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Scimitar syndrome with different features in pediatric patients: a single-center experience

Çocuk hastalarda farklı özellikleriyle Scimitar sendromu: tek merkez deneyimi

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Abstract

Purpose: Scimitar syndrome is a rare congenital cardiac anomaly in which pulmonary veins are drained to the inferior vena cava (IVC) instead of the left atrium and it is often associated with additional cardiac and pulmonary anomalies. In this article, the different clinical features of pediatric patients with Scimitar syndrome are reported.

Material and methods: All patients with scimitar syndrome were diagnosed in our hospital between 2000 and 2020. The clinical findings, angiographic and surgical procedures, and follow-up of the patients are evaluated.

Results: Five pediatric patients aged between 1.5 months and 10 years were diagnosed as Scimitar syndrome. All patients were symptomatic. One patient had cor triatriatum and pulmonary venous stenosis, one patient had left atrial isomerism and absence of the IVC (azygous continuity), and another patient had coarctation of the aorta. One patient had dual drainage: the IVC and left atrium with meandering pulmonary veins, which we determined as a Scimitar variant. One infant patient with pulmonary hypertension and two patients with pulmonary to systemic flow ratio (Qp/Qs)>1.5 were treated surgically with the reanastomosis technique. Vascular embolization of the aortopulmonary collaterals was performed in two patients using vascular plugs, coils, and onyx.

Conclusion: Treatment should be planned individually in this syndrome due to Scimitar syndrome has a wide range of anatomic and clinical variations.

Key words: Aortopulmonary collateral, children, meandering pulmonary veins, Scimitar syndrome.

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Öz

Amaç: Scimitar sendromu, pulmoner venlerin sol atriyum yerine inferior vena kavaya (IVC) açıldığı ve sıklıkla ek kardiyak ve pulmoner anomalilerle ilişkili olduğu nadir görülen bir doğuştan kalp anomalisidir. Bu yazıda farklı klinik özellikleri olan Scimitar sendromlu pediyatrik hastalar tartışılmıştır.

Gereç ve yöntem: Hastanemizde 2000-2020 yılları arasında Scimitar sendromlu tanısı alan olgular çalışmaya alındı. Hastaların klinik bulguları, anjiyografik ve cerrahi işlemler ve takipleri değerlendirildi.

Bulgular: Çalışma süresi içinde yaşları 1,5 ay ile 10 yaş arasında Scimitar sendromu tanısı olan beş hasta değerlendirildi. Tüm hastalar semptomatikti. Bir hastada kor triatriatum ve pulmoner venöz stenoz, bir hastada sol atriyal izomerizm ve IVC yokluğu (azigos ven devamlılığı) ve diğer hastada aort koarktasyonu vardı. Diğer bir hastada ise çift drenaj saptandı. Bu hastada İVC'nin meandering pulmoner venler aracılığıyla sol atriyum ile olan bağlantısı Scimitar varyantı olarak değerlendirildi. Pulmoner hipertansiyonlu infantil bir hasta ve pulmoner / sistemik akım oranı (Qp/Qs)>1,5 olan iki hasta reanastomoz tekniği ile cerrahi olarak tedavi edildi. İki hastada ise aortopulmoner kollateraller saptanarak bu olgulara vasküler embolizasyon (vasküler plug, koil ve onyx kullanılarak) yapıldı.

Sonuç: Scimitar sendromunun çok çeşitli anatomik ve klinik varyasyonları olması nedeniyle tedavi ve takip kişiye özel olarak planlanmalıdır.

Anahtar kelimeler: Aortopulmoner kollateral, çocuk, meandering pulmoner venler, Scimitar sendromu.

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Introduction

Scimitar syndrome is a rare congenital heart anomaly in which the pulmonary veins drain into the inferior vena cava (IVC) instead of the left atrium and is often associated with additional anomalies such as aortopulmonary collaterals (APCs), right lung hypoplasia, right pulmonary artery hypoplasia, atrial septal defect (ASD), pulmonary sequestration, and dextrocardia [1, 2]. Treatment and follow-up of the Scimitar syndrome should be planned individually according to patient because of its wide range of presenting age, anatomic, and clinical variations [3]. In this article, the clinical findings, angiographic and surgical procedures, and follow-up of pediatric patients with Scimitar syndrome with different presentations are reported.

