

Volume-dependent superior vena cava syndrome related to stenosis after central venous catheterization

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Stenosis of the central veins, most commonly due to thrombosis, represents a common and serious complication of long-term central venous catheterization.¹⁻⁶ Thickening of the vascular intima or thrombosis involving the superior vena cava (SVC), if severe enough, can present clinically as the SVC syndrome. Typically, the symptoms of SVC syndrome, including headaches, brawny edema of the upper extremities, head and neck, and prominent collateral vascularization are present while the patient is euvolemic. As a result, treatment modalities for SVC syndrome focus on restoring patency of the stenotic vessel with early fibrinolysis, thrombolytic therapy, venous angioplasty, expandable metal stenting or surgical thrombectomy.⁵⁻¹⁵ We report a case of SVC syndrome in a hypervolemic patient who was treated by restoring euvoolemia with furosemide diuresis.

Case report

A 67-year-old woman, weighing 53 kg and suffering from chronic idiopathic intestinal pseudo-obstruction (CIIP) for at least 5 years, was receiving total parenteral nutrition (TPN) through her first left subclavian 9.6-French single-lumen Hickman catheter (Bard Access Systems, Salt Lake City, Utah) for 2.5 years to supplement her usual enteral feeding. She had a 40-year pack-per-day smoking history and had undergone diagnostic

laparotomy in 1992 and in 1995. Otherwise she was well and on no regular medications. She was admitted to hospital because of abdominal pain. She became febrile and complained of fatigue, and 1 week after admission blood cultures demonstrated *Candida parapsilosis* fungemia with corresponding growth on line-tip culture. The Hickman line was removed, and amphotericin was started intravenously followed by itraconazole intravenously. The patient vomited all of her enteral feeds, so TPN was initiated through a right subclavian double-lumen silicone rubber catheter inserted without complication 3 days after the left subclavian line was removed. Two days later, facial and neck fullness as well as headaches developed and patient positioning had no effect on these findings. Ultrasonography demonstrated obstructed flow at the tip of the new subclavian venous catheter. Heparin was given intravenously to a partial thromboplastin time of 60 to 85 seconds, and the right subclavian line was replaced by a right femoral central line. Edema in the distribution of the SVC became more prominent, and growth of *Candida* persisted on blood cultures. Spiral computed tomography demonstrated mild pulmonary edema and small bilateral pleural effusions but was inconclusive for the SVC syndrome, despite brawny edema of the head, neck and upper extremities, mild dyspnea and a body weight of 59.5 kg. A ventilation-perfusion scan

demonstrated low probability for pulmonary embolism. For the next 5 days, intravenous fluids were reduced, and 40 mg of furosemide daily was administered intravenously. Her body weight dropped to 54.5 kg, and she demonstrated marked symptomatic improvement. Digital subtraction venography performed 6 days after the onset of symptoms revealed minimal intraluminal thrombotic debris. Still, there was complete occlusion of the left subclavian and innominate veins and marked stenosis of the right subclavian and innominate veins with incomplete obstruction at the superior-most aspect of the SVC (Fig. 1). There was marked bilateral collateralization both above and below the diaphragm via intercostal collateral vessels leading to the azygous system and SVC inferior to the level of obstruction (Fig. 2).

In the event of a recurrence of the symptoms of SVC syndrome, angioplasty and possible stenting were considered once her sepsis had cleared. However, even after anticoagulation was terminated, intermittent furosemide diuresis with careful assessment of volume status and maintenance of a body weight of approximately 52 kg have prevented further recurrence of SVC syndrome during her subsequent 32-month follow-up period.

Discussion

Extravascular compression of the SVC due to malignant disease of the thorax,

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such as bronchogenic carcinoma, remains the most common cause of SVC syndrome, accounting for 85% to 97% of all cases; central venous stenosis due to mediastinitis or intraluminal vascular malignant disease are other reported causes.¹⁶⁻²¹ Long-term central venous catheterization for parenteral nutrition or chemotherapy is an increasingly common cause of SVC obstruction, occurring at a rate of 3.9 cases per 100 patient-years in this population.²² The development of central venous thrombosis due to a chronically implanted catheter is commonly implicated and occurs 0.03 times for every 100 catheter-days, or in 12.9% to 33% of all TPN catheters, although only a fraction of patients are symptomatic.^{1-4,23} Even though our review of the literature produced no data on the incidence of central venous stenosis without thrombosis, venography of our patient's stenotic central veins failed to demonstrate thrombosis.

Pulmonary embolism is the most devastating complication of long-term central venous lines, particularly when they are thrombosed, occurring in approximately 12% of patients with upper extremity deep venous thrombosis.²⁴ Septic

thrombophlebitis, cardiac tamponade and acute right ventricular infarction are also complications of central lines.²⁵⁻²⁸ When central venous catheters are used for parenteral nutrition, the development of SVC syndrome has been associated with sepsis in approximately 40% of cases, and both these conditions increase the mortality associated with home parenteral nutrition and must be treated.^{22,29} This patient's *Candida* sepsis cleared with intravenous administration of amphotericin B, interrupted briefly by an experimental trial with intravenous itraconazole.

