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Case Report

Scimitar Syndrome with Tetralogy of Fallot: A Very Rare Association

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Abstract

Scimitar syndrome is a rare variant of partial anomalous pulmonary venous connection. The association of Scimitar syndrome with another cardiac congenital anomaly such as tetralogy of Fallot is very rare; It may proceed unnoticed, as cyanosis already predominates in the clinical picture. Data about optimal therapeutic strategy in this rare variant of SS are scarce. We are reporting the case of an eighteen-month old boy diagnosed with Scimitar syndrome and tetralogy of Fallot treated with pneumonectomy.

Keywords: *Scimitar Syndrome; Scimitar Vein; Tetralogy of Fallot; Anomalous Pulmonary Venous Drainage*

Abbreviations

ASD: Atrial Septal Defect; ECHSA: European Congenital Heart Surgeons Association; CT: Cardiac Tomography; IVC: Inferior Vena Cava; LA: Left Atrium; LV: Left Ventricle; LPA: Left Pulmonary Artery; PH: Pulmonary Hypertension; PDA: Patent Ductus Arteriosus; RPA: Right Pulmonary Artery; RV: Right Ventricle; SS: Scimitar Syndrome; SV: Scimittar Vein; TAPSE: Tricuspid Annular Plane Systolic Excursion; TOF: Tetralogy of Fallot; VSD: Ventricular Septal Defect

Introduction

Scimitar syndrome (SS), also known as pulmonary venolobar syndrome or hypogenetic lung syndrome, is a rare congenital heart malformation characterized by an anomalous vein that drains part (usually the right lower lobe) or all of the right lung blood flow into the systemic venous circulation, usually the inferior vena cava (IVC), associated with an ipsilateral pulmonary hypoplasia, underdevelopment of the right pulmonary artery (RPA), and sometimes an anomalous systemic arterial supply from the descending aorta or its branches to the hypoplastic lung supplying aberrant nonfunctioning lung parenchyma (pulmonary sequestration) [1-3].

The association of SS with another cardiac congenital anomaly such as tetralogy of Fallot (TOF) is very rare [4]. Very few published cases of SS and TOF have discussed therapeutic strategies and related outcomes and prognosis in this rare variant of SS.

We are reporting the case of an eighteen-month old boy diagnosed with SS associated to TOF treated by pneumonectomy and discussing outcomes of therapeutic strategies according to literature review.

Case presentation

- Patient information

We describe the case of an 18 months-old Moroccan infant with a history of recurrent respiratory infections, who was referred to our

department with cyanosis, dyspnea, and feeding difficulties. His parents were not related. He was a product of full term normal vaginal delivery and the first child in his family. He was not on any medication at the time of presentation.

- Clinical findings

On physical examination, he appeared to be a well-grown and healthy appearing boy with a weight of 10 kg. His blood pressure was 90/60 mmHg and his pulse rate was 100 beats per minute (bpm). His oxygen saturation was 70% on room air. He had a 3/6 ejection systolic murmur, there was no hepatomegaly and peripheral pulses were palpable. Neurological and general physical examination were normal.

- Diagnostic assessment

An echocardiographic examination showed normal abdominal and atrial situs, dextroposition of the heart in the right side of his chest, normal atrioventricular relationship, a TOF, with a large subaortic ventricular septal defect (VSD) with a right-left shunt, an overriding aorta, a valvar pulmonary stenosis, continuous wave Doppler interrogation across the stenosis showed a peak systolic pressure gradient at 49,7 mmHg without other subvalvular or supravalvular stenosis, Right (RPA) and left pulmonary arteries (LPA) were confluent with a relatively hypoplatsic RPA measuring 4.3mm vs 6mm for LPA. The right ventricle looked markedly dilated and hypertrophied with grossly good systolic function on visual inspection and tricuspid annular plane systolic excursion (TAPSE) (Figure 1).

