Facial swelling and somnolence in a patient with cancer

A 46-year-old female patient with a history of drug abuse (now on methadone), chronic hepatitis C and heavy smoking was admitted with a 4-month history of increasing low back pain radiating to her left leg, weight loss and sweats. Examination was non-contributory. Laboratory tests showed hemoglobin 12.7 g/dl, white blood cells $18.6 \times 10^3/\mu l$ (neutrophils 14.8), platelets $411 \times 10^3/\mu l$, normal electrolytes, minimal liver enzymes disturbance and Erythrocyte sedimentation rate 86 mm/h. Contrast computed tomography (CT) demonstrated lytic lesions in the left iliac and sacral bones, an adjacent large soft tissue mass with multiple (necrotic) lymph nodes in the retroperitoneum and liver hilum and two 28 to 36 mm liver masses. Multiple pulmonary nodules were found, as well as (necrotic) lymph nodes in the mediastinum, right lung hilum, right axilla and neck. Imaging-guided biopsy confirmed poorly differentiated adenocarcinoma of unknown primary.

While considering therapeutic options, the patient suddenly developed bilateral eyelid edema (right > left), face swelling and arm edema and gradually became increasingly somnolent with venous distention in the neck and superficial chest wall veins (Figure 1). CT now revealed a mass of enlarged hypodense mediastinal lymph nodes compressing the superior vena cava (SVC) (Figure 2). Prophylactic low molecular weight heparin was started.

Figure 1. The patient’s photograph showing eyelid edema, face swelling and venous distention in the neck and superficial chest wall veins. Even her somnolence can be sensed.

Figure 2. Contrast-enhanced CT of the chest (axial and sagittal planes) demonstrating massive mediastinal lymphadenopathy with necrotic hypodense regions compressing the SVC (arrow). A proximal dilatation of the SVC can be seen in the sagittal view.
increased to full dose, a peripherally inserted central catheter line was inserted and two courses of carboplatin and taxol were administered with marked improvement in the SVC obstruction and general wellbeing. However, the patient died suddenly within 2 weeks of her presentation.

Comment

The patient presented with disseminated cancer of unknown primary, a presentation that is not rare, encountered in ~4% of oncology patients.¹ Her most important symptom was pain, until she developed bilateral face swelling, arm edema and somnolence—typical signs of SVC obstruction² that can be recognized at a glance (Figure 1). The SVC syndrome occurs in ~15 000 persons in the USA each year. When not related to indwelling intravascular devices (about a third of the cases), an intrathoracic malignancy (often lung cancer, lymphoma or metastatic cancer) is usually responsible.²,³ SVC syndrome may not infrequently be the presenting symptom of an undiagnosed tumor. Either direct invasion, compression or thrombosis may obstruct the SVC. Contrast-enhanced chest CT (or magnetic resonance venography) is most useful for diagnosis. Severe cases are medical emergencies and require immediate stent placement and radiotherapy according to American College of Chest Physicians guidelines.⁴ Combination chemotherapy is another viable option.

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References