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Case Report

Superior vena cava syndrome: a case report of a rare form of presentation

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ABSTRACT

Superior vena cava syndrome (SVCS) is the clinical manifestation of superior vena cava (SVC) obstruction, with severe reduction in venous return from the head, neck and upper extremities. It is a medical emergency. Obstruction to the SVC may be caused by neoplastic invasion of the venous wall associated with intravascular thrombosis or, more simply, by extrinsic pressure of a tumour mass against the relatively thin-walled SVC. More than 80% cases of SVCS are caused by malignant mediastinal tumours. We present a 42-year-old female patient who was admitted with 15 days history of swelling of face, neck and upper limbs and dyspnea of 5 days duration at MKCG Medical College Hospital, Berhampur, Odisha, India. The patient was diagnosed as a case of Non-Hodgkin's Lymphoma (NHL) which is a rare form of presentation. In this case report we would like to highlight that although lung carcinoma being the most common cause of acute superior vena cava syndrome, other malignancies like NHL should also be kept in mind and the importance of a detailed history, clinical examination and thorough investigations cannot be over emphasised.

Keywords: Contrast enhanced computed tomography, Fine needle aspiration cytology, Non-Hodgkin's lymphoma, Superior vena cava syndrome

INTRODUCTION

Superior vena cava syndrome (SVCS) is the clinical manifestation of superior vena cava (SVC) obstruction, with severe reduction in venous return from the head, neck and upper extremities.[1]

The obstruction of blood flow through SVC can occur through invasion, external compression or thrombosis in the setting of a malignancy and the severity and rapidity of symptom onset depends on the rate at which the SVC is occluded and if there has been enough time for collateral venous drainage to develop. Lung cancer, lymphomas, thymic neoplasms, germ cell tumours, mesotheliomas and other solid tumours are associated with SVC syndrome.[2]

It is a medical emergency and most often manifests in patients with a malignant disease process within the thorax. It is devastating in nature and causes a great concern to the patient. Most cases of SVC syndrome are diagnosed by clinical examination but other investigations like chest X-ray, CT scan, MRI, ultrasonography and venography can be helpful in determining etiology and further management.

Correlation of imaging studies with clinical findings suggests that severity depends on the level of obstruction, that is, above or below the level of azygous vein.[3] Early diagnosis and treatment with diuretics and oxygen can produce temporary symptom relief and glucocorticoids may be useful in shrinking lymphoma masses but of no benefit in lung cancer. Radiation therapy is the treatment

of choice for SVCS caused by non-small cell lung cancer and chemotherapy for small cell carcinoma of lung. According to clinical observations, approximately 10% patients with bronchogenic carcinoma and 45% patients with lymphoma treated with irradiation can survive for atleast 30 months, whereas patients with untreated malignant SVC syndrome survive only for 30 days.[4] Percutaneous stent placement in malignant SVC syndrome is an effective technique to relieve symptoms.[5] SVC syndrome due to thrombus formation should be treated by anticoagulation. Prognosis in patients with SVC syndrome depends on the underlying condition. In benign cases of SVC syndrome, life expectancy is normal, but if SVC syndrome is secondary to malignancy, the prognosis depends on tumour histology.

Complications of SVC syndrome include laryngeal edema, cerebral edema (characterised by headache, confusion and coma), low cardiac output leading to hypotension and pulmonary embolism. Of these, laryngeal edema and cerebral edema can cause sudden death. We describe here a case of rare form of presentation of superior vena cava syndrome (SVCS).

CASE REPORT

A 42-year-old female patient was admitted at MKCG Medical College Hospital, Berhampur, Odisha, India with 15 days history of swelling of face, neck and upper limbs and dyspnea of 5 days duration and there was no history of fever or decreased urination (Figure 1).

Figure 1: A 42-year-old female presenting with swelling of face, neck and upper limbs and dyspnoea of 5 days duration.

On admission, her pulse rate was 100/min and BP were 100/60mm Hg and SpO2 was only 82%. Head to foot examination revealed the presence of multiple cervical and bilateral axillary nodes, firm and discrete. Examination of all systems were normal. A provisional diagnosis of Superior Vena Cava Syndrome was made and she was started on oxygen in propped up position and Tab Dexamethasone 8 mg TDS.