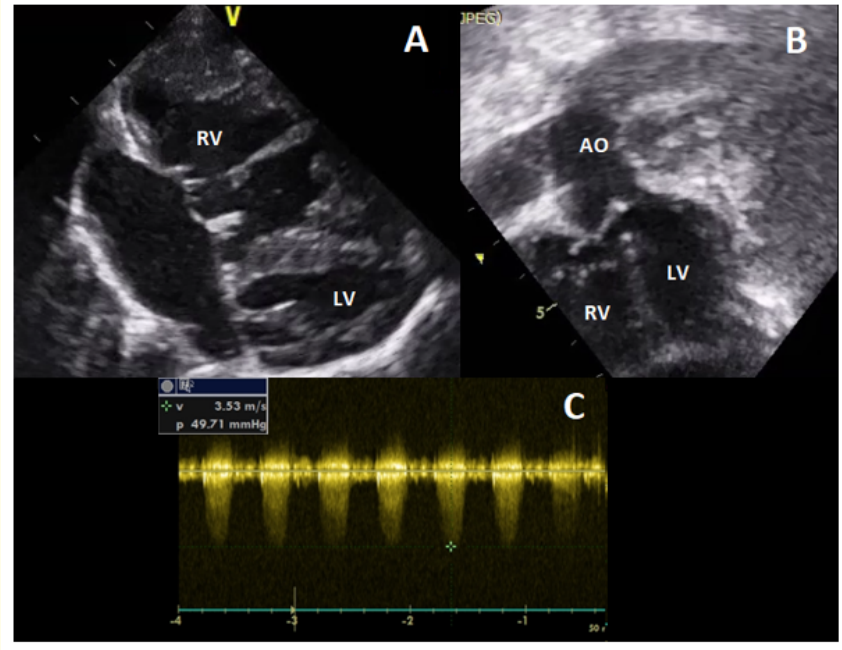

Figure 1: *Transthoracic echocardiography (A): Subcostal four chamber view showing a dilated and hypertrophied RV; (B) Subcostal view showing an overriding aorta with a large subaortic VSD; (C): continuous wave Doppler on pulmonary valve showing a peak systolic pressure gradient at 49,7 mmHg suggestive of pulmonary valvular stenosis. *RV: Right ventricule; LV: Left ventricule; Ao: Aorta; VSD: Ventricular septal defect.*

Echocardiography showed only left pulmonary veins drainage into the left atrium while drainage of right pulmonary veins towards the left atrium was not seen, with an unexpected flow towards the inferior vena cava raising the suspicion of partial abnormal pulmonary venous return (Figure 2). In addition, echocardiogram revealed a persistent left superior vena cava drained intro a dilated coronary sinus, and a patent ductus arteriosus (PDA). The left ventricle was not dilated nor hypertrophic, with a paradoxical interventricular septal motion and preserved global systolic function. The mitral and aortic valves were structurally normal. The interatrial septum appeared intact, there was no other associated VSD, and the aortic arch was left-sided and had no coarctation.

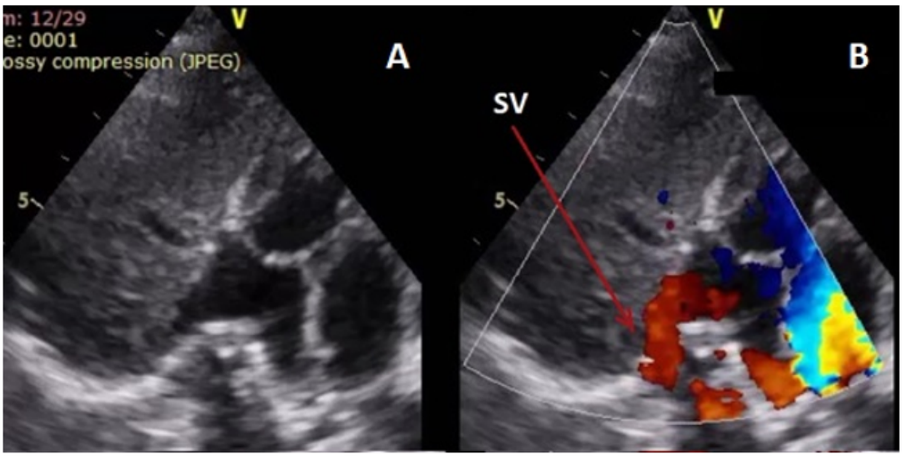

Figure 2: *Transthoracic echocardiography subcostal bicaval view (A) 2D and (B) with color Doppler showing a vascular sctructure with a directed flow towards the inferior vena cava suggestive of scimitar vein. *SV: Scimittar vein.*

Chest computed tomography (CT) was performed to clarify the anatomical connections. It showed normal left pulmonary venous drainage into the left atrium (LA) and abnormal right pulmonary venous drainage through a descending vein (scimitar vein) connected to the inferior cavo-atrial junction (Figure 3), with hypoplasia of the right pulmonary artery, which measured approximately 4.5 mm with hypoplasia of ipsilateral lung (Figure 4). The right lower lobe of the right lung was sequestrated, and supplied by a large collateral arising from the descending aorta below the diaphragm which is divided into two large collaterals (Figure 5).

