CASE STUDY

Superior Vena Cava Syndrome

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Approximately 15,000 cases of superior vena cava (SVC) obstruction are diagnosed in the United States annually. Malignancies (primarily lung cancer) are the underlying cause of 80–85% of cases, leaving 15–20% caused by various benign conditions, including sclerosing mediastinitis (the diagnosis in our case). Thrombolytic therapy and major advances in vascular techniques in recent years have improved the outcome and lessened the morbidity of SVC obstruction. However, even though a benign condition, sclerosing mediastinitis is a dynamic, ongoing fibrotic condition that seldom can be totally removed surgically. It frequently causes recurrent episodes of SVC obstruction, requiring further repetitive vascular procedures that can result in major morbidity and even mortality.

CASE PRESENTATION

A 51-year-old male applied for a $500,000 special whole life policy. On the application, he admitted to a history of a superior vena cava (SVC) obstruction with surgical correction in 1992, followed by several restenosis events requiring additional procedures in 1998 and 2000. An attending physician statement was obtained and revealed an extensive history.

In September 1991, he experienced acute chest pain, which was diagnosed as acute pericarditis by electrocardiogram (ECG) changes and echocardiogram findings of a small to moderate pericardial effusion. Also noted was mild mitral valve prolapse. There was also a history of mild hypertension treated with a diuretic. A chest x-ray and a treadmill stress test done as part of his evaluation were both normal. The etiology of his pericarditis was unknown but felt to be viral.

In September 1992, he presented to his physician with facial and neck swelling and prominent facial flushing, especially when bending over. He also was experiencing mild lightheadedness and occasional headaches. His symptoms had been steadily increasing for approximately 2 months. His physical exam was described as normal, with a comment by his physician that he had the patient bend over and try to reproduce his symptoms but with unimpressive results except for perhaps mild flushing. His work-up at that time included the following normal labs: a chemistry profile, CBC, thyroid profile, and sedimentation rate. He continued to experience worsening symptoms and 2 weeks later had a chest x-ray, which was read as normal and unchanged since his 1991 x-ray. Further evaluation for the continued flushing episodes revealed normal results for 24-hour urinary catecholamines, 5 HIAA, VMA, metanephrines, and free cortisol. An abdominal CT scan for adrenal evaluation was also normal. At this point, his physician first entertained the possibility of a thoracic inlet cause such as con-
strictive pericarditis or SVC obstruction as the basis for his symptoms.

Within the next 2 weeks, additional symptoms of a rushing sensation in his head with exercise plus worsening headaches and further elevation of his blood pressure (156/100) led to a chest CT, which revealed apparent obstruction with thrombus formation in the SVC and both right and left brachiocephalic veins. No mass lesion was present in the mediastinum or elsewhere in the chest. A venogram of the SVC and brachiocephalic veins revealed concentric narrowing of the proximal SVC resulting in approximately 75% stenosis of the SVC, but the brachiocephalic veins appeared moderately dilated and free of thrombus or stenosis.

In December 1992, he was taken to surgery. Upon entering the chest, the surgeon noted he had moderate pericardial adhesions from his previous pericarditis. These adhesions were lysed and the surgeon proceeded to expose the SVC and left brachiocephalic vessels. A dense mass was found surrounding the proximal SVC. Frozen section biopsies of this mass revealed intense fibrous reaction but no malignancy. An attempt to separate this mass from the SVC was unsuccessful since no separation plane could be accomplished in the very dense fibrous growth. Fortunately, it was possible to separate the mass from the ascending aorta. Because the SVC obstruction could not be relieved, a Goretex graft was placed between the left brachiocephalic vein and the SVC beyond the obstructed area. He was postoperatively placed on continuous coumadin therapy, maintaining his INR between 2 and 3.

Final pathologic determination of the mass was that of "sclerosing mediastinitis" (SM). Clinically, he did well for 6 years. However, in early 1998, symptoms of plethora, facial edema, flushing, headaches, and increasing blood pressure returned. Studies revealed new obstructions of the graft and the left brachiocephalic vein. An angioplasty was performed with satisfactory results and relief of symptoms. There was return of his symptoms 10 months later. His evaluation now revealed obstructions of the SVC and the graft. These obstructions were successfully reopened by angioplasty, and stents were placed in both brachiocephalic veins and the SVC. He was continued on coumadin and did well until December 2000, when once again symptoms returned. The repeat vascular studies revealed stenosis at both ends of the left brachiocephalic stent and mid-stent stenosis of the right brachiocephalic stent. Restenting with improved, more rigid stents was performed successfully. He remained on adequate coumadin therapy plus pletal until the time of his application for life insurance.

