Superior Vena Cava Syndrome

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The superior vena cava syndrome (SVCS) is characterized by edema of the face, head, neck, arms, and upper trunk with dilated collateral veins, and can be accompanied by cyanosis, dyspnea, headache, and altered mental status.[1] The syndrome is confirmed by superior vena cavagram and venous pressure measurements. Often viewed as a medical emergency, SVCS is commonly a sequela of malignant thoracic neoplasm, a disease requiring immediate therapeutic attention. In patients with a known diagnosis of malignant neoplasm, further histological study is unnecessary, and aggressive management with a combination of surgical, radiation, and oncological therapies is often employed.[2] When SVCS is of unknown etiology, a full diagnostic evaluation and specific therapeutic plan are desirable. Empiric emergency thoracic irradiation to reduce assumed tumor growth and obstruction has been advocated.[2-5] The following case illustrates an unusual cause of SVCS for which such radiotherapy would have been inappropriate and possibly detrimental.

CASE REPORT

A 57-year-old man with congenital blindness, obesity, and mild hypertension, controlled with thiazide diuretics, developed swelling of his face and neck. Seven days later he presented to the emergency department with these complaints. Six weeks earlier he had developed soreness in the right neck with spontaneous resolution followed by recurrence on the left side, also resolving spontaneously. The patient denied any other problems, associated circumstances, or symptoms including trauma, dyspnea, cough, dysphagia, or chest discomfort. He was a nonsmoker for 28 years. Family history was noncontributory.

On physical examination the patient weighed 120 kg, his blood pressure was 120/70 mmHg in the right arm, and 130/70 mmHg in the left arm, and his pulse was 80 beats per min. There was obvious swelling of the face and neck with partial closure of the eyelids due to periorbital edema. No swelling was noted in the upper extremities. The chest revealed several small but prominent distended venules. The remainder of the physical examination was either normal or not associated to the current condition.

Chest x-ray examination showed a widened mediastinum with a small right pleural effusion. Results of a hemogram, urinalysis, sedimentation rate, and full chemistry profile were all within normal limits. Arterial blood gases in room air were pH 7.45, partial pressure of carbon dioxide ($PaCO_2$) 33.1 mmHg, arterial pressure of oxygen (PaO_2) 63.5 mmHg, and bicarbonate 24 mEq/L. A computed tomographic (CT) scan of the chest demonstrated complete occlusion of the superior vena cava without other masses or abnormalities. A superior vena cavagram showed complete intrinsic obstruction of the right subclavian vein and opacification of multiple collateral neck veins and the superior vena cava. Primary thrombosis became the working diagnosis.

Hematology consultation and study failed to identify a cause. Coagulation studies and bone marrow biopsy results were all normal. No source was found for hypercoagulability or occult tumor. Carcinoembryonic antigen was normal. An intravenous pyelogram, thyroid scan, bronchoscopy, barium enema, upper gastrointestinal series, and abdominal CT scan all showed no abnormality. The patient was given the anticoagulant heparin for ten days and was discharged on warfarin and hydrochlorothiazide with follow-up as an outpatient.

Four weeks later he was rehospitalized for progressive shortness of breath and marked neck edema. Breath sounds were diminished over both lower lung

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fields, and chest x-ray films showed bilateral pleural effusions. Thoracentesis revealed sterile chylous effusions with atypical cell structure but no evidence of malignancy. Repeat chest CT scan and superior vena cavagram demonstrated extension of a suspected thrombus to include the innominate and base of the internal jugular veins. Persistent reaccumulating chylous effusion was thought to be from obstruction to the thoracic duct at its entry to the major venous system. The patient was transferred to a university medical center, where he was treated with multiple chest tube drainage, intravenous streptokinase, and finally thoracic surgery in an attempt to redirect the flow from the thoracic duct. He died postoperatively.

An autopsy demonstrated occlusive thrombosis of the right subclavian vein, right internal jugular, and left subclavian veins. There was no evidence of neoplastic or hematological disease. There was no evidence of vasculitis or traumatic injury to the veins or internal structures of the thorax, neck, or arms. Additional findings at the time of death correlated with the clinically documented sequelae of thrombosis and subsequent therapeutic procedures. The final diagnosis was spontaneous thrombosis of the superior vena cava and tributory veins in the absence of known preceding trauma, catheterization, occult neoplasm, hematological disease, infection, or abnormal muscular exertion. Death occurred from the sequelae of this disease and the attempted therapies required by his clinical deterioration.