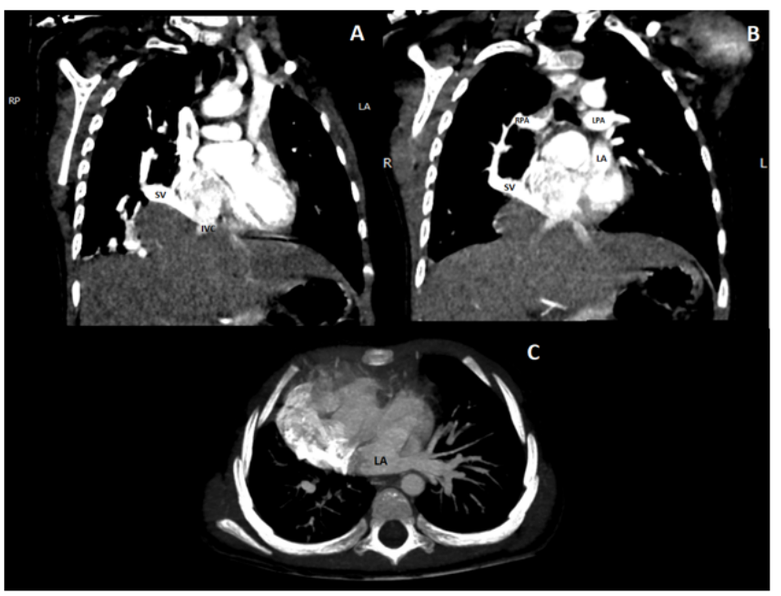

Figure 3: *Chest angio-CT, (A) and (B): Coronal sections showing an abnormal right pulmonary venous drainage through a descending vein connected to the inferior cavoatrial junction; (C): Axial section showing a normal left pulmonary venous drainage into the left atrium *LA: left atrium; RPA: right pulmonary artery; LPA: Left pulmonary artery; SV: scimitar vein; IVC: inferior veina cava.*

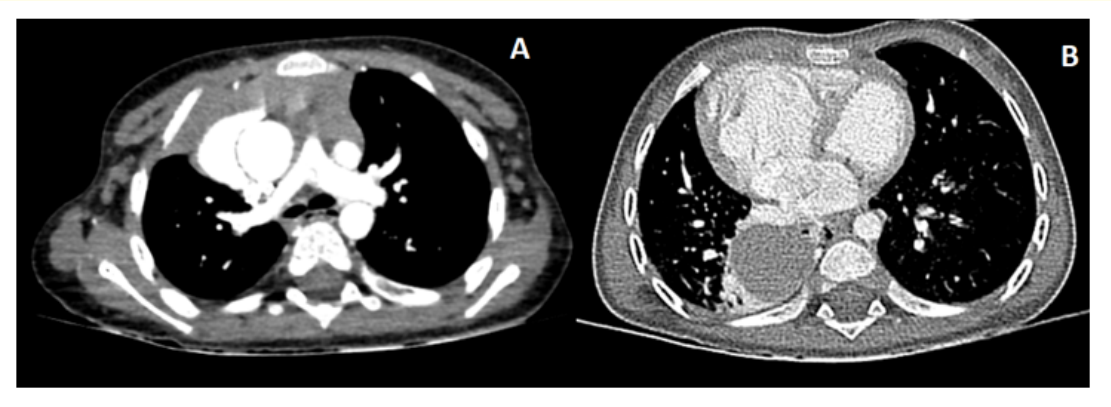

Figure 4: *Chest angio-CT axial sections, (A): hypoplasia of the right pulmonary artery, which measured approximately 4.5 mm with hypoplasia of ipsilateral lung; (B): sequestration of the lower lobe of the right lung.*

