Superior Vena Cava Syndrome: A Case Report

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We report a case of superior vena cava syndrome (SVCS) caused by squamous cell lung carcinoma in a 49-year-old man. He presented with severe edema of the face and upper half of the body with numerous dilated and tortuous cutaneous veins. Although we initially considered this case to be a drug eruption because the patient had repeated episodes of worsening clinical presentation whenever he took nonsteroidal anti-inflammatory drugs (NSAIDs), persistent and characteristic dermatologic findings led us to the diagnosis of SVCS. After balloon angioplasty of the left subclavian vein to the SVC, dermatologic findings markedly disappeared. Bronchoscopy and biopsy results revealed underlying squamous cell lung carcinoma.

Cutis. 2006;77:305-309.

Superior vena cava syndrome (SVCS) presents as edema and dilatations of superficial veins in the face and upper half of the body, as well as dyspnea, headache, dizziness, and syncope. These signs and symptoms result from a disturbance of venous flow from the upper half of the body caused by SVC obstruction due to compression, invasion, or thrombus formation. Mediastinal invasion by bronchogenic carcinoma is the most common cause of SVCS, accounting for 70% to 80% of all cases.[1-4] Unfortunately, dermatologists infrequently see patients with SVCS at their initial presentation, and there have been few reports of SVCS in dermatologic journals.[2,5,6] We report a case of SVCS caused by squamous cell lung carcinoma where the patient, at his initial visit, presented characteristic cutaneous features that rapidly disappeared after balloon angioplasty and venous stenting.

Accepted for publication October 25, 2005.

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The authors report no conflict of interest.

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Case Report

A 49-year-old Japanese man presented with severe edema that appeared to be limited to the face and upper half of the body with extensive purple-pink, scattered, bifurcated vasodilatation from the chest to the back and upper extremities without pain and itching (Figures 1, 2, and 3). The patient had dyspnea in the supine position with a blood pressure level and pulse rate within reference range and no palpable superficial lymph nodes. He had lost 7 kg of his weight. Laboratory evaluations taken at initial presentation had results within reference range except for an elevated level of ferritin (280 ng/mL; reference range, 10–250 ng/mL). Results of a chest x-ray showed no abnormal shadow. Results of a skin biopsy from a specimen of the chest vasodilatation revealed only dilatation of vessels in the upper dermis with no infiltration of inflammatory cells.

About 2 months before his initial presentation, the patient had taken ceftibuten 600 mg/d and diclofenac sodium 75 mg/d orally for pleuritis for a week, after which 3 lesions of 2 to 3 cm in diameter appeared on his chest with bifurcated vasodilatation. One month later, after he took oral mecobalamin 1500 μg/d, eperisone hydrochloride 150 mg/d, and zaltoprofen 240 mg/d for ischialgia, facial edema appeared and the vasodilated lesions on his chest were extended. However, the facial edema rapidly disappeared with the discontinuation of all drugs except mecobalamin. Two weeks later, after he took oral ibuprofen 600 mg/d for symptoms of a common cold, severe edema of the upper half of the body including the face reappeared, with vasodilated lesions extending from the upper extremity to the back.

Because the patient's symptoms worsened whenever he took nonsteroidal anti-inflammatory drugs (NSAIDs), we initially diagnosed the patient with a drug eruption by NSAIDs. All of his previous medications were discontinued and administration of prednisolone 30 mg/d was started. Despite treatment

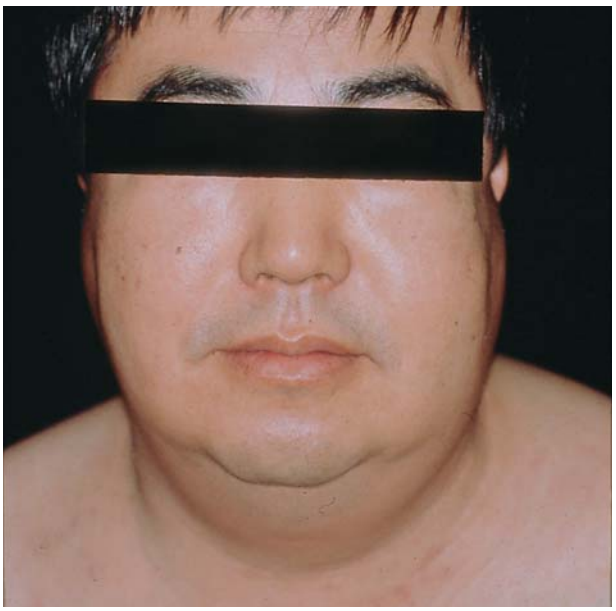

Figure 1. Facial and neck edema.

