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Case Report

Superior Vena Cava Syndrome in a Patient with Small-Cell Lung Cancer: A Case Report

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Abstract
Superior vena cava (SVC) syndrome, a potential oncologic emergency, is closely associated with malignancy and right-sided lung cancer in particular. A case of SVC syndrome presenting with facial swelling, neck distension, and enlarged veins of the upper chest, which developed over a period of 5 weeks in a 46-year-old patient on a clinical trial with small-cell lung cancer, is reported. Computed tomography scan of the chest revealed slight enlargement of a superior conglomerate mediastinal lymphadenopathy and intramural thrombus of the SVC. The etiology, diagnosis, and treatment of the SVC syndrome are discussed.

Introduction

The superior vena cava (SVC) syndrome, which occurs in approximately 15,000 people in the United States each year [1], refers to characteristic symptoms and signs that result from the increase in venous pressure upstream from the SVC obstacle (Fig. 1), impairment of

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venous return to the heart from the head, neck, thorax, and upper extremities, and the subsequent development of venous shunts to bypass the obstruction [2]. Hence, diagnosis is usually made on the basis of clinical suspicion coupled with confirmatory imaging (plain radiography, computed tomography (CT), and venography).

The intensity of the symptomatology depends (1) on the degree of edema, resulting from the egress of fluid at high pressure into the adjacent tissues, e.g., face, neck, upper extremities, upper thorax, and brain, and (2) the speed of collateral venous development, which may result in distension of the superficial veins in the chest wall, to restore venous return; if the time for collateral vessel development is insufficient, decreased right atrial filling and cardiac output may ensue [3]. It is this potential for the development of cerebral edema with increased intracranial pressure, laryngeal edema with airway obstruction, and hemodynamic compromise, which defines SVC as an oncologic emergency, although death is only very rarely an outcome [4].

Anatomically, as a thin-walled vein that courses along the right middle mediastinum [5] (Fig. 2), the SVC is readily compressed, most commonly by right-sided malignancies and lymphadenopathies and/or a thrombus within the vessel, although iatrogenic causes from intravascular devices (catheters, cardiac defibrillators, and pacemaker wires) are increasingly implicated [6]. Since small-cell tumors are centrally located, with mediastinal adenopathy [7], they account for the majority of cases of malignant SVC syndrome followed by squamous cell carcinoma, large-cell carcinoma, and non-Hodgkin's lymphoma [8, 9].

Treatment options include percutaneous stent placement, corticosteroids, radiotherapy, and chemotherapy as well as thrombolytics, anticoagulation, and elevating the head of the patient's bed [10]; however, randomized clinical trials on which to base "best therapy" recommendations are lacking [11].

Case Presentation

A 46-year-old overweight white male (BMI 31) with refractory small-cell lung cancer (SCLC) metastatic to the brain status post multiple lines of therapy including cisplatin/etoposide, whole brain radiotherapy, single agent gemcitabine, carboplatin/paclitaxel, single agent docetaxel, single agent irinotecan, single agent temozolomide, and single agent vinorelbine enrolled on a clinical trial in which an experimental intravenous anticancer "priming" agent is followed at progression by re-introduction of first-line platinum doublets, in this case cisplatin etoposide. During a regularly scheduled infusion visit, facial plethora, neck swelling, and purplish discoloration across his chest was noted (Fig. 3).

Except for mild dyspnea when supine and a cough, the patient was asymptomatic and in no acute distress. Blood pressure, respirations, and oxygen saturation were all within normal range. Pertinent physical findings were: diffuse edema in the neck and dilated, engorged blood vessels on the chest as seen in the image above. Stridor, edema, shortness of breath, difficulty swallowing, visual disturbances, headache, and altered mental status were not present. Labs revealed a normal CBC and BMP.

Because his symptoms were suggestive of SVC syndrome, common with SCLC, and potentially life-threatening, he was immediately admitted to the hospital. A PET scan identified slight enlargement of a superior mediastinal adenopathy conglomerate (but with lower metabolic activity potentially indicating a favorable response to treatment) likely exerting pressure on the SVC. A CT revealed tumor encasement of the SVC with luminal compression (Fig. 4).

