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The complex form of anomalous pulmonary venous return; new generation imaging and surgical approach of scimitar syndrome

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Summary

Scimitar syndrome is a rare congenital heart disease where anamolous pulmonary veins drain into inferior vena cava. Its pathogenesis is unknown and it is caused by abnormal development of the pulmonary bud during early embryogenesis. In the chest radiography, the shadow of anomalous veins resembles a Turkish sword (better known as scimitar) located between the lower right lung and the heart. It is associated with other anomalies such as right pulmonary hypoplasia, cardiac dextroposition, anomalous systemic arterial supply to the right lung, pulmonary sequestration and atrial septal defect. In this article, we present an 8-year-old boy operated for scimitar syndrome with partial anomalous pulmonary venous return draining into the inferior vena cava, atrial septal defect, dextropositon of the heart and hypoplastic right lung. The diagnostic significance of new generation imaging methods and surgical strategy have been scrutinized. *(Turk Arch Ped 2013; 48: 173-5)*

Key words: Computerized tomography, congenital heart disease, "scimitar" syndrome

Introduction

Scimitar syndrome is a very rare pathology which occurs with a rate of 2/100 000 and which presents in the infancy, childhood or adulthood with a female/male ratio of 2/1. In addition to partial abnormal pulmonary venous return into the vena cava inferior, cardiac dextroposition and right lung hypoplasia, the other significant abnormalities accompanying the syndrome include receiving of arterial blood directly from the aorta by the right lung with a rate of 60% and atrial septal defect (ASD) with a rate of 40% (1). In "scimitar" syndrome, anomalously returning pulmonary veins, the place where the pulmonary veins are drained and the relation of the pulmonary veins with ASD are significant in terms of surgery. Generally, a part or all of the pulmonary veins of the lower lobe of the right lung and sometimes middle lobe veins drain into the vena cava inferior. However, cases in which the whole right lung and rarely left pulmonary veins are drained anomalously have also been reported (2).

A careful anatomic examination is required in "scimitar" syndrome. In this article, successful surgical intervention of a patient with "scimitar" syndrome and the importance of new generation imaging methods in the diagnosis have been emphasized.

Case

A 8-year-old male patient presented to our clinic with dyspnea and easy fatigability in December 2009. His physical examination and electrocardiogram were found to be normal. On chest X-ray, the heart had a normal size, but the heart and mediastinal structures were observed to be shifted to the right side because of right lung hypoplasia. On echocardiography, partial anomalous pulmonary venous return draining into the vena cava inferior and ASD (secundum type) were found. Cardiac catheterization revealed dextroposition and right lung hypoplasia. Following right pulmonary artery injection the vena cava inferior and right atrium were visualized in the venous return phase.

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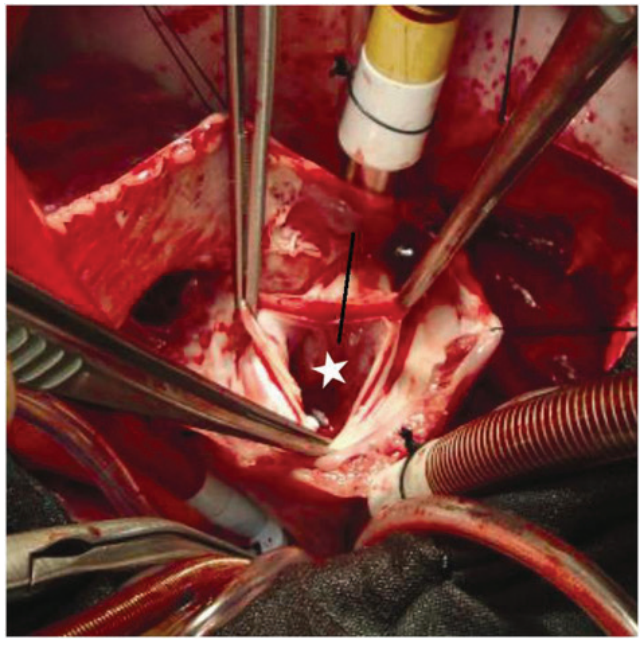
The Qp/Qs ratio was found to be 1,91. Pulmonary artery and right ventricular pressures were found to be normal (27/5 mmHg and 27/0 mmHg, respectively). On 64-section-multidetector computarized tomography (MDCT), three main veins were observed to be combined at the level of the lower lobe of the right lung and to be drained into the vena cava inferior by way of a truncal vein at the level of the diaphragm. The patient was operated under cardiopulmonary bypass. During right atriotomy, it was observed that there was a fossa ovalis type 1x2 cm ASD and the right pulmonary veins were combined and drained into the vena cava inferior by way of a truncal vein. Atrial septal defect was expanded towards the vena cava inferior (Picture 1) and was closed using pericard patch such as the pulmonary veins were kept in the left atrium. The patient who had no problem in the postoperative period was discharged on the 6th day after operation.

Discussion

The first two articles published by Cooper and Chassinat (3,4) in 1836 on the subject of "scimitar" syndrome pioneered many studies written on this subject. The drainage of the weird vein in the shape of "scimitar" into the vena cava inferior was described on chest x-ray by Halasz et al. (5) in 1956. However, the name of "scimitar" syndrome was used by Catherine A. Neill et al (6) for the first time in 1960.

