Transcatheter management of superior vena cava obstruction following cardiac surgery: A case report from a resource-limited set-up

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ABSTRACT

Obstruction of the superior vena cava (SVC) is a rare complication after cardiac surgery in infants and children. We present the case of an 8-year-old boy who underwent bi-directional Glenn shunt followed by takedown of Glenn shunt and complete repair for cyanotic congenital heart disease. After 4 years of surgery, the child developed features of superior vena caval (SVC) syndrome. Echocardiography and CT angiography revealed complete obstruction of SVC without any forward flow. Transcatheter intervention was performed successfully to re-canalize and stent the SVC to maintain its patency. The patient was doing well at follow-up appointments, with good laminar flow through the stent. In conclusion, transcatheter management of post cardiac surgery SVC obstruction was successful in this patient.

Keywords: Postsurgical, superior vena cava stenting, superior vena cava syndrome

INTRODUCTION

Superior vena cava (SVC) syndrome is the constellation of clinical manifestations caused by obstruction of venous flow due to external compression, internal stenosis, or occlusion of the SVC. SVC syndrome affects ~15,000 patients in the United States annually.[1] No Indian data are available regarding the incidence of SVC syndrome among children following cardiac surgery. Malignancies such as primary lung cancer are the most common cause, accounting for 70% of cases in adults.[2] SVC syndrome is rare in children.[3,4] In children, surgery for congenital heart disease accounts for the majority of cases, followed by mediastinal tumors.[5] We are presenting a case of SVC syndrome resulting from cardiac surgeries for congenital heart disease and the successful endovascular therapy of it.

CASE REPORT

An 8-year-old boy, body weight 15 kg, presented with complaints of swelling of the face, neck, and upper torso, prominence of the neck, and superficial chest veins, along with headache.

The child was a known case of cyanotic congenital heart disease with a diagnosis of double-outlet right ventricle, large ventricular septal defect (VSD), with severe pulmonary stenosis, for which he had undergone a right-sided bidirectional Glenn shunt at the age of 2 years. After cardiac catheterization at 4 years of age, he underwent biventricular repair in the form of taking down of Glenn shunt, re-anastomosis of SVC to the right atrium (RA), VSD closure, and transannular patch augmentation of the right ventricular outflow tract.

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Four years after his complete corrective cardiac surgery, the patient presented with features suggestive of SVC syndrome, like a swollen face, conjunctival congestion, and prominent superficial veins on the upper torso.

Transthoracic echocardiography (TTE) revealed an intact VSD patch, no residual shunt, a well-opened right ventricular outflow tract with good flow across bilateral branch pulmonary arteries, and good bi-ventricular function. On the subcostal bi-caval view, the SVC flow could not be seen draining into the RA. A few venous collaterals were seen on the suprasternal view, which could not be well profiled on TTE.

To identify the etiology of SVC syndrome, we planned for a computerized tomography angiogram, which revealed an old, organized thrombus at SVC-RA anastomosis with complete obstruction and multiple decompressing veno-venous collaterals [Figure 1]. The dilated proximal segment of the SVC measured 16 mm. There was adequate space between the SVC and the right upper pulmonary vein [Figure 2].

After informed consent, we decided to recanalize the SVC-RA anastomosis in the catheterization laboratory. After proper heparinization, SVC angiography through an internal jugular vein (IJV) showed complete occlusion of SVC near the SVC-RA junction with increased SVC pressure (mean: 20 mm of Hg) with multiple venovenous collaterals (decompressing SVC through an azygous vein) [Figure 3]. Through the IJV route, we punctured the thrombus at the SVC-RA junction with the help of the Amplatzer right coronary catheter and 0.018 straight tip Terumo wire. After piercing the thrombus, the wire tip was placed in the inferior vena cava and snared through the right femoral venous route to make a venovenous loop. We predilated the obstructed area at the SVC-RA junction with a 2.5 mm

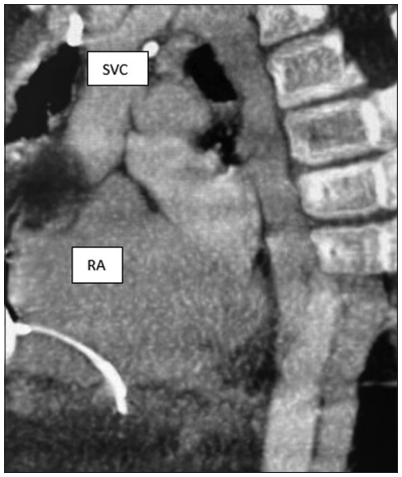

Figure 1: Old organized thrombus at superior vena cava - right atrium anastomosis with complete obstruction. SVC: Superior vena cava, RA: Right atrium

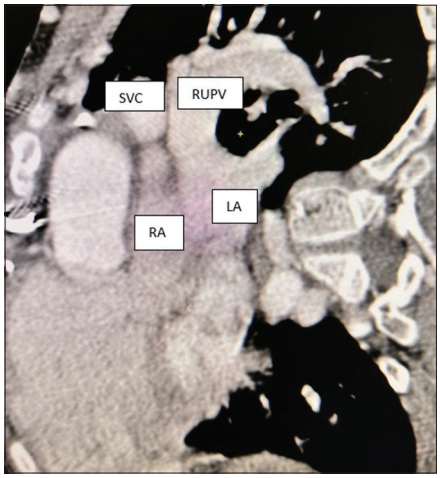

Figure 2: Spatial relationship between superior vena cava and right upper pulmonary vein. SVC: Superior vena cava, LA: Left atrium, RA: Right atrium, RUPV: Right upper pulmonary vein