DISCUSSION

Superior vena cava syndrome is a well-recognized complication of many mediastinal conditions. Malignant neoplasm is considered the most common. Lokich and Goodman[3] estimated that 97 percent of all patients with SVCS have intrathoracic malignant neoplasm, approximately three fourths of which are bronchogenic carcinoma, with the remainder lymphoma or metastases from tumors outside the thorax. In other studies, the remaining 3 percent of cases presenting as SVCS are known to result from nonmalignant causes.[2,6] Primary clot formation is a rare cause of SVCS.[7]

Other documented causes include mediastinal fibrosis secondary to histoplasmosis, goiter, aneurysm, and radiation fibrosis.[8] SVCS is a serious complication of many diseases and usually requires prompt treatment. In most instances, however, there is sufficient time for diagnostic evaluation to permit specific therapy design.[1,9] Empiric therapy might best be reserved for those with a rapidly deteriorating clinical illness.

Thrombosis of the superior vena cava accounts for less than 1 percent of all published cases of SVCS.[3] The etiology of the thrombosis can be further divided into two general groups, primary and secondary.[10] Primary thrombosis generally occurs following exertion, activity, or trauma. Approximately one fourth of the primary thromboses arise spontaneously without a predisposing event.[11] The more prevalent, secondary thromboses have widely varied causes including local inflammation of the vein (infection, catheter, or irritating intravenous solutions) as a manifestation of a serious predisposing systemic disease (Trousseau's syndrome) or as a migratory thrombophlebitis.[10]

Whether the SVCS constitutes a medical emergency remains debatable. Schraufnagel et al[8] reviewed 107 cases of superior vena cava obstruction in adult patients, seeking information regarding the interval between onset of symptoms and treatment. They examined the complications and survival of patients with SVCS from all causes. Prognosis and response to treatment were dependent on the underlying cause of the obstruction. No support was found for the theory that superior vena cava obstruction and SVCS represent a radiotherapeutic emergency, but tracheal obstruction often accompanying SVCS may require emergency management.

The question remains whether the fears of impending disaster should provoke emergency empiric treatment for SVCS. Shimm et al[1] reported 28 patients presenting with SVCS who tolerated invasive and noninvasive diagnostic procedures without major complications. Teirstein[9] editorializes that "in almost every patient presenting with SVCS, therapy should begin after the indicated diagnostic procedures are completed. With the diagnosis in hand, the physician may proceed immediately with the most appropriate therapy and a cogent, thoughtful treatment plan for the future." Experience with this patient supports Teirstein's view.

References

1. Shimm DS, Logue GL, Rigsby LC: Evaluating the superior vena cava syndrome. JAMA 1981; 245:951-953
2. Perez CA, Presant CA, Van Amberg AL III: Management of superior vena cava syndrome. Semin Oncol 1978; 5:123-134
3. Lokich JJ, Goodman R: Superior vena cava syndrome: Clinical management. JAMA 1975; 231:58-61
4. Davenport D, Feree C, Blake D, Raben M: Response of superior vena cava syndrome to radiation therapy. Cancer 1968; 38:1577-1580
5. Salsali M, Cliffton EE: Superior vena cava obstruction with lung cancer. Ann Thoracic Surg 1968; 6:437-442
6. Gomes MN, Hufnagel CA: Superior vena cava obstruction. Ann Thoracic Surg 1975; 20:345-359
7. Ochsner A, Dixon JL: Superior vena caval thrombosis. J Thoracic Surg 1936; 5:641-672
8. Schraufnagel DE, Hill R, Leech JA, Pare JAP: Superior vena caval obstruction: Is it a medical emergency? Am J Med 1981; 70:1169-1174
9. Teirstein AS: Diagnostic procedures in patients with superior vena cava syndrome. JAMA 1981; 245:956
10. Tilney NL, Griffiths HJG, Edwards EA: Natural history of major venous thrombosis of the upper extremity. Arch Surg 1970; 101:792-796
11. Prescott SM, Tikoff G: Deep venous thrombosis of the upper extremity. A reappraisal. Circulation 1979; 59:350-355