On further review of her past history, there was a history of multiple cervical lymph node enlargement 8 months ago which was aspirated by fine needle (FNAC) and revealed necrotising lymphadenitis. A TB-PCR done was found to be negative, nevertheless, she was started on anti-tubercular treatment (ATT). A biopsy of the enlarged right submandibular node was done but, the report of which was not available. A CT scan of neck showed bulging of nasopharynx on the right side (Figure 2). A punch biopsy ruled out malignant change according to the reports with the patient. Following then she was asymptomatic until her present visit to us 8 months later.

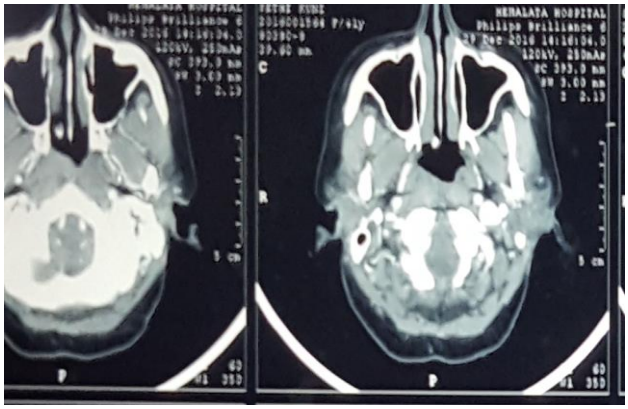

Figure 2: CT scan of neck showing bulging of nasopharynx on the right side.

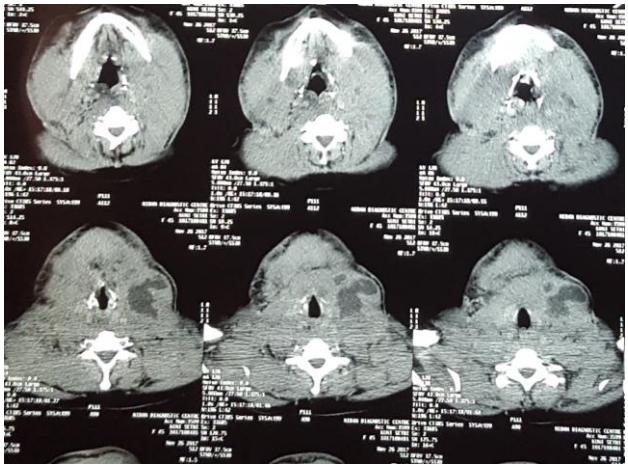

Figure 3: CT scan of neck showing soft tissue over lower part of neck with mass effect over neck and mediastinal vessels.

Among her initial blood investigations, her haemoglobin was 9.6g%, TLC 34,810/mm^3, DC-$N_{96}L_3M_1E_0B_0$, ESR 12, sodium 128, potassium 3.5. Urine routine showed 8-10 pus cells.

Patient improved on the following day and on day 3 her SpO_2 was 96% in room air.

During patient's present admission, a CECT thorax was done on day 3 which showed large necrotic lymph node

at lower neck region. There was moderate edema involving lower neck soft tissue extending to anterosuperior mediastinum associated with mass effect over the neck and mediastinal vessels (Figure 3). Large bilateral axillary lymph nodes and moderate right pleural effusion was present (Figure 4).

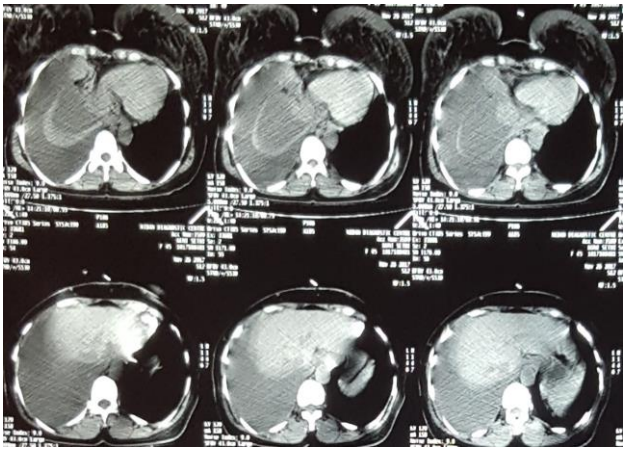

Figure 4: CT scan of thorax showing moderate right pleural effusion.