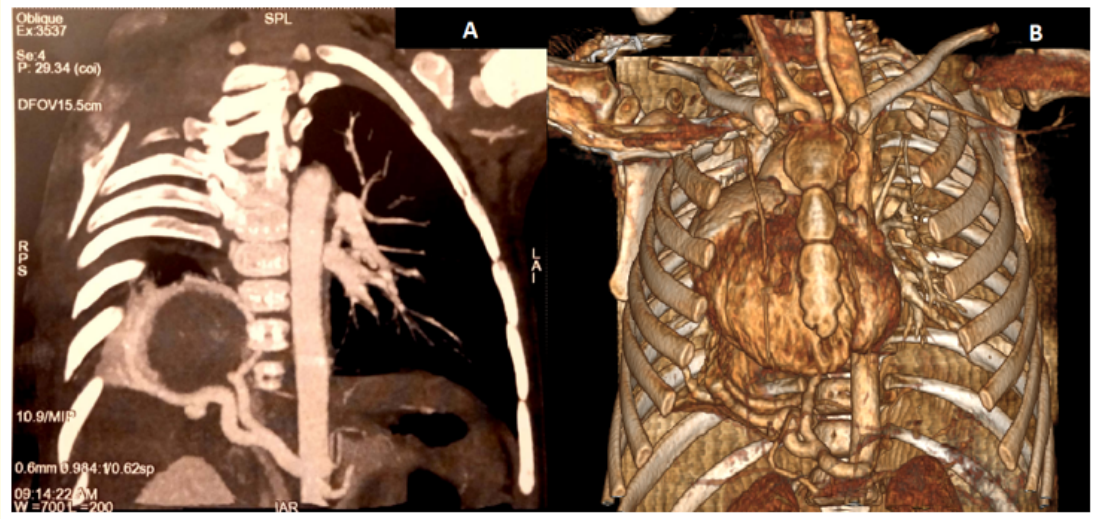

Figure 5: *Chest angio-CT, (A) coronal section and (B) 3D angiographic reconstruction showing a sequestration of the right lower lobe supplied by a large collateral arising from the descending aorta below the diaphragm.*

- Therapeutic intervention, follow-up, and outcomes

After longitudinal sternotomy; systemic heparinization, the patient underwent under cardiopulmonary bypass a right pneumonectomy with ligature of abnormal aorto-pulmonary collateral arteries as a primary surgical time. Repair of TOF was planned in a secondary operative time. The patient's clinical condition deteriorated during the early postoperative period and death occurred 3 days after the surgery. An autopsy was not performed.

Discussion

Scimitar syndrome (SS) is a rare variant of partial anomalous pulmonary venous connection. It associates an anomalous vein that drains part or all right lung with an hypoplasia of the ipsilateral lung and pulmonary artery and sometimes an anomalous systemic arterial supply [5].

In general, the most commonly associated congenital heart malformation in SS is persistent left superior vena cava draining to the coronary sinus [6]. Atrial septal defect (ASD) is also frequently associated to SS (60%–65% of all the patients). Ventricular septal defect (VSD) and PDA are present (alone or in association) in about a quarter of patients with SS. Other congenital heart disease are rarely associated (< 10%) to SS, like aortic coarctation and valvular disease [7]. The association with TOF is even more rare [8-10]. To the best of our knowledge, less than ten cases of TOF associated to SS have been reported in literature [11].

Isolated forms of SS may be asymptomatic for a very long time. Symptoms are more frequent when SS is associated with other malformations or in the presence of lung sequestration [7]. In our case, the clinical presentation consisted of cyanosis, dyspnea and recurrent infection. As patients with TOF are usually cyanotic, associated abnormal pulmonary venous connections are infrequently suspected. However, recurrent pulmonary infection is not usual in the setting of TOF and should led to search for other associated malformation; in our case these infections were explained by the presence of SS with sequestrated lung tissue. The sequestrated lung is vulnerable to surinfection [3]. Our patient had the typical anatomic description of SS associated to TOF, PDA and persistent left superior vena cava drained into the coronary sinus.

The diagnosis of SS when associated to TOF, can be suspected on chest X-Ray showing the scimitar sign [12]. Echocardiography is a simple non-invasive tool, that should be used to evaluate intra-cardiac structures and to precisely track all pulmonary venous returns including connections to the IVC. The detection of an abnormal flow-connection to IVC on echocardiography may lead to suspect SS, particularly in patients with TOF [8,13,14]. In most cases, the diagnosis is established by non-invasive techniques (CT angiogram and angio-MRI) to evaluated intra and extra-cardia structures and clarify anatomical connections. The diagnosis is complemented by cardiac catheterization only in some cases [15,16].

