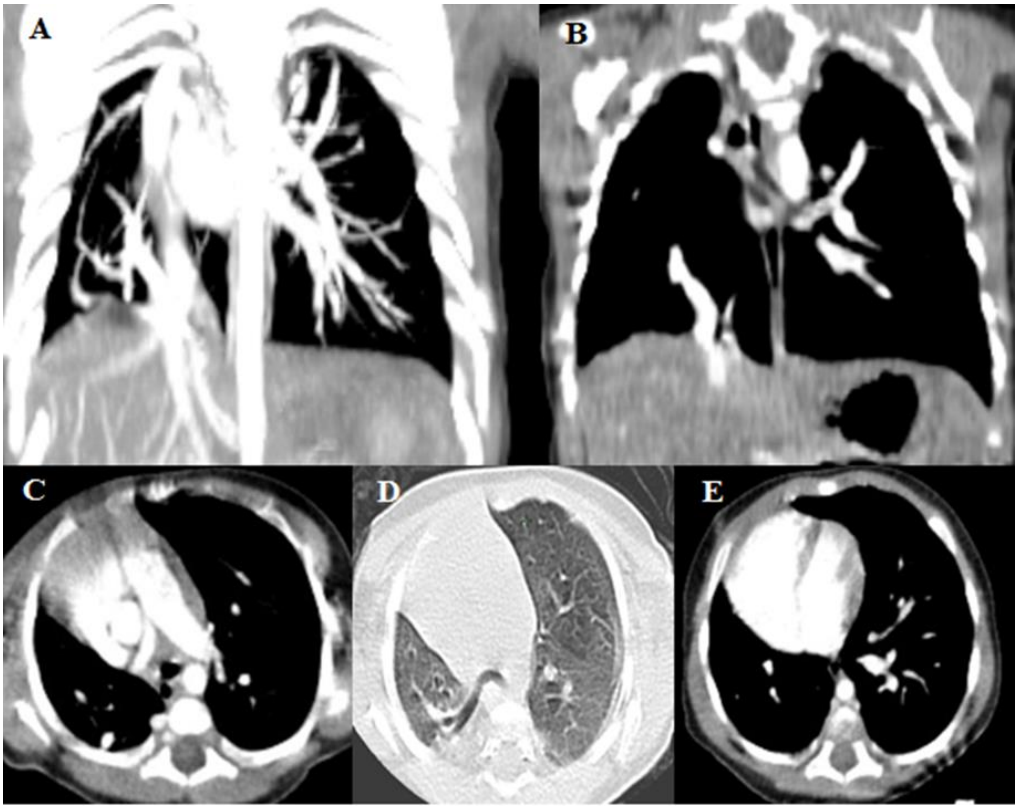

Figure 3. A, B: computed tomography scan (Coronal section) showing an abnormal right pulmonary venous return to the interior vena cava C: Hypoplasia of the right pulmonary artery D: Right pulmonary hypoplasia E: Dilatation of cardiac cavities, paradoxical septum

DISCUSSION

Symptoms of scimitar syndrome are determined by the age of diagnosis and may include problems of respiratory, growth, and cardiac systems. The treatment is based mostly on surgical intervention with a high risk of mortality [3, 4]. The anomaly is characterized by hypoplasia of right pulmonary artery and right lung replacing cardiac structures to the right as a result of the abnormal insertion of the right pulmonary vein into the inferior vena cava, taking the shape of scimitar sword from where the name. The diagnosis is based on chest X-ray which may present a characteristic Scimitar sign, beside echocardiogram, angiography, computed tomography scan, and magnetic resonance imaging, which is an excellent non-invasive exam to describe such a complex congenital defect well [5-8]. Often, patients with scimitar syndrome remain without symptoms for many years before the declaration of pulmonary artery hypertension and right ventricle failure. When there is an association with pulmonary sequestration or repeated lung infections, surgical treatment may be needed while it is not required for asymptomatic patients [9].

The surgical correction case by case is the definitive therapy, which indication depends on the shunt size, presence or not of pulmonary artery hypertension, right ventricle failure, and eventual hepatic cirrhosis.

CONCLUSION

Scimitar syndrome is a rare congenital anomaly that can be declared early in the majority of cases as well as at advanced age. The prognosis depends on early diagnosis and the surgical strategy. The results tend to be promising with a low incidence of morbidity and mortality

Conflict of Interest

None

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