What Is Your Diagnosis?

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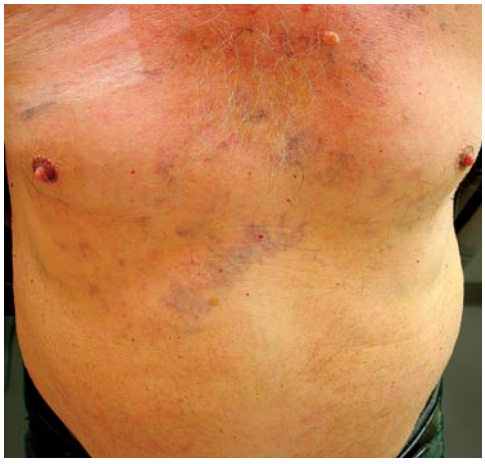

A poor postoperative cosmetic result of a left infraorbital squamous cell carcinoma. Ruddy erythema and facial plethora were present. Superficial collateral veins emerged as grossly visible varicosities on the chest.

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The authors report no conflict of interest.

The Diagnosis: Superior Vena Cava Syndrome

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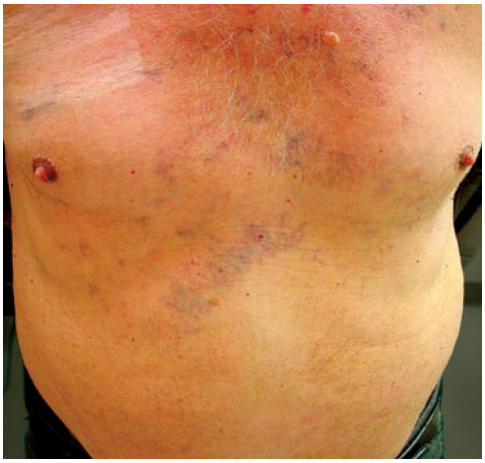

A 70-year-old man returned to clinic 4 weeks after excision of a left infraorbital squamous cell carcinoma with a poor postoperative cosmetic result. In addition to skin cancer and numerous actinic keratoses, the patient had a history of rosacea, hypertension, and chronic obstructive pulmonary disease. He complained of increasing dyspnea and intermittent chest pain on deep inspiration that started 1 week after surgery. He also reported headaches exacerbated by bending at the waist and bloodshot eyes since the surgery. He denied trauma or pain at the excision site and had followed a proven wound care regimen. He took no new medications over the past 6 months other than 5 mg midazolam hydrochloride administered before his surgery.

Superior vena cava (SVC) syndrome is a condition in which hypertension of the vessels feeding the SVC is caused by increased hydrostatic pressure from SVC flow obstruction. In this patient, the combination of pulmonary symptoms and examination of dilated angulated veins of the upper trunk and face overlying ruddy, erythematous, edematous skin explained the poor cosmetic result and prompted pulmonary examination, which revealed right-sided bronchial breath sounds and egophony. Computed tomography showed a 6.2-cm density in the right hilum compressing the SVC, and subsequent biopsy was consistent with squamous cell carcinoma of the lung.

SVC syndrome often is the presenting symptom of underlying mediastinal disease obstructing the venous drainage from the head, neck, and upper extremities.[1] Patients with early SVC syndrome frequently seek consultation for cutaneous signs, such as subacute facial plethora, and may lack the characteristic symptoms of SVC syndrome. Recognizing the dermatologic findings associated with SVC flow obstruction can aid in the timely diagnosis of an underlying malignancy. In those patients presenting to the dermatologist with facial plethora and swelling, the leading differential diagnosis often is rosacea. Other considerations in patients with a similar constellation of symptoms include angioedema, actinic dermatitis, irritant contact dermatitis, drug reaction, photodermatitis, and dermatomyositis.[2] Rare diseases causing facial edema, such as Melkersson-Rosenthal syndrome or Morbihan disease, may be considered in the remaining undiagnosed cases.[3] We found no reports of poor postoperative healing leading to a new diagnosis of SVC syndrome, but the condition is likely underreported, as the importance of cosmesis is overshadowed by potential mortality.