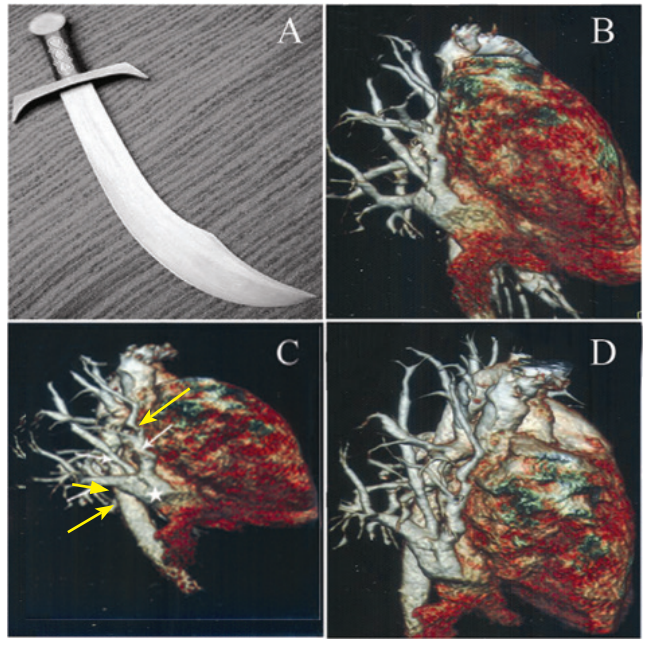
Historically, the sign of "scimitar" (Turkish sword, Persian sword or shamshir) which is one of the 100 classical radiological signs (Picture 2A) is an important chest X-ray finding (7).



Picture 1. Atrial septal defect (ASD) (star) and the cut line (between ASD and vena cava inferior); view from the right atrium

In patients with anomalous pulmonary venous return, it is substantially difficult to evaluate pulmonary vein anatomy fully by transthoracic echocardiography (TTE). For the pulmonary veins the width, course and the area where they are drained are important. In patients with congenital cardiac anomaly, the rate of a definite diagnosis on TTE is 85-90%, but arcus aorta anomalies and pulmonary veins may sometimes be overlooked. In some conditions, sufficient acoustic window may not be found for examination (8). Complete anatomical evaluation is important before surgical or transcatheter ASD closure procedure (9).

Development of advanced imaging methods in recent years have led to beneficial results in this subject. Interestingly, the Turkish sword sign obtained especially in the frontal plane with 64-section MDCT is similar to the appearance on chest X-ray (Picture 2B). The vena cava inferior into which all pulmonary veins and the truncal vein where they are combined are drained is clearly visualized on these advanced imaging methods and provide a great convenience during operation for the surgeon (Picture 2C and D). On the other hand, MDCT is substantially beneficial also in conditions where the arterial circulation of the right



Picture 2. A: Turkish sword, B: Finding of Turkish sword in the venous phase of MDCT cardiac anterior projection, C: The truncal vein (star) where all right pulmonary veins (arrows) are combined and drainage into the vena cava inferior, D: The truncal vein where the right pulmonary veins are combined and MDCT appearance in the posterior projection

lung is provided by the collaterals of the descending aorta, because ligation of the collateral web is important surgically. No such collateral vessels were observed in our patient.

In patients who are diagnosed with "scimitar" syndrome during the infancy, the sign of "Turkish sword" is generally absent. In patients who are diagnosed during the childhood and adulthood, complaints including easy fatigability and dyspnea may be present and the sign of "Turkish sword" is found on chest X-ray with a rate of 70%. In this group of patients, right lung hypoplasia is generally absent and the sign of "Turkish sword" can be determined more easily. If ASD is present (25-50% of older children and adults), specific findings are noted. Although our patient had ASD, he lived until the age of 8 years without being diagnosed. He only had easy fatigability and dyspnea. Chest X-ray revealed right lung hypoplasia, but the sign of "Turkish blade" was absent. The important indications for surgery in "scimitar" syndrome include ASD accompanying the "scimitar" vein, pulmonary hypertension and stenosis of the anomalous vein. Patients who are asymptomatic in infancy may be followed up clinically until they reach the sufficient body weight. Surgical intervention is necessary, if pulmonary hypertension develops. Some recomend early surgical intervention, if "cardiopulmonary bypass" and "cardiac arrest" are not going to be applied (10).

The first physiological correction was performed by Kirklin et al. (11) in 1956. In this procedure, the right main pulmonary vein was anastomosed with the right atrial wall adjacent to ASD and the atrial wall was stitched to the border of ASD.

Curently, the "scimitar" vein is seperated from the vena cava inferior and bound to the left atrium by right thoracotomy without "cardiopulmonary bypass" in patients without ASD. In patients with atrial septal defect, following closure of ASD with pericard patch under "cardiopulmonary bypass" the anomalous pulmonary vein seperated from the inferior vena cava is anastomosed with the left atrium using a "polytetrafluoroethylene" (Gore-Tex) graft (10, 12). In our patient, the truncal vein was draining into the vena cava inferior at the level of the diaphragm at a point close to the heart. The space between the mouth of the truncal vein and ASD was closed. Atrial septal defect was closed using a patch processed with glutheraldehide such as the truncal vein was kept on the side of the left atrium. Here, it was taken care of that the enterance of the vena cava inferior was not narrowed. Conclusively, "scimitar" syndrome is a rare but severe congenital heart disease which can be corrected successfully independent of the method used. Detailed evaluation by advanced imaging methods before surgery will allow a more convenient and trouble-free intervention and increase the success of treatment.

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