A rare combination of the Scimitar Syndrome and aneurysmal dilatation of the pulmonary artery in an adult

Tiange Li[1], Yunfei Ling[1], Yue Zhuo[1], and Yongjun Qian[1]

[1]Sichuan University West China Hospital

May 30, 2022

Abstract

In this case, we reported an old female with a rare combination of the Scimitar Syndrome and pulmonary artery aneurysm, and she presented with chest pain, fatigue and shortness when admitted in our hospital. Further cardiac examination including echocardiography and computed tomography angiogram were performed and revealed these rare anomalies. Surgical procedure was carried out to cure the patient. This patient stayed in ICU for few days without any major complications and discharged from hospital after 10 days.

A rare combination of the Scimitar Syndrome and aneurysmal dilatation of the pulmonary artery in an adult

Tiange Li[1*], Yunfei Ling[1*], Yue Zhuo[2], Yongjun Qian[1#]

[1] Department of cardiovascular surgery, West China Hospital, Sichuan University, Chengdu 610041, People's Republic of China

[2] West China School of Medicine, West China Hospital, Sichuan University, Chengdu, 610041, People's Republic of China

[*] These two authors contributed equally to this work and should be considered co-first authors.

[#] Corresponding author: Yongjun Qian, MD, PhD

Phone: + 86 28 85422897

Fax: + 86 28 85422897

Email:*qianyongjun@scu.edu.cn*

A rare combination ofthe Scimitar Syndrome and aneurysmal dilatation of the pulmonary artery in an adult

Tiange Li[1*], Yunfei Ling[1*], Yue Zhuo[2], Yongjun Qian[1#]

[1] Department of cardiovascular surgery, West China Hospital, Sichuan University, Chengdu 610041, People's Republic of China

[2] West China School of Medicine, West China Hospital, Sichuan University, Chengdu, 610041, People's Republic of China

[*] These two authors contributed equally to this work and should be considered co-first authors.

[#] Corresponding author: Yongjun Qian, MD, PhD

Phone: + 86 28 85422897

Fax: + 86 28 85422897

Email:*qianyongjun@scu.edu.cn*

Abstract

In this case, we reported an old female with a rare combination of the Scimitar Syndrome and pulmonary artery aneurysm, and she presented with chest pain, fatigue and shortness when admitted in our hospital. Further cardiac examination including echocardiography and computed tomography angiogram were performed and revealed these rare anomalies. Surgical procedure was carried out to cure the patient. This patient stayed in ICU for few days without any major complications and discharged from hospital after 10 days.

Keywords : Congennital heart disease

A 62-year-old female was referred to department of cardiovascular surgery in our hospital presented with spontaneous chest pain, fatigue and shortness of breath for 2 months. She had been found of dextroposition of the heart and heart murmur for over 30 years, since remained asymptomatic all along except catching cold easily, treatment was not carried out before. Physical examination revealed a little displacement of apex beat site to the right, and grade III systolic murmur located at the left margin of the sternum, second intercostal space.

Among further cardiac examination, transthoracic echocardiography (TTE) revealed severe dilation of pulmonary artery accompanied with pulmonary valve stenosis, and the inferior venous cava (IVC) was significantly widened. Computed tomographic images demonstrated hypoplasia of right pulmonary artery and lung. Computed tomography angiogram (CTA) indicated significant dilatation of pulmonary artery trunk and the left pulmonary artery with a maximum diameter of 6.0 cm (Figure 1, panel A), three-dimensional reconstruction visually demonstrated the aneurysmal dilatation of pulmonary artery (Figure 1, panel B). Two branches of left pulmonary vein merged into the left atrium, while right pulmonary veins were not found to enter into the left atrium (Figure 1, panel C). The CTA image demonstrated the right common pulmonary vein directly merged into the inferior vena cava above the diaphragm, while the diameter of the IVC was significantly enlarged (Figure 1, panel D).

Open heart surgery through middle sternotomy under cardiopulmonary bypass was performed for the correction of scimitar vein. Autologous pericardial patch was used to create an intra-atrium tunnel to baffle the scimitar vein into left atrium via the artificial atrial septal defect. In addition, the enlarged pulmonary artery was repaired close to the normal diameter. Intraoperative transesophageal echocardiography revealed the flow of inferior vena cava and scimitar vein were constant and scimitar vein merged into the left atrium smoothly through the atrial septal defect, which proved operation was successful. This patient stayed in ICU for few days without any major complications and discharged uneventfully after 10 days.

The scimitar syndrome is a congenital cardiovascular defect consisted following anomalies: partial or total anomalous right pulmonary venous drainage to the inferior vena vein with variable degrees of right pulmonary artery and lung hypoplasia; one or more anomalous abdominal aortopulmonary collateral to the right lung, usually right lower lobe; and variable degrees of dextroposition of the heart[1~3]. The syndrome is usually appeared with other defects, atrial septal defect (80%) is the most common comorbidity, following is patent ductus arteriosus (75%), ventricular septal defect (30%) and pulmonary vein stenosis (20%)[4]. We reported this rare case of the scimitar syndrome combined with aneurysmal dilatation of the pulmonary artery. Further speculation, long-term overload of right heart volume, existence of aortopulmonary collateral and mild pulmonary valve stenosis common caused the significant dilation of the pulmonary artery, and finally lead to symptoms of heart failure.

CONFLICT OF INTERESTS

All authors declare that there are no conflict of interests.

ETHICS STATEMENT

This manuscript and all of its content meet the ethical guidelines, including adherence to the legal requirements of the study country. The need for patient consent was waived.

FUNDING

This study was funded by 1·3·5 project for disciplines of excellence-Clinical Research Incubation Project, West China Hospital, Sichuan University (ZYJC21064).

References

1. Dusenbery, S. M. et al. Outcome predictors and implications for management of scimitar syndrome. American heart journal 165, 770-777, doi:10.1016/j.ahj.2013.01.016 (2013).
2. Midyat, L. et al. Eponym. Scimitar syndrome. European journal of pediatrics 169, 1171-1177, doi:10.1007/s00431-010-1152-4 (2010).
3. Bo I, Carvalho JS, Cheasty E, Rubens M, Rigby ML. Variants of the scimitar syndrome. Cardiol Young. 2016 Jun;26(5):941-7. doi: 10.1017/S1047951115001651. Epub 2015 Sep 16.
4. Diaz-Frias J, Widrich J. Scimitar Syndrome. 2022 May 8. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan–.

Figure

Figure 1. A and B. Computed tomography angiography and three-dimensional reconstruction demonstrated the aneurysmal dilatation of the pulmonary artery trunk and the left pulmonary artery. **C.**Computed tomography angiography revealed left pulmonary veins merged into the left atrium, while the right pulmonary vein was not found.**D.** Computed tomography angiography demonstrated the right common pulmonary vein entered into the significantly enlarged inferior vena cava. (LPA, left pulmonary vein; RPA, right pulmonary vein; LA, left atrium; IVC, inferior venous cava.)