The treatment of SS in symptomatic patients is generally based on the repair of the anomalous venous drainage by redirecting the scimitar vein into the left atrium and ligation of systemic arterial supply [17]. Occlusion of systemic arteries feeding the pulmonary sequestration allow to reduce the blood flow, the amount of shunting [18,19], the pulmonary arterial pressure [20,21], and thus improve clinical symptoms in infants with congestive heart failure. The main post-operative complication after SV re-routing is stenosis or occlusion of the SV anastomosis. It occurs in almost 25% of SS patients, more frequently in young children and requires interventional cardiac catheterization or reoperation [22].

Right lung lobectomy or pneumonectomy can be considered; as a first line surgical strategy or a secondary resort after a failed repair [23,24]; in patients with repeated pulmonary infections, hemoptysis, and severe heart failure symptoms [25]. Right lung pneumonectomy in SS patients can be well tolerated in children with right lung hypoplasia and adequate remaining lung function [26]. In the multicentric study of European Congenital Heart Surgeons Association (ECHSA) [19], Patients who underwent right pneumonectomy had more severe degree of preoperative congestive heart failure and respiratory symptoms, in addition to a severe degree of pulmonary arterial hypertension at the catheterization study. The incidence of mortality and morbidity is higher in this group of patients [17,19,20,26].

In our case the patient underwent right pneumonectomy with ligature of the abnormal aorto-pulmonary arteries as a primary surgical time. This choice of surgical procedure was justified by the clinical status of the patient presenting severe dyspnea with recurrent infection, and by chest CT findings, showing an hypoplastic right lung with a large abnormal collateral feeding artery supplying an important pulmonary sequestration responsible for pulmonary over circulation. Therefore, right pneumonectomy associated to collateral artery

ligature may allow to reduce the blood flow and the amount of shunting as well as to remove the non-functional pulmonary sequestration responsible for recurrent infection and severe heart failure symptoms.

In a case of SS associated to TOF reported by Çınar., et al [4], the patient was treated by a hybrid combination of initial transcatheter occlusion of the arterial supply to the sequestration by a vascular plug, and corrective surgery for both TOF and disoriented pulmonary vein shortly after. Transcatheter occlusion of the anomalous systemic arterial supply has been reported as an alternative and less invasive method to surgical ligation in symptomatic patients with SS [19]. A Hybrid approach combining interventional catheterization that eliminates the arterial collateral flow, and surgery that corrects intracardiac abnormality as well as venous drainage, contributed to good short and long term results in this case management [4]. A similar therapeutic approach was adopted by Ismail., et al [5] in published case report of an 18 month-old infant with good results.

In another case reported by Azhari., *et al* [27], the patient underwent transcatheter occlusion of the collateral artery supplying the sequestered lobe, as well as pulmonary valve transcatheter balloon dilation in the catheterization laboratory, followed by a right-modified Blalock-Taussig shunt. Total repair of TOF, rerouting of the anomalous pulmonary vein to the LA and Blalock-Taussig shunt removal were performed few months after with good outcomes. An initial palliation strategy by surgical construction of a systemic-to-pulmonary shunt before total repair has been carried out in this patient in order to preserve the right lung function of the right lung and induce growth of its hypoplastic pulmonary artery.

The association of SS with other cardiac abnormalities have a major impact on long term results [19]. Only few cases of SS associated to TOF have been reported in the literature, and not all of them discussed therapeutic strategies and outcomes, which make the optimal surgical approach and related prognosis in this unusual variant of SS questionable. Moreover, no cases of SS with TOF treated by pneumonectomy as first surgery have been previously published. However, the association of SS to other cardiac malformations and to a sever clinical presentation suggest additional mortality when pneumonectomy is performed in comparison with patients with isolated SS. In our case, the patient's clinical condition deteriorated during the early postoperative period, with acute pulmonary and hemodynamic distress and death occurred 3 days after the surgery.

Conclusion

Scimitar syndrome is rarely associated to TOF, management of this association can be challenging. According to the literature, a hybrid approach including catheter intervention followed by surgical repair seems to be associated with good results, especially when symptoms are mild and when there is no recurrent infection. However, in some cases, the severity of symptoms and recurrent infection make pneumonectomy necessary even though it is associated to higher mortality.

Conflict of Interest

I Declare no conflict of interest.

Authors' Contributions

Contributions

HM conceived the study, participated in its design, acquired the data, performed a literature review, and drafted the manuscript. AD confirmed the diagnosis, performed patient's follow-up, and helped in literature review, and to draft and edit the manuscript. Both authors read and approved the final manuscript.

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