Material and methods

All cases with Scimitar syndrome were diagnosed in our hospital between 2000 and 2020. One of the patients was reported previously from our clinic [4]. The research protocol was approved by the local ethics committee of Baskent University(Project No: KA20/344). Informed consent was obtained from all patients. The study was conducted in accordance with the principles of the Declaration of Helsinki. The clinical findings, angiographic and surgical procedures, and follow-up of the patients with Scimitar syndrome are evaluated.

Results

Five pediatric patients (2 female,3 male) with Scimitar syndrome diagnosis, were evaluated. Diagnosis age of patients was between 20 days and 9 years. All patients were symptomatic. Their symptoms were sweating, fatigue during feeding, cyanosis, cough and restlessness. One infant patient had a history of recurrent respiratory infections. The clinical features, angiographic and surgical procedures of the patients were given in Table 1.

Table 1. The demographic, clinical, and laboratory features of the patients with Scimitar syndrome

Case	1	2	3	4	5
Age	17 years	66 days	6 years	6.5 years	10 years
Age of diagnosis	1.5 month	20 days	4.5 years	5 years	9 years
Gender	Female	Male	Male	Female	Male
Complaint	Sweating, fatigue	Cyanosis	Sweating, weakness	Cough	Fatigue, fainting
Concomitant congenital heart disease	Collateral artery Pulmonary sequestration Right pulmonary artery hypoplasia Coarctation of the aorta	Cor triatriatum Hypoplastic pulmonary veins Pulmonary hypertension ASD	Collateral artery Pulmonary sequestration ASD	Meandering light pulmonary veins	Left atrial isomerism Total abnormal hepatic venous connection
mPAP(mmHg)*	19	32	31	22	18
Qp/Qs	1.59		4.6	1	2.21
Interventional	Collateral occlusion(coil, onyx) CoA balloon angioplasty	No collateral	Collateral occlusion (vascular plug)	No collateral	No collateral
Surgery	No	Pulmonary vein repair Cor triatriatum	Pulmonary vein repair ASD	No (Qp/Qs:1)	Pulmonary vein repair
Surgery time	No	42 days	5 years	No	9.2 years
Follow up time (years)	17	Exitus at 2.5 months old (Hypoplastic pulmonary veins)	1.5	1.5	1

* mPAP: Mean pulmonary arterial pressure

A pulmonary venous return anomaly was suspected in echocardiography in two patients. In the echocardiography findings of the other three patients, concomitant congenital anomalies were detected. Concomitant congenital heart disease(CHD) is described in Table 1. Coarctation of the aorta in the first patient; cor triatriatum, hypoplastic pulmonary veins, pulmonary hypertension and ASD in the second patient; left atrial isomerism, absence of the infrahepatic segment of the IVC (azygos vein connected to the right superior vena cava), total abnormal hepatic venous connection, and foramen ovale were diagnosed in the fifth patient.

Angiography was performed in four patients. Pulmonary-systemic flow ratio (Qp/Qs) was calculated range 1-4.6. In addition to the scimitar vein, collateral artery was seen in two patients. Collateral arteries were closed interventionally in two patients. Angiography of the first patient showed a large and three feeding artery branches arising from the aorta and another feeding artery arising from the celiac trunk. (Figure 1a) We performed occlusion of arteries and branches with microcoils (Figure 1b) Post-closure control angiography revealed incomplete occlusion. An embolic agent for occlusion of the all feeding arteries was used. Ultraflow microcatheter (Covidien, USA) was used during Onyx embolization, dimethyl sulfoxide (DMSO) was injected in the ultraflow microcatheter to fill the feeding arteries and then Onyx (ev3, USA) was injected slowly under fluoroscopy (Figure 1c). In the third patient, two collateral arteries with diameters of 5.3 mm and 3.2 mm originating from the descending aorta were supplying blood to the lower zone of the right hypoplastic lung segment. Both collaterals were closed respectively by inserting 8-mm and 6-mm vascular plugs (Lifetech Cera).