A pleural fluid study was done on the patient and the cell count was 600/mm^3 with lymphocytes 70%, mesothelial cells 25% and atypical lymphoid cells 5%. There were suspicious atypical moderate sized cells with bluish vacuolated cytoplasm. A malignant cell block was prepared from pleural fluid aspirate which showed the presence of abnormal lymphoid cells in a monotonous pattern on a hemorrhagic background with cells showing scanty to moderate cytoplasm and coarsely clumped chromatin with a few cells showing intracytoplasmic vacuoles, suggestive of effusion secondary to lymphoma. FNAC was done from neck, axillary and submandibular nodes which showed atypical lymphoid cells on a background of tumoral necrosis suggestive of Non-Hodgkin's Lymphoma (NHL). A bone marrow study done was normal.

Hence, authors arrived at a final diagnosis of Non-Hodgkin's Lymphoma (NHL) and she was referred to another tertiary centre for further management as we did not have a department for oncology. The patient was then lost to follow up.

DISCUSSION

SVCS was first described by William Hunter in 1757 in a patient with syphilitic aortic aneurysm.[6] In 1954, Schechter reviewed 274 well-documented cases of SVCS reported in the literature; 40% of them were due to syphilitic aneurysms or tuberculous mediastinitis.[7] The most common cause of this condition is lung cancer in 75% cases.

SVC is the major drainage vessel for venous blood from the head, neck, upper extremities, and upper thorax. Located in the middle mediastinum, it is surrounded by sternum, trachea, right bronchus, aorta, pulmonary artery, and the perihilar and paratracheal lymph nodes. It extends from the junction of the right and left innominate veins to the right atrium, a distance of 6-8cm. It is a thin-walled, low-pressure, vascular structure. This wall is easily compressed as it traverses the right side of the mediastinum.[8]

Obstruction of the SVC may be caused by neoplastic invasion of the venous wall associated with intravascular thrombosis or, more simply, by extrinsic pressure of a tumour mass against the relatively thin-walled SVC.

More than 80% of cases of SVCS are caused by malignant mediastinal tumors.[9-11] Bronchogenic carcinomas account for 75-80% of all these cases, with majority being small-cell carcinomas.[12] Non-Hodgkin's Lymphoma (especially the large-cell type) account for 10-15%. Causes of SVCS appear similar to the relative incidence of primary lung and mediastinal tumours. Rare malignant diseases include Hodgkin's disease, metastatic cancers, primary leiomyosarcomas of the mediastinal vessels, and plasmocytomas.[13-16]

Non-malignant conditions that cause SVCS include mediastinal fibrosis, aortic aneurysm, vasculitis, and arteriovenous fistulas; infections like histoplasmosis, tuberculosis, syphilis, and actinomycosis; benign mediastinal tumours like teratoma, cystic hygroma, thymoma, and dermoid cyst, Cardiac causes, such as pericarditis and atrial myxoma; thrombosis related to the presence of central vein catheters. These account for approximately 22% of cases of SVCS.[17-20]

NHL can present as SVC syndrome in 10% cases. This patient had no other prior symptoms like fatigue, fever or weight loss or night sweats. Her only complaint was the presence of some cervical nodes 8 months ago and she was put on anti-tubercular treatment (ATT) and then she went on to develop acute swelling of face and neck with pleural effusion. This is a rare form of presentation.

CONCLUSION

Lung carcinoma being the most common cause for acute superior vena cava syndrome which can be associated with pleural effusion, the possibility of other malignancies like NHL should also be kept in mind and the importance of a detailed history, clinical examination and thorough investigations cannot be over emphasised.

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