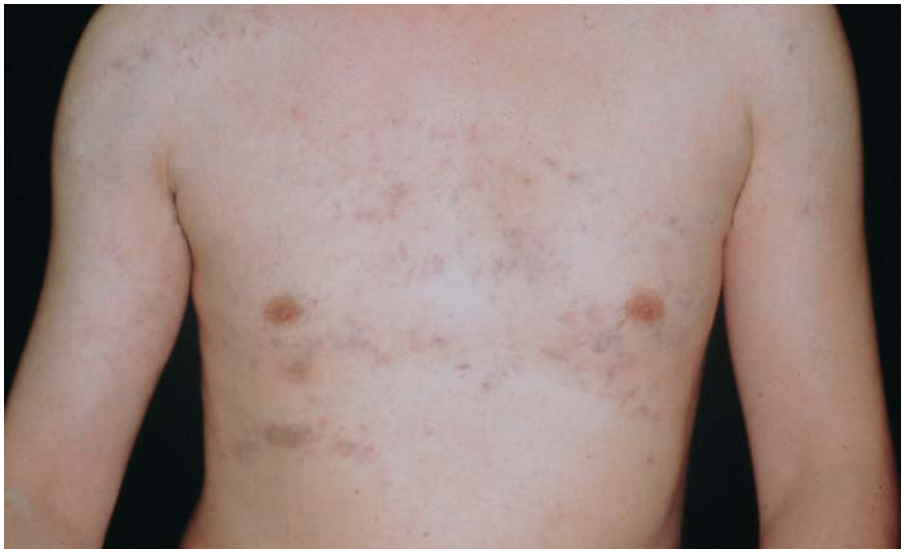

Figure 2. Upper body edema with mats of dilated vessels on the chest wall and upper extremities.

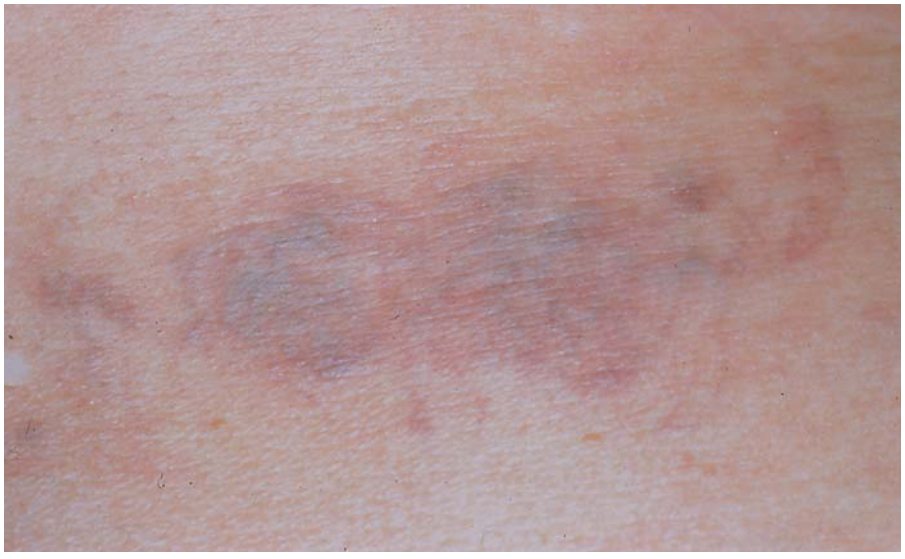

Figure 3. Dilated superficial vessels.

