Case Reports

Endovascular Stenting in 2 Patients with Benign Superior Vena Cava Syndrome

Jared B. Hooker, MD
Beau M. Hawkins, MD
Mazen S. Abu-Fadel, MD

Superior vena cava syndrome has typically been associated with malignant conditions; however, the number of benign cases has started to grow as the use of upper-extremity venous lines and implantable cardiac devices increases. Whereas endovascular techniques are standardly used to treat patients with malignancies, the optimal care of patients with benign causes is less clear because they typically have longer life expectancies. We describe 2 cases of benign superior vena cava syndrome successfully managed with endovascular stenting, and we review the relevant literature. Of 145 cases in 10 series (average follow-up time, 24 mo), 96% of patients experienced symptomatic relief after endovascular management, with a primary patency rate of 66% and a secondary rate of 93%. Although few data exist to compare open surgical and endovascular techniques directly, both approaches appear to produce similar rates of patency. Both approaches frequently necessitate secondary intervention to maintain patency, but endovascular management is associated with fewer complications. We conclude that endovascular management of benign superior vena cava syndrome is a safe, effective, and reasonable initial management approach. ***(Tex Heart Inst J 2018;45(4):264-9)***

Key words: *Blood vessel prosthesis implantation/instrumentation; chest pain/etiology; dyspnea/etiology; edema/etiology; stents; superior vena cava syndrome/diagnostic imaging/etiology/pathology/therapy; treatment outcome; vascular patency*

From: *Department of Internal Medicine (Dr. Hooker) and Cardiovascular Diseases Section, Department of Internal Medicine (Drs. Abu-Fadel and Hawkins), University of Oklahoma College of Medicine, Oklahoma City, Oklahoma 73104*

Address for reprints: *Mazen S. Abu-Fadel, MD, Cardiovascular Diseases Section, University of Oklahoma College of Medicine, Andrews Academic Tower, Suite 5400, 800 Stanton L. Young Blvd., Oklahoma City, OK 73104*

E-mail: *Mazen-Abufadel@ouhsc.edu*

An obstruction that impedes venous flow from the face or upper extremity to the right side of the heart may cause superior vena cava (SVC) syndrome. Symptoms include facial or upper-extremity edema, dyspnea, hoarseness, headaches, and dysphagia. From the time of its description in the 18th century through the 1990s, most SVC syndrome cases were attributed to malignancies.[1] In recent years, the number of benign SVC syndrome cases—that is, SVC occlusion from causes other than malignancy—has started to increase, mainly because the use of upper-extremity venous lines and implantable cardiac devices has increased.[1] Although benign SVC syndrome generally presents less acutely than the malignant syndrome does, many patients have severe symptoms and need aggressive treatment.

Optimal treatment guidelines for benign SVC syndrome are not yet established, and they may vary from those for malignant cases. Patients with benign SVC syndrome are generally younger and have longer life expectancies than those with malignant obstructions, making long-term patency of utmost importance. We describe the cases of 2 patients with benign SVC syndrome who underwent endovascular treatment of SVC occlusion, and we review the literature pertaining to these issues.

Case Reports

Patient 1

A 42-year-old woman presented with a one-month history of dysphagia, face and neck swelling, and dyspnea. She had a history of renal cell carcinoma and had undergone partial nephrectomy. She also had recurrent genitourinary infections that necessitated prolonged intravenous antibiotic therapy, administered through a remote right subclavian port, which extended to the SVC–right atrial (RA) junction.

Physical examination revealed ptosis of the patient's left eye and edema affecting the left side of her face, neck, and chest. Upper-extremity duplex ultrasonography revealed deep vein thrombosis (DVT) involving the left internal jugular, left brachiocephalic, left subclavian, and left axillary veins. In addition, diminished phasicity on her right upper extremity suggested a proximal obstruction. Computed tomographic (CT) an-

giograms of the patient's chest showed right subclavian vein thrombosis associated with an indwelling port, thrombosis at the confluence of the left subclavian and jugular veins, thrombus in the azygos vein, and subcutaneous collateral vessels that had not been seen on CT 4 months previously. The patient was given low-molecular-weight heparin for anticoagulation.