The SVC is particularly vulnerable to extrinsic compression because of its relatively thin wall and the rigid confines of its surrounding anatomy.[4] The severity of symptoms is largely dependent on the rate of obstruction and presence of collateral vessel formation. In slower growing tumors, obstruction of blood flow develops gradually, and collateral vessels may arise to accommodate venous return to the heart.[4] These superficial collateral veins, most commonly in the trunk, become increasingly dilated and tortuous, eventually emerging as grossly visible varicosities. They may be quite numerous, appear rather suddenly, and often are vertically oriented.[5] Alternatively, a rapidly enlarging malignant mass may result in

substantial obstruction before collateral veins sufficiently develop, resulting in an acute onset of facial or upper extremity edema, the classic symptom of SVC syndrome. Patients with malignant conditions tend to present more promptly, within days to weeks, than do patients with benign conditions.

Although first described by Hunter[6] in conjunction with a syphilitic aneurysm of the aorta, infectious causes of SVC syndrome are now quite rare.[7] Malignancy comprises the vast majority of cases, with more than 85% of cases attributed to an intrathoracic malignant mass compressing or invading the SVC.[4] Bronchogenic carcinoma (67%−82% of cases), particularly of the small cell variety, and lymphoma (5%−15% of cases) are responsible for the vast majority of these malignant masses[5]. Thymomas and metastatic malignancies invading regional lymph nodes also are well-recognized causes. Metastatic lesions are most likely to be associated with primary malignancies of the breasts, testicles, kidney, bladder, uterus, ovaries, or pharynx.[5] Despite the overwhelming etiology from tumor infiltration or compression, the incidence of nonmalignant etiologies has increased due to widespread use of permanent central venous catheters, chemotherapy success, and the common placement of multilead implantable pacemakers and defibrillators that can lead to in situ venous thrombosis and fibrosis.[8] Other conditions include mediastinal fibrosis, retrosternal goiter, aortic aneurysm, and thrombosis.[9]

Dyspnea secondary to laryngeal edema is the most common presenting symptom of SVC syndrome, occurring in more than half of cases. Stridor and hoarseness also may be associated with laryngeal edema.[10] Other common complaints include chest pain, orthopnea, syncope, and a sensation of head fullness that is exacerbated by bending forward. Hemoptysis, dysphagia, and dysphonia are relatively rare presenting symptoms.[11]

On physical examination, the most common findings of SVC syndrome are dilated veins on the face and neck and overlying the chest wall, which are present in a high percentage of cases.[9] The veins may appear quite suddenly and may be displayed in a remarkable vertical pattern. In contrast to the familiar telangiectases often found on the chests of elderly patients, these veins are typically more widespread, palpably dilated, and often can be traced several centimeters across the upper chest.[5] Swelling of the face, tongue, and upper extremities also are common, and these symptoms typically are exaggerated when the patient is placed in the horizontal position.[7] Other findings include upper extremity edema, facial plethora, proptosis, and conjunctival vascular injection. Cyanosis of the skin due to venous stasis also may occur.[7]

Treatment of SVC syndrome is aimed at relief of venous obstruction.[10] In patients with poorly prognostic malignant etiologies, rapid symptomatic palliation usually is achieved by radiation, chemotherapy, or endovascular stenting. Patients with better prognoses usually are treated surgically via bypass grafting, but percutaneous endovascular stenting is growing increasingly common in this population.[10]

Although SVC syndrome is not a primary dermatologic disorder, the constellation of skin and pulmonary findings are unique and patients may present to the dermatology clinic. Early diagnosis offers the best opportunity for treatment, and 75% of patients with malignancy report substantial improvement of symptoms within days of beginning chemotherapy or radiation therapy.[11]

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