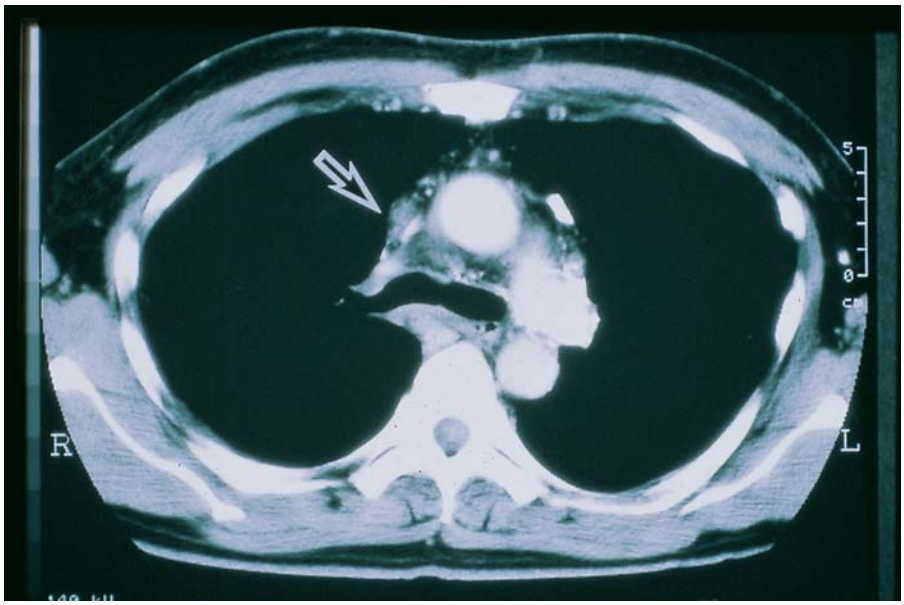

Figure 4. Compression and stenosis of superior vena cava by surrounding tissues, including enlarged mediastinal lymph nodes at the T5 level indicated by the arrow, revealed by chest computed tomography.

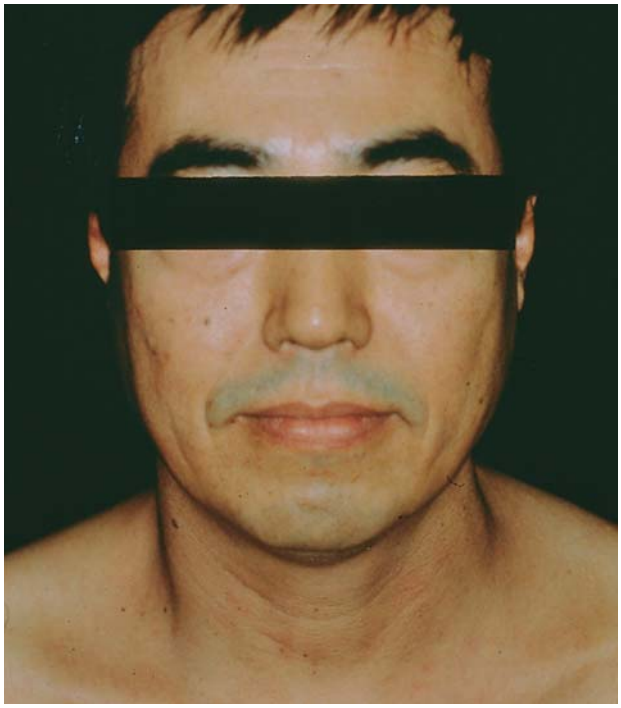

Figure 5. Resolution of facial and neck edema after balloon angioplasty and venous stenting.

with a total dose of 180 mg of prednisolone for a week, the symptoms were not alleviated. Therefore, a diagnosis of SVCS was considered because of the characteristic dermatologic features (eg, localized edema of the upper half of the body, vasodilatation). The patient was referred to the Department of Medicine at Nippon Medical School, Tokyo, Japan, and a general examination was performed. A computed tomography of the chest revealed compression and stenosis of the SVC by surrounding tissues, including enlarged mediastinal lymph nodes at the level of T5 (Figure 4). Superior venacavography from the ambilateral medial cubital veins showed compression and stenosis of the ambilateral brachiocephalic veins. Collateral circulation from the distal site of the stenosis also was seen. The veins flowing to the azygos system and the left internal thoracic vein were dilated. On the basis of these findings, we made a diagnosis of SVCS. Angioplasty then was performed with 6- and 8-mm balloon catheters introduced from the right subclavian vein to the SVC. After the balloon angioplasty was introduced from the left subclavian vein to the SVC, 2 stents (10 mm in diameter and 7 cm in length, and 7 mm in diameter and 10 cm in length) were placed. These procedures provided good recovery of circulation and rapid improvement of symptoms (Figures 5 and 6). A bronchoscopy then was performed and a protruded lesion was found on the bifurcation of the trachea. Biopsy of a specimen from the lesion revealed squamous cell carcinoma, and the diagnosis of stage IIIB squamous cell lung carcinoma (T4, N3, M0) was made. The patient was treated with radiotherapy, as well as chemotherapy, which resulted in complete remission; no recurrence of SVCS has been detected.