The experimental priming agent was discontinued for suspected progression (even though radiologically his tumor was stable), 18,000 units of Fragmin was administered for anticoagulation of the intraluminal thrombosis, and per protocol cisplatin-etoposide was started without incident to relieve compression/invasion of the SVC by the tumor. He was discharged 2 days later in stable condition. On routine follow-up in the clinic, the patient's neck swelling and facial plethora were improved. Partial resolution of venous distension, after initiation of platinum doublet, was seen (Fig. 5).

Discussion

A rare emergency that occurs in roughly 15,000 persons in the United States each year, SVC syndrome, first described by William Hunter in 1757 in a patient with a large syphilitic aortic aneurysm [12], is most commonly (and almost exclusively) encountered in patients with malignancies (especially SCLC). As a thin-walled vein approximately 7 cm in length and 2 cm in diameter [13] that drains deoxygenated blood from the upper body, the SVC is vulnerable to external compression, thrombosis, or invasion of the vein. Depending on degree of SVC compromise and the extent of collaterality, the clinical presentation is varied, ranging from gradual in onset and mild with facial and upper extremity edema to immediate in onset and dire with intracranial hypertension, hemodynamic instability, and tracheal obstruction. The subject of this case report, who fell into the mild category, evinced only slight orthopnea, intermittent dry cough, facial plethora, and superficial blue vessels on the skin surface venous on the chest wall, which required a high degree of clinical suspicion to make the diagnosis. A chest CT with intravenous contrast confirmed the diagnosis, which was due to external compression by enlarged mediastinal lymph nodes.

Chemotherapy, radiation, or intravascular stents are the mainstays of treatment. In this case, the patient was restarted per clinical trial protocol on cisplatin-etoposide with almost immediate resolution of clinical symptoms.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors have no conflicts of interest to declare.

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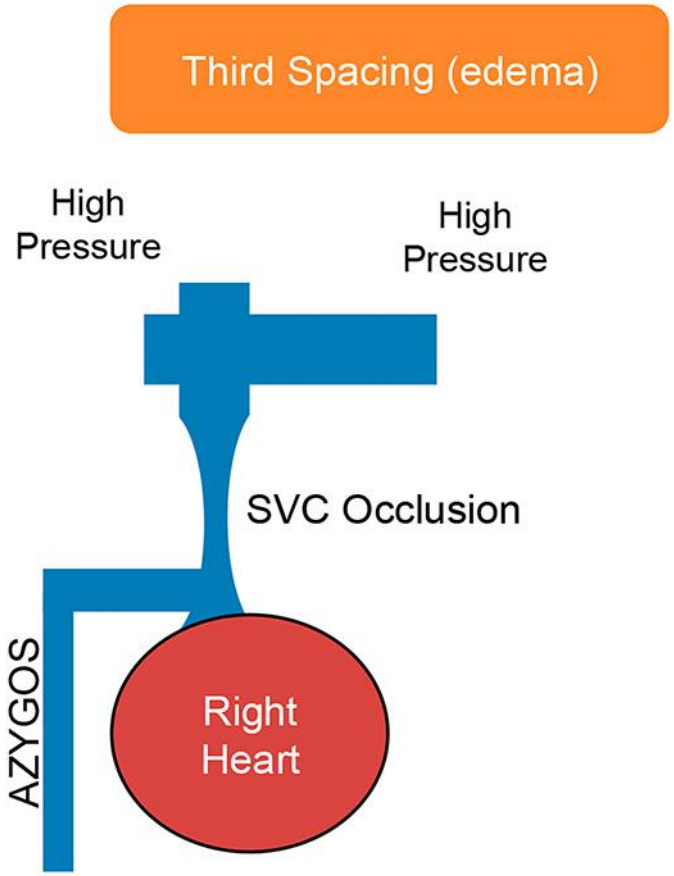

Fig. 1. Superior vena cava (SVC) occlusion causes edema (third spacing) upstream from the site of obstruction.