The patient's symptoms and the findings on the CT angiograms suggested SVC syndrome. Venograms confirmed an occlusion at the SVC–RA junction (Fig. 1A–B). In addition, the bilateral brachiocephalic veins, left subclavian vein, and distal left-arm deep veins were occluded with thrombus. A 5F right internal jugular venous sheath and a 6F left basilic venous sheath were placed. An angled 0.035-in Glidewire® (Terumo Interventional Systems) and a 0.035-in support catheter were inserted through each sheath and advanced across the SVC–RA obstruction. After venoplasty with small-caliber balloons, ultrasound-facilitated catheter-directed thrombolysis was administered in standard fashion through the left arm access, beginning with a 5-mg slow bolus dose of tissue plasminogen activator, followed

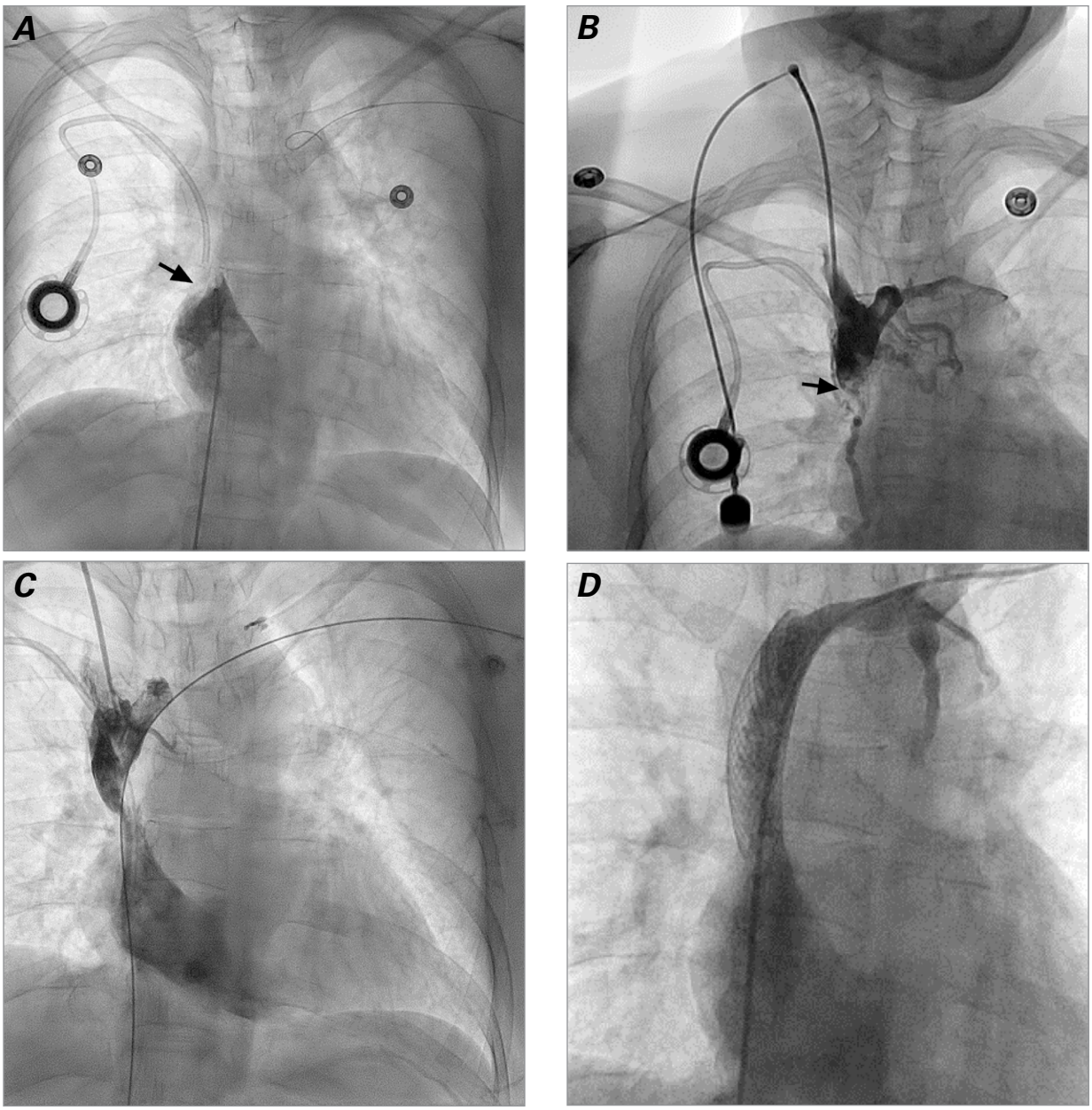

Fig. 1 *Patient 1. Venograms after injection of contrast medium through the* ***A****) right atrium and* ***B****) right internal jugular vein show superior vena caval occlusion (arrows).* ***C****) Venogram after catheter-directed thrombolysis and angioplasty shows substantially reduced thrombus burden and some residual stenosis at the superior vena cava–right atrial junction.* ***D****) Final venogram shows the stented superior vena cava; the flow was so brisk that the entire caval width could not be fully opacified with a unilateral injection.*

by 1 mg/hr for 8 hours, then 0.5 mg/hr for 16 hours. The next day, venograms showed that the thrombus burden was substantially reduced (Fig. 1C), but there was residual stenosis at the SVC–RA junction. Further endovascular treatment was suspended until the right-upper-extremity port could be removed 3 days later.

Ten days after the initial procedure, the patient was taken to the catheterization laboratory to complete therapy for SVC syndrome. Right femoral access was obtained, and a 10F sheath was placed. The residual SVC–RA junction stenosis was predilated with a 14-mm balloon, and an 18 × 40-mm Wallstent® (Boston Scientific Corporation) was deployed. After postdilation with a 16-mm balloon, the final venogram revealed excellent results (Fig. 1D). The SVC was widely patent, and there was no residual stenosis.

