CASE REPORT

Percutaneous stenting of malignant superior vena cava syndrome in a patient with persistent left and absent right superior vena cava

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ABSTRACT

Stent placement is the preferred means of managing malignant obstruction of the superior vena cava (SVC). Persistent left and absent right SVC is a very rare venous anomaly. We here report the case of a 58-year-old man who underwent percutaneous stenting for malignant persistent left and absent right SVC obstruction caused by advancement of adenocarcinoma of the upper lobe of the left lung. The patient became symptom-free one day after endovascular stenting through the right femoral vein. However, he experienced repeated supraventricular tachycardia during the procedure. To our knowledge, this is the first report of stenting for malignant SVC obstruction with this congenital anomaly.

Keywords: superior vena cava syndrome; malignant obstruction; stent; anatomical variation.

INTRODUCTION

Superior vena cava syndrome (SVCS) is caused by obstruction of the flow of venous blood from the upper body into the right atrium. Approximately 73% to 97% of cases of SVCS are secondary to malignancy, and they are often caused by the advancement of lung cancer (1). Ever since the study reported by Charnsangavej et al. (2) in 1986, endovascular stenting has been well established as an effective means of treating SVCS. Stenting is considered the primary treatment option for patients with malignant SVCS (3). The anatomical variation of the superior vena cava (SVC) can make this interventional procedure challenging. We report our experience of percutaneous stenting of a rare malignant persistent left and absent right SVC obstruction cause by advanced left lung cancer.

CASE REPORT

Our institutional review board approved this study. Patient informed consent was waived for this retrospective study. A 58-year-old man was admitted to our hospital with a 1-week history of facial and arm swelling, dyspnea, and cough. Thirty months prior, he was diagnosed with advanced non-small-cell lung cancer (left upper lobe, T4 N3 M0, stage IIIIB adenocarcinoma) and repeatedly underwent chemotherapy and radiotherapy in our hospital. Previous chest CT scans revealed an anatomical variation consisting of persistent left and absent right
SVC (Figure 1a). Upon this admission, enhanced chest CT results showed advancement of the tumor, which almost completely encompassed the SVC (Figure 1b). Although the patient was given anti-inflammatory treatment with dexamethasone, diuresis with furosemide, and mannitol with dehydration, his signs did not resolve. Therefore, the patient was offered endovascular stenting for the SVC obstruction.

Prior to the procedure, the electrocardiogram monitoring showed a normal sinus rhythm and a heart rate of 80–90 bpm. Procedural access for catheterization was obtained through the right femoral vein. A 7-French sheath (Cordis Corp; Miami Lakes, FL, US) was inserted into the femoral vein and 3500 IU (50 IU/kg) of heparin was injected through the sheath. The stenosis was traversed with a 0.035-inch, 150-cm hydrophilic guidewire (Terumo Corp; Tokyo, Japan) and a 4-French C2 catheter (Cordis Corp) through the inferior vena cava, right atrium, coronary sinus, and persistent left SVC. An angiogram showed near complete obstruction of the proximal left brachiocephalic vein and an extensive collateral venous network bypassing the SVC into the left side azygos vein (Figure 2a). A 5-French O catheter (Cook Medical, Bloomington, IN, US) with 10 side ports was exchanged using a 0.035-inch, 260-cm Amplatz® guidewire (Cordis Corp). Angiography was again performed and the proximal stenosis was revealed. The length of the stenosis was approximately 6 cm. Stent placement was performed using a 14 × 80-mm self-expanding Smart stent (Cordis Corp), and angiography showed good patency of the obstructed region and dilatation of the coronary sinus opening (Figure 2b). Supraventricular tachycardia (160–200 beats/min) occurred repeatedly during the procedure, and this was relieved using the Valsalva maneuver and/or adjusting the position of equipment in the right atrium.

The patient received conventional anticoagulant therapies with continuous infusion of heparin (500 IU/h) immediately after the procedure for 2 days and subsequently received an oral anticoagulant agent (warfarin; titrated to an INR of 2.0) for 6 months. One day after stent placement, the symptoms of SVC obstruction completely disappeared. Two weeks after the procedure, the follow-up chest radiograph showed the stent had spread effectively (Figure 3). Although the patient again underwent chemotherapy and the tumor was advanced, he experienced no further recurrence of SVCS at 13-month follow-up until he died of advancement of the cancer.

**DISCUSSION**

Persistent left SVC is the most common venous anomaly of the thorax, affecting 0.47% of the general population (4). In most patients with persistent left SVC, the right SVC is also present (5-7). However,
very rarely the right SVC is absent, as in the case presented here. This anatomical variation presented a left SVC that drained the venous blood from both upper extremities and the head into the right atrium through the coronary sinus. In our case, the left side azygos vein was also present. The embryology of a persistent left and absent right SVC involves absent regression of the fetal left anterior cardinal vein and regression of the right anterior cardinal vein (7). In most cases, the left SVC drains into the right atrium through the coronary sinus, but in a few cases, it drains into the left atrium, even if the coronary sinus has developed normally, creating a right-to-left shunt (5,6). This anomaly may cause difficulty when introducing central venous catheters, pacemakers, or defibrillators (4–7).

SVCS with persistent left SVC is rare. Stavropoulos et al. (8) reported a case of SVCS following a Mustard repair for congenital heart disease in a patient who underwent a direct end-to-end anastomosis of the left SVC to the left atrial appendage. Jolly et al. (9) reported a case of SVCS after heart transplantation who underwent stenting for obstruction at the anastomotic site of the donor SVC and native persistent left SVC. To the best of our knowledge, this is the first report of stenting for malignant obstruction with persistent left and absent right SVC.

Miller et al. (10) reported that the subclavian approach was advantageous when stenting for the management of SVCS using the self-expanding Wallstent. However, a variety of nitinol stents with little contraction have been used in SVC stenosis, as in our case. The femoral approach is typically used when stenting for SVCS. In our case, the left SVC was easily catheterized using a Cobra catheter with a guidewire via the femoral approach through the inferior vena cava, right atrium, and coronary sinus. The main complication encountered, repeated supraventricular tachycardia, might have been caused because the equipment more easily stimulated the wall of the right atrium, atrioventricular node, or both because of the femoral approach. Performing the procedure via an upper approach may decrease this complication.

CONCLUSION

We report the first case of successful stenting for malignant obstruction with persistent left and absent right SVC. It is important to perform pre-therapeutic evaluation of the anatomy of the SVC when planning and performing interventional procedures in the thorax.

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