



RESEARCH ARTICLE

SUPERIOR VENA CAVA SYNDROME SECONDARY TO ADULT T-CELL LYMPHOBLASTIC LYMPHOMA

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Abstract

Superior vena cave (SVC) obstruction can be a substantial contributor to morbidity and mortality in cancer and non-cancer patients. It is currently known to be almost exclusively secondary to malignancy, most frequently lung cancer. A case of SVC syndrome presenting with worsening right-sided chest pain, shortness of breath, facial swelling, neck distension, and cough developing over a period of 12 days is reported. The approach to diagnosis included imaging studies and tissue diagnosis. CT scan of the chest revealed a large right pericardial mass, hilar infrahilar adenopathy, moderate increased right base posterior effusion, and compressive atelectasis. A mediastinal lymph node CT guided core biopsy subsequently revealed adult T-cell lymphoblastic lymphoma. Due to rapid symptom progression, the patient was started on hyper-CVAD regimen and tumor lysis prophylaxis. The etiology, diagnosis, and treatment modalities of the SVC syndrome are discussed further in detail. Given the wide range of symptom severity, identifying and treating severe SVC syndrome promptly can be critical to patient outcomes.

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Introduction:-

The patient described in this case report has been diagnosed with adult T-cell lymphoblastic lymphoma leading to superior vena cava (SVC) syndrome, an unusual clinical presentation that warrants further development of efficacious and safe treatments for improving patient outcomes.

Purpose

A 31-year-old Latino male presented with sharp, 9/10 right-sided chest and upper quadrant pain radiating to his back, aggravated with movement. History revealed non-productive cough, fatigue, subjective fevers, and night sweats. Patient had decreased right lower breath sounds. Computed tomography (CT) angiography with contrast revealed moderate effusion with perihilar compressive atelectasis and multiple diffuse lymphatic adenopathy. Thoracentesis revealed exudative effusion with 81% lymphocytes. A CT-guided mediastinal lymph node core biopsy showed adult T-cell lymphoblastic lymphoma, staining positive for CD3, BCL2, CD1a, TDT, and CD10.

Repeat CT angiography revealed left displacement of mediastinal structures, severely narrowing the SVC to 7mm. The patient was started on hyper-CVAD regimen (Cyclophosphamide 300 mg/m², Mesna 600 mg/m², Vincristine 1.4 mg/m², Doxorubicin/Adriamycin 50 mg/m², Dexamethasone 40 mg). During bone marrow biopsy, SVC syndrome was noted. While prone, his right upper extremity and face swelled, followed by dyspnea and cough. He was repositioned; however, symptoms reemerged with a positive Pemberton sign.

Discussion:-

SVC syndrome, arising from any condition that blocks blood flow through the SVC, can be a substantial contributor to mortality. In >90% of cases, SVC syndrome is caused by malignancy (e.g. T-cell lymphoblastic lymphoma). Obstruction can occur via direct tumor invasion or external compression of the vessel. Moreover, disease rate varies depending on etiology; our patient developed symptoms within 12 days, favoring malignancy. Upon getting a bone marrow biopsy, our patient showed the classic findings of SVC syndrome.

Once the patient was stable, a work-up was done for malignancy-associated SVC syndrome. This approach was preferred as histological diagnosis may be obscured by pre-biopsy radiation. Nearly all cases of SVC syndrome are diagnosed clinically, but certain diagnostic tests and procedures are helpful. Magnetic resonance imaging (MRI) and CT scans provide information regarding possible etiologies and, therefore, direct the approach to a tissue diagnosis. Tissue evaluation with immunohistochemistry remains the mainstay in diagnosis of lymphomas. Our patient's immunohistochemistry was positive for tumor markers discovered in only a subset of tumors, directing towards treatment with combination chemotherapy instead of local measures such as radiation therapy or percutaneous vascular procedures.

Therapeutic maneuvers for clinically stable patients such as head elevation can reduce hydrostatic pressure and relieve edema. For symptomatic patients, radiation therapy and intravascular stent placement are recommended. Nonetheless, initial management should be led by the severity of symptoms and the underlying malignancy. The primary therapy for T-cell lymphoblastic lymphoma is hyper-CVAD chemotherapy, which our patient received.

Conclusion:-

While the severity of symptoms differs widely, SVC syndrome may be fatal. Reported is a rare case of SVC syndrome caused by adult T-cell lymphoblastic lymphoma. Given the sporadic presentation and aggressive nature of this lymphoma, early recognition and identifying when intervention is necessary are critical factors for positive patient outcome.

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