Giant superior vena caval aneurysm in a post-Glenn patient

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Abstract

Aneurysms of mediastinal systemic veins are extremely rare, usually asymptomatic and incidentally diagnosed during chest radiography. We describe the case of a giant superior vena caval aneurysm in a 14-year old male following Glenn surgery and discuss its complications and management.

Keywords: Superior vena caval aneurysm

INTRODUCTION

Aneurysms of mediastinal systemic veins are rare, usually asymptomatic and incidentally diagnosed during chest radiography [1]. A literature review shows the reporting of only 35 cases of superior vena cava (SVC) aneurysm. We describe the case of a giant SVC aneurysm in a 14-year old male following Glenn surgery and discuss its complications and management.

CASE

A 14-year old male was diagnosed with complex cyanotic congenital heart disease in infancy. He was detected to have transposition of the great arteries with ventricular septal defect, persistent ductus arteriosus and pulmonary artery bifurcation stenosis with an increased pulmonary blood flow in infancy. At the age of 1 year, he underwent pulmonary artery banding with left modified Blalock-Taussig shunt. At the age of 3 years, he underwent bidirectional Glenn anastomosis along with atrial septectomy, repair of pulmonary artery bifurcation stenosis, division, suturing of persistent ductus arteriosus and the closure of a left Blalock-Taussig shunt. A year after the Glenn procedure, he underwent a cardiac catheterization study and was detected to have an SVC aneurysm measuring 3.8 cm. Four years after Glenn surgery, he suffered an embolic cerebrovascular accident (CVA) (left-side hemiparesis) from which he completely recovered. In the year 2008, he was admitted for a pre-Fontan catheterization study. Incidentally, his chest radiograph showed a large mediastinal shadow and transthoracic echocardiogram demonstrated no aortic aneurysm, but there was aneurysmal dilatation of the SVC. A catheterization study revealed acceptable haemodynamics for a Fontan procedure, but he had severe right ventricular (systemic ventricle) dysfunction and hence a medical follow-up was recommended. An SVC angiogram done during the catheterization procedure demonstrated a large SVC aneurysm measuring 6.0 cm (Fig. 1, Supplementary Video 1). He was started on aspirin, carvedilol and captopril and was followed-up in the cardiology clinic with no significant symptoms reported. As a follow-up of his SVC aneurysm, a chest computed tomography (CT) scan was performed. The scan demonstrated a contrast-enhancing sac adjacent to the ascending aorta suggesting a giant SVC saccular aneurysm measuring 6.6 × 5.1 cm with no significant stasis and no evidence of thrombus or filling defects in the pulmonary arteries (Fig. 2A and B, arrowheads).

Figure 1: Catheter angiography of SVC shows a giant size SVC saccular aneurysm with good flow into the pulmonary arteries (PA) in a patient after a bidirectional Glenn procedure.
Conservative therapy for the aneurysm was recommended in view of the high surgical risk.

DISCUSSION

Venous aneurysms are known to develop thrombosis with or without pulmonary embolism or venous obstruction, and may result in rupture [2, 3]. This is true for SVC aneurysms as well [2]. The majority of SVC aneurysms are fusiform with only a few saccular aneurysms reported [4]. SVC aneurysms are usually primary or congenital with histopathology revealing no defect, but there are reports of deficiency in the longitudinal muscle layer of the adventitia [4]. In addition, there is an association with cystic hygroma [4]. Management of a large SVC aneurysm is challenging. As a general consensus, conservative management is advised for fusiform SVC aneurysms [2]. However, with regard to saccular SVC aneurysms, in view of the risk of a sudden increase in size and thrombus formation, surgical therapy is recommended even in asymptomatic cases [2, 4]. In patients with a fusiform SVC aneurysm, long-term oral anticoagulation is recommended to prevent thrombosis and subsequent pulmonary embolism, but in patients with a saccular SVC aneurysm, a rupture during anticoagulation may be catastrophic. Hence, the benefit of anticoagulation should be weighed against the risk of rupture [2]. Therefore, in this patient with a saccular SVC aneurysm, aspirin therapy was recommended instead of anticoagulation.

CONCLUSION

In conclusion, we present a patient who developed an SVC aneurysm after a bidirectional Glenn procedure, which has not been reported previously. Physicians and surgeons caring for such patients should be aware of this rare complication of SVC in patients after the Glenn procedure and should manage appropriately, considering other comorbidity problems.

SUPPLEMENTARY MATERIAL

Supplementary material is available at ICVTS online.

Conflict of interest: none declared.

REFERENCES