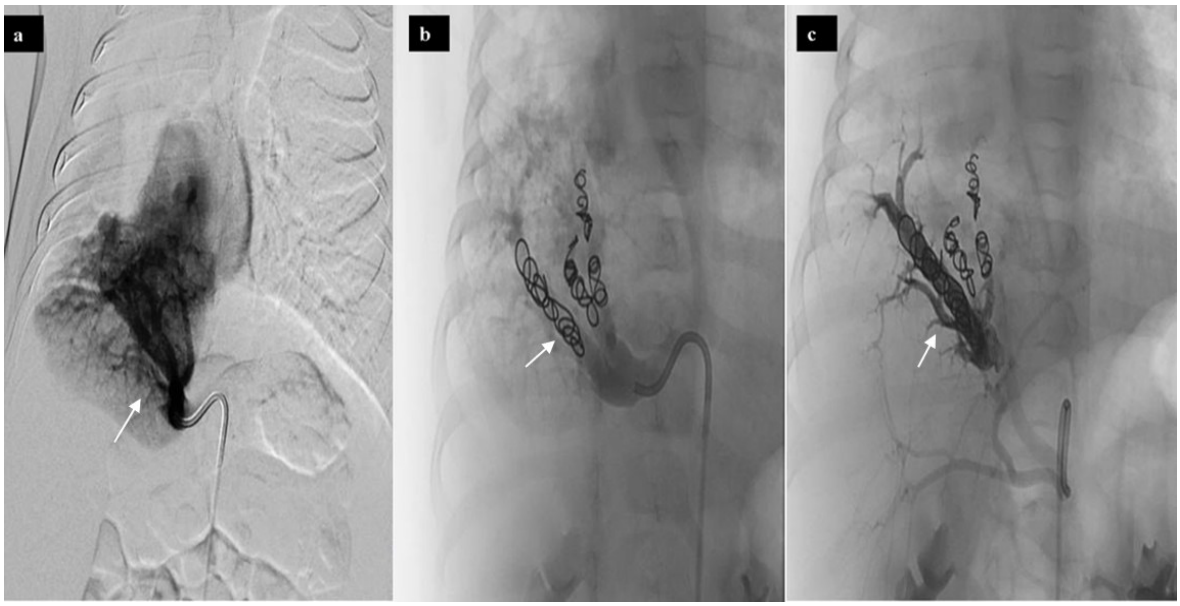

Figure 1. The angiogram images of case 1. The images show; a) the arterial collaterals were taking origin from the aorta, b) the three feeding artery branches after occlusion with micro-coils, c) a large artery embolization with onyx

Meandering right pulmonary veins were detected in the fourth patient. The right pulmonary veins connected with the IVC via the right vertical descending vein. (Figure 2a, 2b) The right lower lung segmental vein drained to the IVC via the right descending vertical vein. The right upper and middle pulmonary veins connected to the left atrium and the vertical vein via meandering pulmonary veins (Figure 3a, 3b). In the fifth patient the angiogram showed that the left pulmonary veins drained to the left atrium, and the right pulmonary veins drained into the hepatic vein via the right lateral vertical vein. The scimitar sign was present.

Surgery was performed in three patients. The surgery age was 42 days-9.2 years. The decision of surgery was made in one infant patient (case 2) due to pulmonary hypoplastic veins and in the other two patients (case 3 and 5) due to pulmonary to systemic flow ratio (Qp/Qs)>1.5. In the second patient during surgery, two separate right pulmonary veins