Six weeks after stenting, the patient presented at the emergency department with swelling of her left upper extremity and left side of her face. Computed tomographic angiograms confirmed that the SVC stent was patent but showed thrombosis of the left axillary vein, which raised concern about thoracic outlet syndrome. One week later, our vascular surgeons performed left pectoralis minor tenotomy to relieve compression of the axillary vein. Eighteen months after the procedure, a repeat ultrasonogram showed no evidence of recurrent obstruction.

Patient 2

A 25-year-old man had been diagnosed with a neuroectodermal brain tumor when he was in his early teens, and he had undergone chemotherapy and radiation therapy. Approximately 10 years later, the tumor recurred, and a port was placed in his left subclavian vein for repeat chemotherapy. Approximately 18 months later, the patient presented at the emergency department with dyspnea and swelling of his face, neck, and upper chest. Upper-extremity duplex ultrasonograms showed DVT involving the right internal jugular, right brachiocephalic, right subclavian, right axillary, left subclavian, and left axillary veins. The left-sided thrombosis was associated with the patient's indwelling venous port. He was started on therapeutic low-molecular-weight heparin, with plans to remove the port after anticoagulation therapy.

Nine months later, the port was removed. Duplex ultrasonograms 3 months later showed that the upper-extremity DVT had resolved but suggested that a proximal obstruction was present. The patient had minimal symptoms, so he did not wish to pursue intervention at that time. Six months later, he presented at the emergency department with dyspnea on exertion and worsening swelling of his face, neck, and shoulders (more severe on his left side).

Venograms showed distal SVC occlusion near the RA junction (Fig. 2A–B), as well as occlusion of the left brachiocephalic and right internal jugular veins. The

