Giant superior vena cava aneurysm after Fontan operation

Ibrahim Cansaran Tanidir1*, Aysel Turkvatan2, Taner Kasar1, and Alper Guzeltas1

1Department of Pediatric Cardiology, Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Center and Research Hospital, Istasyon Mah. Turgut Ozal Bulvan No: 11, 34303 Küçükçekmece Istanbul, Turkey and 2Department of Radiology, Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Center and Research Hospital, Istanbul, Turkey

*Corresponding author. Tel: +90 212 692 20 00; Fax: +90 212 471 94 94, Email: cansaran@yahoo.com

A 23-year-old female, as diagnosed with complex cyanotic congenital heart disease in infancy, admitted to the emergency department with acute chest pain and shortness of breath on exertion. At the age of 3 and 7 years, she underwent right bidirectional Glenn anastomosis and extracardiac Fontan operation, respectively, because of the diagnosis of double inlet left ventricle, right ventricular hypoplasia, and ventriculoarterial discordance. Since then she was lost in follow-up.

The initial chest X-ray showed an oval, well-defined opacity with smooth contours causing dilatation of the mediastinal shadow. A chest computed tomography scan demonstrated a contrast-enhancing fusiform sac, suggesting a giant superior vena cava (SVC) aneurysm measuring 5.7 × 8.1 cm with no significant stasis and no evidence of thrombus or filling defects in the pulmonary arteries (Panels A–E, and see Supplementary data online, Video S1). A catheterization study revealed a large SVC aneurysm measuring 5.7 cm (Panel F, and see Supplementary data online, Video S2). Mean superior-inferior caval and right-left pulmonary artery pressures were <6 mmHg and no clinically significant fistula was detected. Medical therapy was recommended.

Aneurysmal dilation of the SVC is a rare anomaly. To the best of our knowledge, there has been only two cases following Glenn anastomosis, and there have been no reports after Fontan operation. The exact pathogenesis of SVC aneurysm is not known and the possibility of the deficiency of longitudinal muscle layer of the adventitia has been reported. The common feature in all reported patients was the region of the aneurysm.

(Panel A) Chest X-ray, arrow shows a mediastinal shadow, (Panel B) axial multiple detector computed tomography (MDCT) image, (Panel C) oblique coronal thin maximum intensity projection MDCT image, (Panel D) anterior volume rendering MDCT image, (Panel E) posterior volume rendering MDCT image, and (Panel F) the angiogram demonstrating the fusiform aneurysm of superior vena cava. Ao, aorta; LIV, left innominate vein; LPA, left pulmonary artery; RIV, right innominate vein; RPA, right pulmonary artery; SVC, superior vena cava.

Supplementary data are available at European Heart Journal — Cardiovascular Imaging online.