Despite promising reports of thrombolysis in patients presenting with SVC syndrome related to central venous catheter use, there was no indication for its use in this patient.¹⁰ Heparin, although initially used, was not indicated once venography demonstrated the absence of intraluminal thrombus. Angioplasty and intravascular stenting have recently become popular methods for treating central venous vascular occlusion and are associated with a low complication rate.^{5,11-15,30} Such treatment was deferred in this patient whose *Candida* sepsis had not fully cleared while still clinically demonstrating SVC syndrome. As a result, furosemide, a common loop diuretic, was used to relieve the patient's symptoms of headache, and swelling of the face and upper extremities resulting from fluid in the distribution of the SVC.

Although not previously reported in an extensive search of Medline from 1966 to the present, we chose to treat SVC syndrome with diuresis in this patient on the basis of brawny edema, positive fluid balance and shortness of breath in the setting of ongoing candidemia. Once a euvolemic state was achieved with diuresis alone, the clinical picture of SVC syndrome resolved and did not recur. This patient subsequently tolerated an enteral diet and did not require further catheterization for TPN.

In this patient, venography-proven SVC syndrome due to central venous stenosis was treated with volume control and intermittent use of furosemide. Patients with SVC syndrome should be fed enterally when possible and through a femoral venous line otherwise. The awareness of stenosis of the central venous system as a result of the long-term use of central venous access is underscored and is another possible cause of SVC syndrome in this group of patients.

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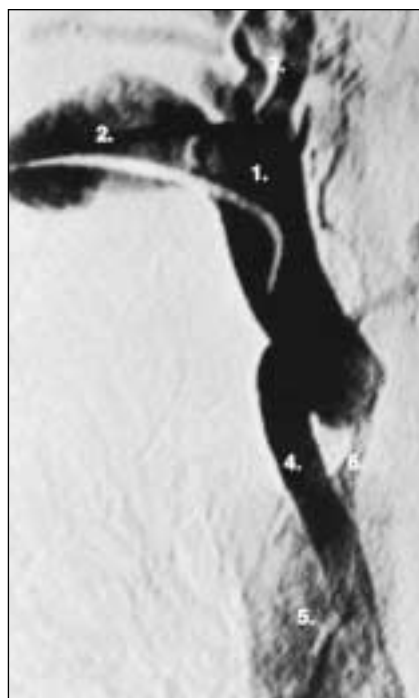


FIG. 1. Digital subtraction venography with contrast injection via the right axillary-subclavian venous system. 1 = right brachiocephalic vein, 2 = right subclavian vein, 3 = right jugular system, 4 = right azygous system, 5 = superior vena cava, 6 = superior vena cava stenosis.

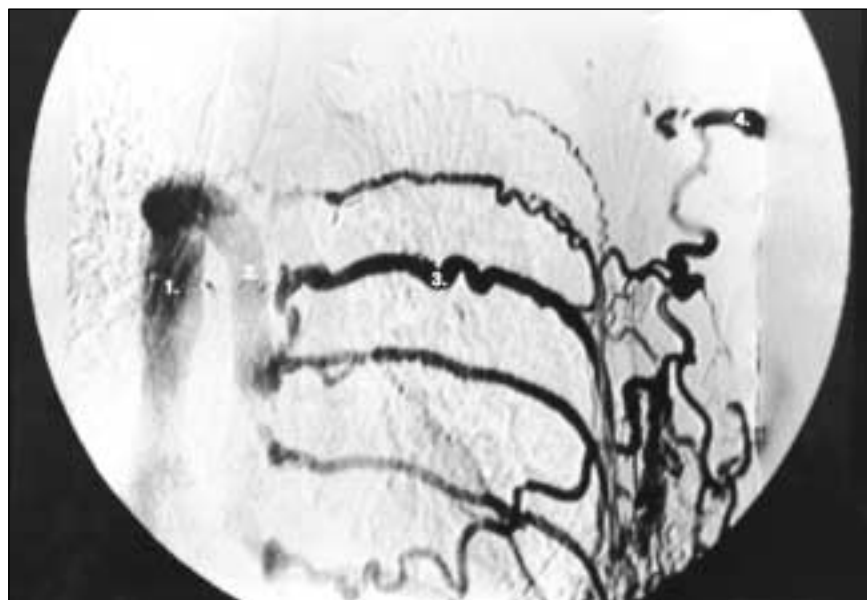


FIG. 2. Digital subtraction venography with contrast injection via the left axillary vein. 1 = superior vena cava, 2 = azygous system, 3 = dilated left intercostal veins, 4 = left axillary vein.

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