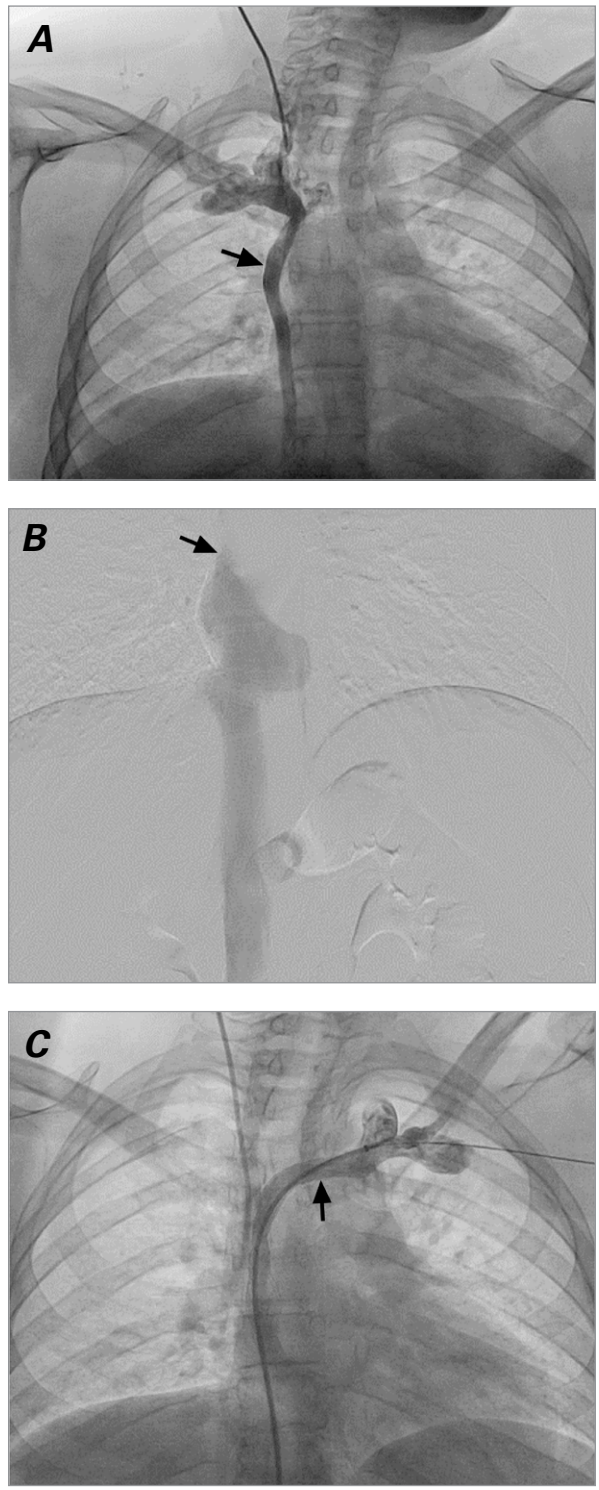

Fig. 2 *Patient 2.* ***A****) Venogram after injection of contrast medium through the right internal jugular vein shows no obvious stump of the superior vena cava, and a large collateral vessel (arrow) diverts blood to the inferior vena cava.* ***B****) Venogram of the inferior vena cava and right atrium shows superior vena caval occlusion (arrow).* ***C****) Venogram shows successful recanalization, with flow through the left brachiocephalic vein (arrow). Note that after injection of contrast medium into the left brachiocephalic vein, the right internal jugular vein is not filled because of competitive flow.*

right internal jugular vein drained through a large collateral vessel that extended down through the chest and abdominal wall, into the right femoral vein. Using a 6F sheath inserted through the left common femoral vein, we crossed the SVC occlusion by using a 0.035-in angled Glidewire and a support catheter. The Glidewire was then snared and externalized through a 6F left basilic sheath. After predilation with a 4 × 40-mm balloon, a 14 × 80-mm Protégé™ (Medtronic) self-expanding stent was deployed and postdilated. The final venogram revealed restored blood flow through the SVC and the left brachiocephalic vein (Fig. 2C).

The patient's symptoms rapidly improved; one week after stenting, his edema and dyspnea had mostly resolved. Thirty months later, he remained asymptomatic, and a duplex ultrasonogram showed no evidence of proximal obstruction.

Discussion

Our 2 cases illustrate the usefulness of percutaneous intervention in patients with benign SVC. The patients had symptoms of SVC occlusion caused by indwelling central venous catheters, and endovascular stenting resulted in complete resolution.

Before antibiotics were available, infections were responsible for most cases of SVC syndrome.[1] By the late 1990s, thoracic malignancy, typically non-small-cell lung cancer, caused about 90% of cases,[2] but, more recently, the frequency of cases from benign causes has increased. In 2006, Rice and colleagues[1] reported that 40% of patients in their study had benign SVC syndrome: of those patients, 71% had an intravascular device, followed by 8% who had fibrosing mediastinitis. Our experience with 2 patients who had indwelling catheters prompted us to review the literature. We found 10 reports that included 145 patients with benign SVC syndrome; 59.1% of those cases were associated with indwelling intravascular catheters or pacemaker wires (Table I).[3-12] As more patients undergo long-term treatment with indwelling catheters or cardiac devices with external leads, this trend is expected to continue.