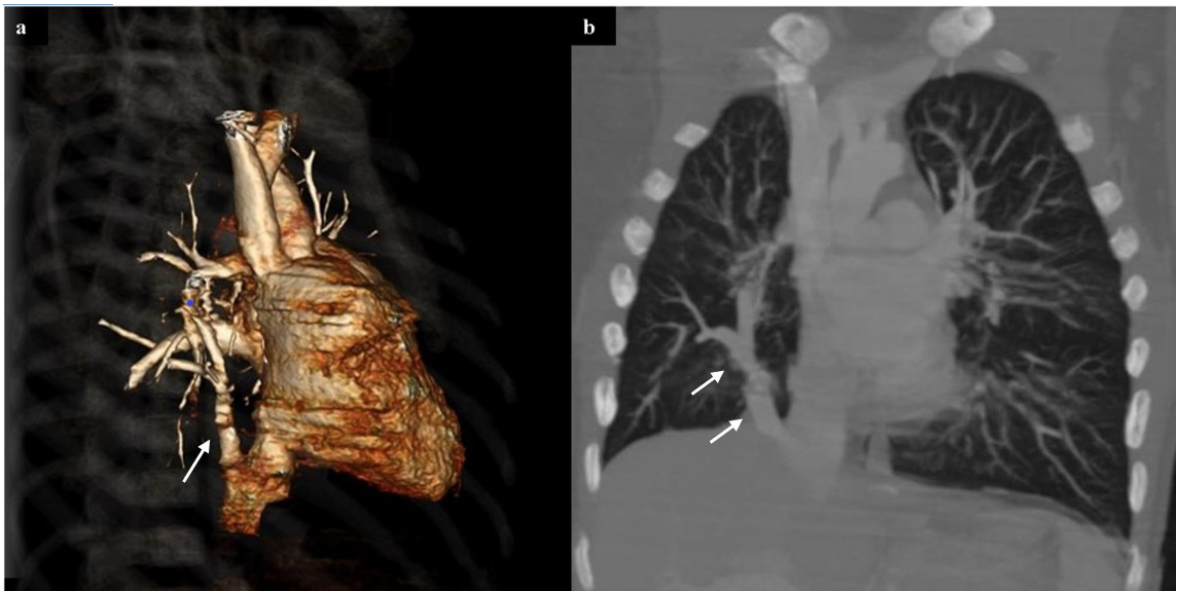

Figure 2. The images of case 4. The images show, a) Scimitar vein 3D Image, b) Scimitar vein coronal plane computed tomography image

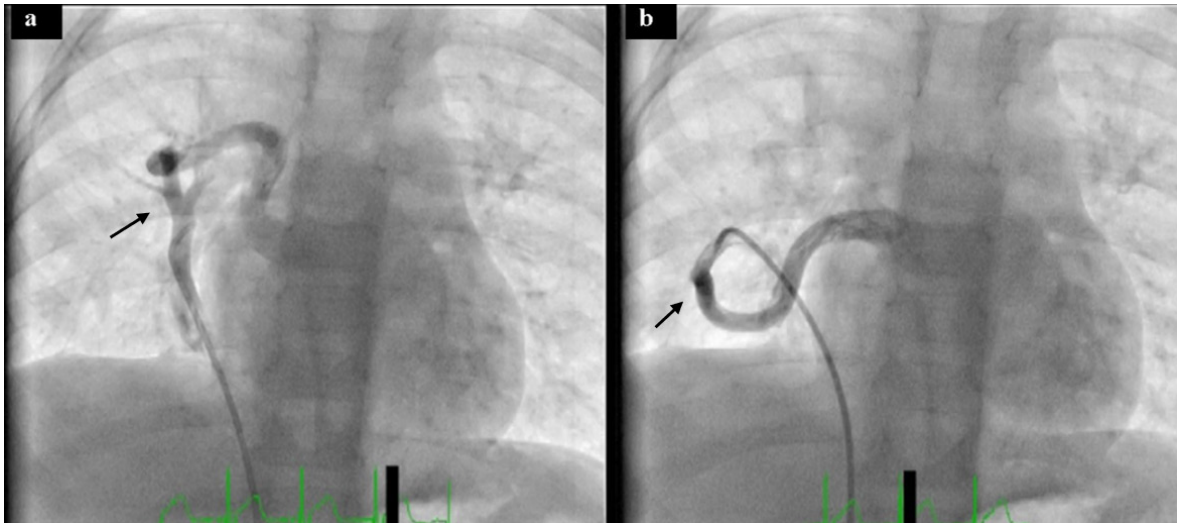

Figure 3. The angiogram images of case 4. The images show a) dual drainage angiography image between the Scimitar vein and the left atrium, b) an angiography image of meandering veins

of 4 mm and 2.5 mm were observed under the diaphragm, which drained to the IVC instead of opening to the left atrium. The left pulmonary veins were connected to the left atrium with a 4-mm diameter tunnel. In addition, sequestered lung segment and large secundum ASD were observed. The left atrium cavity was small. The large right pulmonary vein was anastomosed with the left atrium. The small right pulmonary vein was not anastomosed. The general underdevelopment of the pulmonary veins was not considered appropriate for correction. The ASD was left open. In the other two patients, repair of the anomalous pulmonary venous return was performed by surgery.

The patient with hypoplastic pulmonary vein stenosis died after a postoperative 24th day. The follow-up period of the other 4 patients ranged from 1 year to 17 years with good clinical conditions.

Discussion

Scimitar syndrome is a very rare congenital heart anomaly comprising a partial anomalous pulmonary venous connection to the IVC, lung hypoplasia, hypoplasia in the right pulmonary artery and APCs, and pulmonary sequestration [5, 6]. Scimitar syndrome has a wide range of anatomic and clinic variations and has high morbidity and mortality rates.

