A 59-year-old woman presents with generalized facial swelling and dyspnea that has progressed gradually over the past month. The patient also reports a sensation of pressure in her neck and ears and swelling of the lower eyelids, neck, upper chest, and upper limbs. The blood vessels on her upper chest are prominent. A dry, irritating cough has worsened.

**History.** The patient has smoked about a pack of cigarettes a day for the past 30 years, drinks alcohol socially, and takes no medications.

**Examination.** This mildly obese woman is in moderate respiratory distress. Temperature is 37°C (98.6°F); heart rate, 77 beats per minute and regular; respiration rate, 18 breaths per minute; blood pressure, 137/86 mm Hg. Pulse oximetry, 93% on room air. Thyroid is not palpable. No breast mass or adenopathy is identified. Chest has normal contours with symmetric expansion. Decreased breath sounds on the right side, with dullness to percussion at the right base. No wheezes or rales. Jugular vein pulse is elevated. Point of maximal impulse is not displaced. Heart sounds are normal; no murmur or gallop. Abdomen has normal contours with no tenderness or organomegaly. Carotids are equal and palpable. Neurologic examination is unremarkable.

**Laboratory studies.** White blood cell (WBC) count, 6600/L, with 68% polymorphonuclear leukocytes, 28% lymphocytes, 4% eosinophils. Hemoglobin, 14.6 g/dL; platelet count, 275,000/L; erythrocyte sedimentation rate, 28 mm/h. Levels of serum sodium, 128 mEq/L; potassium, 4 mEq/L; chloride, 90 mEq/L; carbon dioxide, 26 mEq/L. Blood urea nitrogen level, 3 mg/dL; serum creatinine, 0.3 mg/dL. Thyroid-stimulating hormone, 3.4 IU/mL. Urinalysis is negative for protein, glucose, and WBCs. You order a chest film and CT scan.

**What abnormalities are evident on these images, and what do you suspect is the cause?**

A. Cancer of the lung
B. Thymoma
C. Retrosternal goiter
D. Dermoid cyst
E. Sarcoidosis

**WHAT'S WRONG:** The chest radiograph shows a large right perihilar density with infiltrative changes of the right lower lung. The CT scan shows a large mass in the superior mediastinum, aortal pulmonary window, and right perihilar and subcarinal regions, with encasement of the superior vena cava (SVC), right pulmonary artery, lower trachea, and right main-stem bronchus. The mass almost completely occludes the SVC. A right pleural effusion is also evident. In a patient with a long history of smoking, SVC obstruction, and a superior mediastinum mass extending from the perihilar to the subcarinal area, the likely diagnosis is cancer of the lung. Hospital course. Further test results included carcinoembryonic antigen, 3.2 ng/mL; cancer antigen (CA) 19-9, 30 U/mL; CA 125, 70 units; alpha fetoprotein, 1.7 ng/mL; human chorionic gonadotropin, less than 5 mIU/mL; cocci serology, negative. Assays for antineutrophil cytoplasmic antibody (p-ANCA and c-ANCA), purified protein derivative, rheumatoid arthritis factor, and antinuclear antibody are all negative. Urinalysis: sodium, 32 mEq/L; osmolality, 396 mOsm/L; serum creatinine, 200 mOsm/L; serum albumin, 3.7 g/dL. Total bilirubin, 0.8 mg/dL; alkaline phosphatase, 95 U/L; aspartate aminotransferase, 24 U/L; alanine aminotransferase, 30 U/L. A biopsy specimen of the superior mediastinum mass revealed small-cell neuroendocrine carcinoma. A radiation oncologist was consulted, and the patient began daily radiation treatment. Her facial edema improved markedly, and her respiratory distress diminished. Follow-up chemotherapy was arranged. However, 1 month later, the patient was brought to the emergency department in severe respiratory distress; she subsequently died of cardiopulmonary arrest.

**CAUSES OF SVC SYNDROME** SVC syndrome results from:

- Invasion or external compression of the SVC by contiguous pathologic processes that involve the right lung, lymph nodes, and other mediastinal structures.
- Thrombosis within the SVC.
These mechanisms can coexist. About 75% to 85% of cases of SVC syndrome are caused by malignant tumors. **CLINICAL MANIFESTATIONS** SVC syndrome typically affects elderly persons with a significant smoking history. However, this disorder is sometimes seen in younger patients who have a family history of bronchogenic carcinoma or other lung cancers or who have complications of central venous catheterization. Patients with SVC syndrome usually present with neck and facial swelling, especially around the eyes, as well as increasing dyspnea, orthopnea, and cough. Other symptoms can include head, neck, and ear fullness; dysphagia; hoarseness; hemoptysis; nasal congestion; tongue swelling; headache; dizziness; and syncope. Pain in the upper right side of the chest or interscapular area is not uncommon. The characteristic physical findings reflect the underlying pathologic processes. Obstruction of venous return from the head, neck, and upper extremities results in dilated neck veins; an increased number and prominence of anterior chest wall veins; and edema of the face, arms, and chest. Patients with more advanced disease exhibit proptosis, glossal and laryngeal edema, or altered mental status. The clinical picture is more pronounced if the obstruction is located below the azygos vein. **DIFFERENTIAL DIAGNOSIS** Obstruction of recent onset is likely to be malignant in origin, whereas long-standing obstruction is more likely to be benign. Primary lung cancer and lymphoma account for 94% of cases of SVC syndrome. The risk of SVC syndrome is greatest in small-cell lung cancer because of the tendency of this cancer to develop centrally rather than peripherally in the airways. Most patients with SVC syndrome and lymphoma have non-Hodgkin lymphoma. Other causes of SVC syndrome include thymoma, primary mediastinal germ cell neoplasms, breast cancer, fibrosing mediastinitis following primary infection with *Histoplasma capsulatum*, tuberculosis, actinomycosis, aspergillosis, blastomycosis, and nocardiosis. SVC syndrome may also be associated with sarcoidosis and postradiation fibrosis. **LABORATORY AND IMAGING STUDIES** Tumor markers can help identify a malignant process. However, because up to 60% of patients with malignancy-related SVC syndrome present without a known diagnosis of cancer, tissue biopsy for histologic diagnosis is required. Because malignancy is the most common underlying disorder, most patients with SVC syndrome have an abnormal chest radiograph at presentation. Contrast-enhanced CT defines the level and extent of venous blockage, maps collateral pathways of venous drainage, and often permits identification of the cause of venous obstruction. **TREATMENT** Treatment is directed at control of the underlying disease. Radiation therapy is indicated emergently in patients with SVC syndrome, especially when malignancy is suspected. Subsequently, chemotherapy can be given to patients with cancer. In nonmalignant conditions, such as thrombotic occlusion, thrombolytic therapy with or without percutaneous angioplasty within 5 days of clot formation is useful; intravenous heparin is another option. **PROGNOSIS** The overall prognosis for patients with tumor-associated SVC syndrome is closely linked to tumor histology and stage at presentation.

**References:** FOR MORE INFORMATION:


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