Endovascular management of malignant SVC syndrome has become standard, but whether this is the best method for treating benign cases has been questioned. Patients with benign SVC syndrome are typically

TABLE I. Reported Outcomes in 145 Cases of Endovascular Management of Benign SVC Syndrome

Reference	No. of Patients*	CVC or Wire Present (%)	Symptom Relief (%)	Follow-Up Interval** (mo)	Primary Patency Rate (%)	Secondary Patency Rate (%)	Complications (%)
Rosenblum J, et al.[3] (1994)	6	100	100	5–24 (range)	100	100	0
Kee ST, et al.[4] (1998)	16	12	88	17	77	85	10
Qanadli SD, et al.[5] (1999)	12	75	100	11	92	100	0
Smayra T, et al.[6] (2001)	14	0	100	12	32	63	7
Bornak A, et al.[7] (2003)	9	89	100	12	67	100	0
de Gregorio Ariza MA, et al.[8] (2003)	14	21	95	31	57	100	0
Schifferdecker B, et al.[9] (2005)	7	100	100	36	100	100	0
Sheikh MA, et al.[10] (2005)	19	74	100	29	79	93	21
Barshes NR, et al.[11] (2007)	16	94	100	12	75	100	0
Rizvi AZ, et al.[12] (2008)	32	68	88	36	44	96	3
Weighted averages	—	59.1	95.5	23.1	66.2	93	5.2

CVC = indwelling central venous catheter; SVC = superior vena cava

*The data are solely for patients with benign SVC syndrome who were treated with endovascular techniques. Some of the reports also included patients with malignant SVC syndrome and experience with surgical repair.

**The follow-up periods varied among the studies. Unless otherwise indicated, the data are means or medians.

younger and healthier; therefore, they have a longer life expectancy, and they generally tolerate surgical repair with minimal morbidity.[12] However, in the last 2 decades, the results of endovascular treatment in patients with benign SVC syndrome have been favorable. In the 10 series we reviewed, the weighted average of clinical success was 95.5%.[3-12] The follow-up intervals varied among the studies, but during an average of 23.1 months, the primary patency rate was 66.2%, and the secondary rate was 93%. Only 5.2% of the patients had a complication, and most were minor; they included access-site hematoma[12] and asymptomatic stent migration.[10] However, SVC perforation did occur in rare cases[6,10,12] and occasionally resulted in cardiac tamponade.[6,12] Of importance, our review suggests that failure of primary patency is often effectively treated with repeat endovascular intervention. Although some patients in these reports had satisfactory results after angioplasty or catheter-directed thrombolysis, most needed stents, probably because their obstructions had been caused by chronic thrombus or fibrosis refractory to thrombolysis or angioplasty alone.[3-12]

Surgical treatment for benign SVC syndrome is associated with similar patency rates but a higher risk of complications. In one series, the 3-year primary patency rate after open surgical repair was 45%, and the secondary rate was 75%; the respective rates after endovascular repair were 44% and 96%.[12] In 2 studies that compared surgical and endovascular repair, the morbidity rates in the surgical groups were 19% and 14%.[12,13] In addition, a substantial number of surgical patients in both studies needed early repeat surgical or percutaneous intervention to establish patency (14% and 17%), or they needed secondary intervention during follow-up to maintain patency (26% and 28%).[12,13] Secondary intervention is also frequently needed after endovascular repair; however, endovascular procedures are associated with fewer complications. Therefore, we agree with other authors[12] that endovascular techniques are suitable for the initial management of benign SVC syndrome, and that open surgery should be reserved for lesions that are refractory to endovascular techniques.

No stents have been approved for deployment in the SVC, but several commercially available stents have been used successfully, including these models: Wallstent, S.M.A.R.T.® (Cordis, a Johnson & Johnson company), Palmaz® (Cordis), Gianturco-Z (Cook Medical Inc.), Memotherm® (Bard, no longer produced), and Symphony® (Boston Scientific). Regardless of stent, all investigators reported satisfactory results.[3-12]

No guidelines or algorithms have been established to guide follow-up care after SVC stenting. We monitor our patients' clinical symptoms and obtain venous duplex ultrasonograms or CT venograms if symptoms suggesting SVC occlusion recur. Although duplex ultrasonography does not produce satisfactory views of the vena cava, it can frequently suggest proximal obstruction by DVT, evidenced by loss of flow phasicity in the brachiocephalic or subclavian veins. In contrast, CT venography provides excellent caval images but requires contrast administration and radiation exposure, which is not ideal in younger patients. Therefore, we do not perform routine surveillance imaging in patients without symptoms, because it is unlikely to alter their management.