CASE DISCUSSION

Approximately 15,000 cases of SVC obstruction occur annually in the United States, and predictions are that the incidence will increase due to the marked increase in central venous catheter and pacemaker wire usage.1,2 The SVC is susceptible to obstruction for many reasons, including its thin-walled structure, low pressure flow, and close proximity to lymph nodes, lung, and other major vascular structures. Various malignant diseases are by far the most common cause of SVC obstruction (80–85% of all cases), with lung cancer itself accounting for 80% of these malignant cases. In fact, 3–5% of lung cancer patients develop the SVC syndrome sometime during their illness. More important for life insurance considerations are the 15–20% of SVC obstruction cases caused by benign conditions, which include the following: benign tumors (cysts, goiters, thymomas, and teratomas), inflammatory processes (SM, tuberculosis, actinomycosis, and histoplasmosis), vascular causes (aneurysm), chest trauma, radiation fibrosis, and, as mentioned above, the increasing use of catheters and pacemaker wires.3,4

The symptoms of SVC obstruction can develop very acutely or insidiously, depending on the severity of obstruction and, most importantly, on the presence or absence of collateral venous channels of decompression. When collateral development is inadequate,
as in our discussion case, any of the following list of symptoms may ensue: facial, neck, and even arm swelling; hoarseness; cyanosis; dyspnea at rest or with exertion; headaches; and in severe cases, seizures, syncope, or even coma. In a recent series review, the average time from the onset of symptoms of SVC obstruction until presentation to a physician for evaluation was several months.

When the diagnosis of SVC obstruction is suggested by the above-mentioned symptoms, the following testing procedures are utilized to help pinpoint the location, severity, and cause of obstruction: chest x-ray, CT or MRI imaging (which frequently reveals the obstructed area and the lesion of cause), thoracoscopy, venography, and duplex scanning. If a mediastinal mass is demonstrated to be the cause of obstruction, a tissue diagnosis is imperative to exclude malignancy and, if the pathology is benign, to guide therapeutic interventions such as surgical removal of the lesion. When, as in our case, SM is found, total surgical removal is frequently impossible due to the dense, adherent nature of this particular benign lesion. A recent pathology series of 30 cases of idiopathic SM revealed 3 distinct histologic stages of this dynamic benign entity: stage 1 demonstrates edematous fibromyxoid tissue composed of thin-walled blood vessels, lymphocytes, plasma cells, spindle cells, mast cells, and eosinophils; stage 2 consists of haphazardly arranged collagen with focal interstitial spindle cells, mast cells, and eosinophils; stage 3 is characterized by dense acellular collagen containing scattered lymphoid follicles and dystrophic calcifications. These unique stages strongly imply that SM is the result of a dynamic, evolving process similar to abnormal wound healing. Some investigators believe that SM is frequently the result of an abnormal immunologic response to histoplasmosis, but other as yet unknown etiologies surely exist. Because SM is an ongoing process frequently not amenable to total surgical removal and does not respond to any other known medical measures (ie, antifungals, antibiotics, steroids, immunosuppressives), it is common for patients with the SVC syndrome caused by SM to require extensive and recurrent vascular procedures to maintain adequate venous flow.

Therapeutic measures dealing with the SVC syndrome have evolved considerably in recent years due to thrombolytics, angioplasty techniques, vascular stenting, and improved surgical procedures. In a recent review of 59 consecutive patients with the SVC syndrome treated with a combination of thrombolytics and endovascular stenting, 42 had an underlying malignancy and 13 had benign causes. Four cases were lost to follow-up. Of the 13 benign cases, 11 (85%) had clinical venous patency at a mean of 17 months of follow-up. The periprocedural mortality and morbidity for the entire group of 59 patients were 3% and 10%, respectively. There is a considerable need for more quality data regarding the long-term success rate for thrombolysis and stenting of benign SVC syndrome cases, and future data must be compared with the gold standard of surgical bypass. During the decade of the 1990s, at least 5 series of patients with benign SVC syndrome treated surgically with graft procedures have been published. A review of these surgical series reveals 5-year patency rates of up to 86%. As previously stated, regardless of the choice of therapeutic modalities utilized in patients with the SVC syndrome caused by SM, it is common for them to require repetitive additional procedures to maintain adequate patency. The necessity and success of long-term anticoagulation in patients with the SVC syndrome, regardless of which therapeutic vascular procedure they received, is still being debated, with no consensus to date.

**SUMMARY**

Our case represents a fairly typical case of the SVC syndrome caused by SM. He exhibited classic symptoms, underwent an original bypass graft procedure followed by repetitive angioplasties and stenting, despite adequate long-term anticoagulation therapy, and has no distinct prior illness that could be causally
related to his SM. While increased morbidity of such a patient is obvious, the quality of long-term mortality data is sorely lacking. The overall prognosis of such a case is probably good, although statistics to definitely prove this are lacking. Certainly, careful follow-up by experienced vascular physicians is of utmost importance for such a patient.

REFERENCES