In early childhood, patients with Scimitar syndrome are mostly symptomatic with heart failure and recurrent lung infections, but are usually diagnosed incidentally in adult age [6-8]. Scimitar syndrome patients are divided into two groups as infantile under 1 year old and older group over 1 year old [7, 9, 10]. The presence of additional cardiac anomalies and pulmonary hypertension causes patients to be symptomatic earlier and affect prognosis [1, 7]. For these reasons, the prognosis in infants with Scimitar syndrome has been reported to be worse than in the older group [6]. The diagnosis ages of our patients, two of which were infants, ranged between 20 days and 9 years, and all were symptomatic. Different clinical symptoms that reflect heart failure were seen in our patients, and one patient had a history of recurrent lung infections. A two-month old patient died on the 24th day of the post operation due to pulmonary hypertension and severe left pulmonary vein stenosis. There was no reanastomosis stenosis in this patient. Severe left pulmonary vein stenosis was considered as the primary cause of death. In publications, pulmonary vein stenosis is accepted as an independent risk factor for poor prognosis in Scimitar syndrome [2, 9].

Scimitar syndrome can be seen with many congenital heart diseases and the most common association is secundum ASD. Consistent with the literature, secundum ASD was the most common CHD in our patients [3]. In addition, one patient had cor triatriatum and pulmonary venous stenosis, one patient had left atrial isomerism and absence of IVC (azygous continuity), and another patient had coarctation of the aorta.

The term Scimitar variant is used for patients who do not show all the features of Scimitar syndrome and sometimes show additional features. Meandering pulmonary veins with a dual-drain to IVC and left atrium are described as the 'Scimitar variant' [11]. In the literature, Goodman et al. [11] described the presence of the Scimitar sign and an anomalous right pulmonary vein draining to the left atrium [12]. Only the presence of a meandering pulmonary vein does not cause a left to right shunt, whereas a bilateral connection leads to a left to right shunt, which requires treatment [13]. We considered our fourth patient as having a Scimitar variant because of the presence of the dual-drainage to IVC and left atrium with meandering pulmonary veins. However, she was mildly symptomatic and her Qp/Qs was 1 at catheterization, thus an intervention was not considered. There were no problems in the follow-up.

Treatment planning in Scimitar syndrome is determined by the presence of the accompanying anomalies and the patient's symptoms. Patients who are asymptomatic or have mild symptoms do not require surgery. The presence of pulmonary hypertension and additional anomalies may require early surgery and these additional anomalies increase the risk of morbidity and mortality. Patients with pulmonary hypertension who have a Qp/Qs>1.5 in catheterization should undergo surgery [1, 6, 7, 9]. Two surgical techniques, either baffle or reanastomosis, are performed according to the choice of the surgeon [6, 14]. One infant patient with pulmonary hypertension and two patients with Qp/Qs>1.5 were treated surgically with the reanastomosis technique. In addition, the ASD was closed. No postoperative anastomotic stenosis was observed. However, in an infant with pulmonary hypertension, no intervention could be made to his narrow left pulmonary veins. In our first patient with a Qp/Qs of 1.59, firstly, the APCs were closed, after which the Qp/Qs were calculated as 1.5, and the decision was follow-up without intervention.

Two of our patients had pulmonary sequestration, which is a feature of Scimitar syndrome [15]. The treatment of pulmonary sequestration was previously surgical removal of abnormal tissue. Transcatheter occlusion of abnormal arteries is less invasive and has fewer complications than surgery [14, 16]. Coil and vascular plugs are effectively used for transcatheter occlusion of APCs in pediatric patients with CHD. Transcatheter occlusion of APCs was performed in two of our patients. Embolization of the feeding arteries, which originated from the celiac trunk and ascending aorta and their branches, was performed using microcoils in the first patient. Residual flow was managed with onyx. The embolization was performed successfully, and APCs were completely occluded. The third patient had two collateral arteries and two collaterals were closed by a vascular plug.

The limitations of our study are the low number of patients and retrospective evaluation of the patients from a single center. Because Scimitar syndrome is a rare congenital

abnormality, multicentric prospective studies with more number of patients would increase the information about the patients with Scimitar syndrome.

In conclusion, Scimitar syndrome carries an increased risk of morbidity and mortality in the presence of pulmonary hypertension and additional anomalies, are more common in infants than in adults and they may require early surgery. Treatment planning in Scimitar syndrome is determined by the presence of the accompanying anomalies and the patient's symptoms.

Conflict of interest: No conflict of interest was declared by the authors.

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Ethics committee approval: This study was approved by Başkent University Institutional Review Board (project no: KA20/344).

Author contributions

A.O. designed the study, performed the data collection, wrote the draft, and created the tables and figures for the manuscript. B.V., İ.E. and H.A.G. designed the study, designed data collection tools, performed data collection and analysis, and wrote the initial article. M.Ö. performed data collection, analyzed data, created tables and figures N.K.T. and S.A. conceptualized and designed the study, revised the article. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.