Antithrombotic therapy is frequently given to patients treated with SVC stents, but there is no consensus on an optimal regimen, and more data are needed.[10,12] Protocols for antithrombotic care after stenting for benign SVC syndrome are derived largely from those in the more plentiful literature on malignant SVC syndrome. However, these regimens vary widely: some authors recommend months of therapeutic anticoagulation, and others recommend short courses of antiplatelet therapy.[14] Coexistent DVT provided an absolute indication for anticoagulation in our patients. Aspirin and clopidogrel were added to their anticoagulant regimens for 4 weeks after stent placement, and then they continued taking aspirin and an anticoagulant.

References

1. Rice TW, Rodriguez RM, Light RW. The superior vena cava syndrome: clinical characteristics and evolving etiology. Medicine (Baltimore) 2006;85(1):37-42.
2. Wilson LD, Detterbeck FC, Yahalom J. Clinical practice. Superior vena cava syndrome with malignant causes [published erratum appears in N Engl J Med 2008;358(10):1083]. N Engl J Med 2007;356(18):1862-9.
3. Rosenblum J, Leef J, Messersmith R, Tomiak M, Bech F. Intravascular stents in the management of acute superior vena cava obstruction of benign etiology. JPEN J Parenter Enteral Nutr 1994;18(4):362-6.
4. Kee ST, Kinoshita L, Razavi MK, Nyman UR, Semba CP, Dake MD. Superior vena cava syndrome: treatment with catheter-directed thrombolysis and endovascular stent placement. Radiology 1998;206(1):187-93.
5. Qanadli SD, El Hajjam M, Mignon F, de Kerviler E, Rocha P, Barre O, et al. Subacute and chronic benign superior vena cava obstructions: endovascular treatment with self-expanding metallic stents. AJR Am J Roentgenol 1999;173(1):159-64.
6. Smayra T, Otal P, Chabbert V, Chemla P, Romero M, Joffre F, Rousseau H. Long-term results of endovascular stent placement in the superior caval venous system. Cardiovasc Intervent Radiol 2001;24(6):388-94.
7. Bornak A, Wicky S, Ris HB, Probst H, Milesi I, Corpataux JM. Endovascular treatment of stenoses in the superior vena cava syndrome caused by non-tumoral lesions. Eur Radiol 2003;13(5):950-6.
8. de Gregorio Ariza MA, Gamboa P, Gimeno MJ, Alfonso E, Mainar A, Medrano J, et al. Percutaneous treatment of superior vena cava syndrome using metallic stents. Eur Radiol 2003; 13(4):853-62.
9. Schifferdecker B, Shaw JA, Piemonte TC, Eisenhauer AC. Nonmalignant superior vena cava syndrome: pathophysiology and management. Catheter Cardiovasc Interv 2005;65(3): 416-23.

10. Sheikh MA, Fernandez BB Jr, Gray BH, Graham LM, Carman TL. Endovascular stenting of nonmalignant superior vena cava syndrome. Catheter Cardiovasc Interv 2005;65(3): 405-11.
11. Barshes NR, Annambhotla S, El Sayed HF, Huynh TT, Kougias P, Dardik A, Lin PH. Percutaneous stenting of superior vena cava syndrome: treatment outcome in patients with benign and malignant etiology. Vascular 2007;15(5):314-21.
12. Rizvi AZ, Kalra M, Bjarnason H, Bower TC, Schleck C, Gloviczki P. Benign superior vena cava syndrome: stenting is now first line of treatment. J Vasc Surg 2008;47(2):372-80.
13. Kalra M, Gloviczki P, Andrews JC, Cherry KJ Jr, Bower TC, Panneton JM, et al. Open surgical and endovascular treatment of superior vena cava syndrome caused by nonmalignant disease. J Vasc Surg 2003;38(2):215-23.
14. Uberoi R. Quality assurance guidelines for superior vena cava stenting in malignant disease. Cardiovasc Intervent Radiol 2006;29(3):319-22.