Comment

SVCS predominantly is caused by cancer and usually reflects end-stage disease. Mediastinal invasion by bronchogenic carcinoma is the most common cause of SVCS, accounting for 70% to 80% of all cases.[1-4] The remainder of cases are due to other malignant causes, including lymphoma, breast cancer, malignant

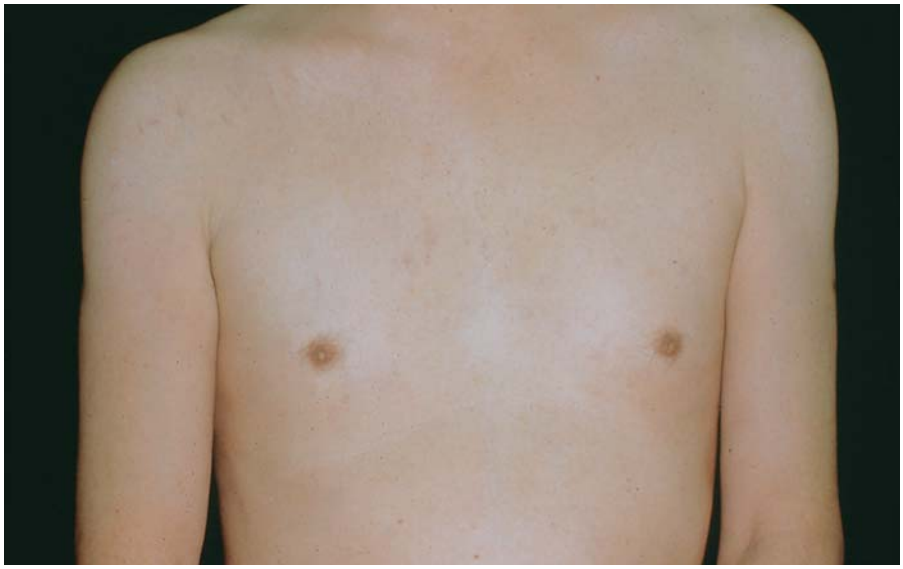

Figure 6. Resolution of upper body edema and mats of dilated vessels after balloon angioplasty and venous stenting.

thymoma, or seminoma, and benign causes, including mediastinal fibrosis, thrombosis, inflammatory nodes, or Behçet disease.[7-9] The most characteristic and often earliest skin finding is dilated, tortuous, and palpable venules or veins in the upper half of the body, which are caused by increased collateral flow through the subcutaneous chest wall veins.[2] Venous congestion of SVC may make the head, upper extremities, and trunk edematous, cyanotic, and ruddy. Our patient showed typical and characteristic skin manifestations of SVCS, with severe facial and upper body edema, as well as dilated and tortuous veins, on the upper half of the body. This case was dramatic; the obvious resolution of these features rapidly occurred after balloon angioplasty and venous stenting. The differences in clinical presentation (between Figures 1–3, and Figures 5 and 6) indicated the high degree of SVC obstruction and venous congestion of this patient.

We initially considered this case to be a drug eruption by NSAIDs because the patient's edema appeared whenever he took NSAIDs and disappeared after they were discontinued. NSAIDs can produce edema in up to 5% of the general population.[10] Cyclooxygenase-2 inhibition by NSAIDs mainly may reduce renal medullary blood flow and subsequently diminish water and sodium excretion, which contribute to the development of edema.[11,12] Similarly, 8 patients with edema caused by zaltoprofen have been reported in Japan, with facial edema occurring in 5 cases.[13] Additionally, edema as an ibuprofen-induced side effect has been reported in 29 patients in Japan with facial edema occurring in 26 cases.[14] In our patient, although impressive facial and upper body edema initially made us consider a diagnosis of drug eruption by NSAIDs, this case rapidly was differentiated in association with another characteristic skin manifestation of SVCS: dilated, tortuous, superficial vessels of the chest wall.

Other than the characteristic skin presentations, SVCS includes dyspnea, headache, dizziness, and syncope. The severity of these symptoms depends on the degree of SVC narrowing and the speed at which it occurs.[1] A malignant neoplasm often has rapid progression, and SVC obstruction frequently develops before there has been time for collateral veins to enlarge. In our patient, the chest computed tomography and venography results led us to consider that squamous cell carcinoma from the mediastinal bronchus progressed and gradually grew in the mediastinum, compressing the ambilateral brachiocephalic veins. However, there might have been enough time for the patient to develop collateral circulation during this process, which differs from the signs of acute onset. Therefore, our patient had no severe symptoms, such as lower level of consciousness and circulatory failure.

Patients with SVCS can be extremely uncomfortable or may develop life-threatening complications, such as laryngeal or cerebral edema. The most common underlying disease of SVCS is a malignant neoplasm, the histology of which affects the prognosis of the patients. Therefore, timely diagnosis and management are critical. Unfortunately, because dermatologists have few chances to see SVCS at the initial presentation, SVCS is an uncommon entity. Moreover, these skin presentations are indirect

dermatologic signs of an underlying malignant neoplasm. However, for 60% to 85% of patients with SVCS, skin changes are the initial signs and patients can seek dermatologic consultation at their initial visit.[2] We think early detection of the characteristic skin manifestations by dermatologists, which leads to rapid findings and treatment of underlying malignant neoplasm, is crucial for improving the prognosis of patients with SVCS.

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