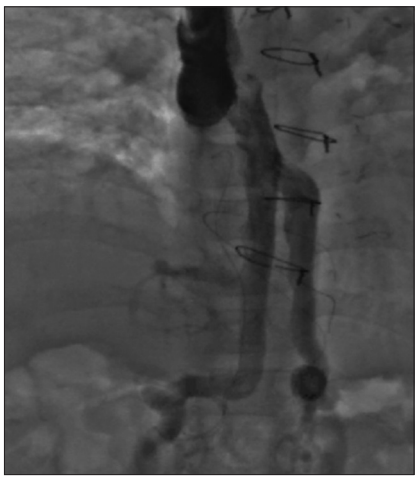

Figure 3: Superior vena cava (SVC) angiography through internal jugular vein showed complete occlusion of SVC near the SVC-right atrium junction with multiple venovenous collateral

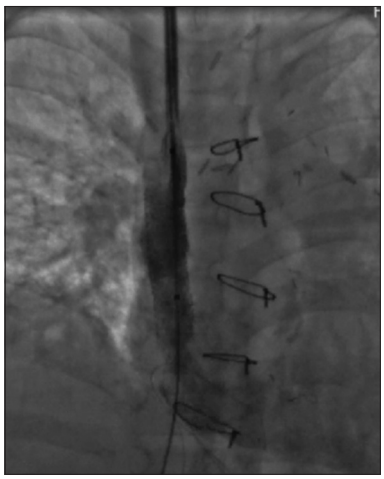

Figure 4: Postinflation angiography showed a nicely opened-up stenosed segment with good flow across the stent

coronary balloon and then took the Cook formula stent (10 mm × 40 mm) across the obstructed area. We placed the stent so that the stent was 60% at SVC and 40% at RA. After confirming the position, the stent was inflated optimally at 12 atm pressure. Postinflation angiography showed a nicely opened-up stenosed segment with good flow across the stent [Figure 4]. The hemodynamic assessment showed decreased SVC pressure to 10 mm of Hg (mean), whereas RA's mean pressure was 6 mmHg. Selective right upper branch pulmonary artery angiogram showed unobstructed pulmonary venous return.

The patient was managed with heparin infusion for the first 24 h, followed by oral dual antiplatelet therapy initiation. Regular echocardiography showed an unaltered stent position with good flow across the stent [Figure 5]. The patient was discharged on the second postprocedure day.

Two postprocedure follow-up visits were unremarkable, with clinical improvement (normalization of facial swelling and disappearance of upper torso venous prominence) and unaltered stent position on chest X-ray and echocardiography.

DISCUSSION

The treatment approach in patients with SVC syndrome is multidisciplinary, and treatment options include radiation therapy with or without chemotherapy, surgical bypass, or endovascular therapy, such as angioplasty, stenting, and catheter-based thrombus removal. In the contemporary era, benign SVC syndrome is usually related to pacemakers and defibrillator leads and less commonly postcardiac surgery.[6] Surgical bypass was once considered the primary treatment option for younger patients as it provides a durable solution. However, more recently, endovascular treatment has been regarded as the first-line therapy as it does not preclude open surgical bypass in the future, and it can be combined with other treatment modalities like hybrid revascularizations.[7]

We used transcatheter stent as the preferred primary treatment modality over open reconstruction surgery because of low postprocedural morbidity and fast recovery time.[8,9] Compared to balloon angioplasty, endovascular stenting relieves SVC obstruction more effectively with a lower recurrence rate.[10] A study by Tzifa *et al.* demonstrated that endovascular therapy is the first-line treatment that alleviates SVC obstruction and relieves accompanying symptoms while providing long-term benefits.[11]

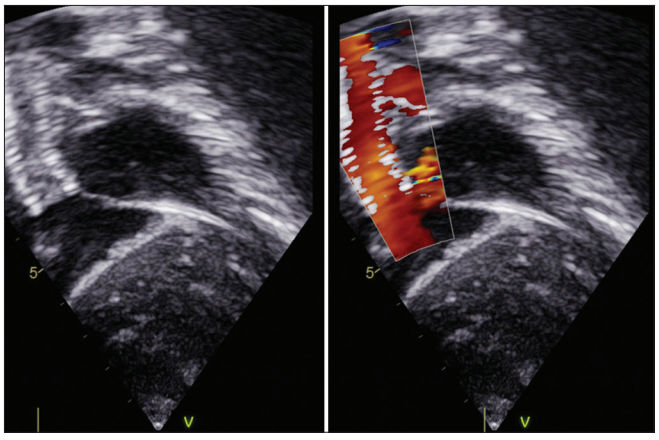

Figure 5: Echocardiography showing the stent in proper position with unobstructed flow through the stent

CONCLUSIONS

SVC syndrome can result from postcardiac surgical complications – mainly following cicatrization at the canulation site, especially where SVC is anastomosed with other structures. Endovascular therapy, like stenting, can be a treatment option for symptomatic relief and hemodynamic improvement, even in a limited-resource setup.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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