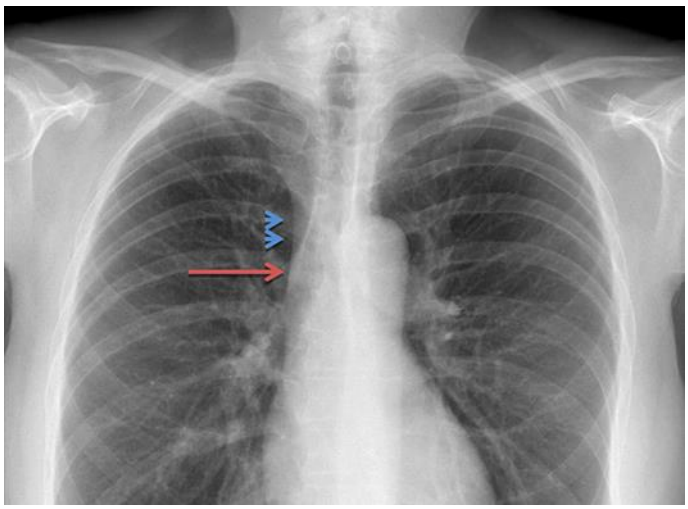

Fig. 2. Frontal chest radiograph with normal superior vena cava border (blue arrowheads) above the azygos vein (red arrow).

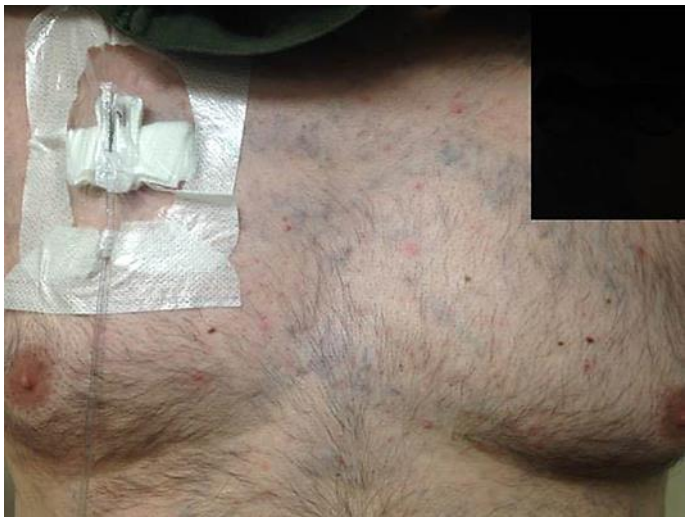

Fig. 3. Collateral venous circulation causing distension of the superficial veins in the chest wall of the patient.

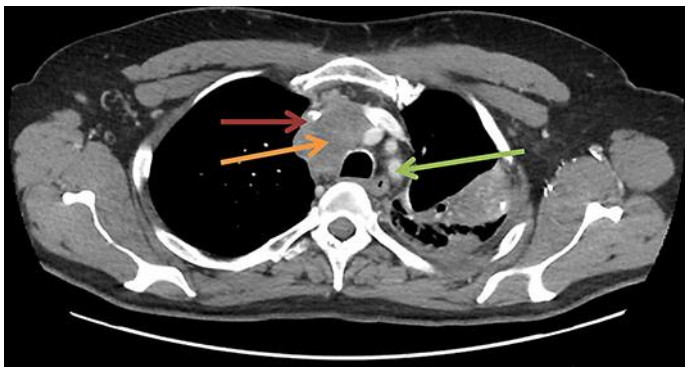

Fig. 4. Axial contrast-enhanced computed tomography scan of upper chest shows encasement and compression of superior vena cava (red arrow) by tumor (orange arrow). Note collateral veins in anterior mediastinum (green arrow).

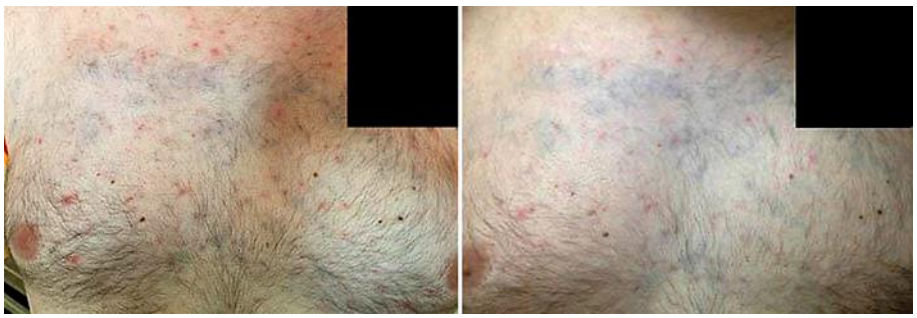

Fig. 5. Collateral venous circulation causing distension of the superficial veins in the chest wall of the patient. Note partial resolution of venous distension after initiation of the platinum doublet.