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Case Report

Superior vena cava syndrome management in limited source setting: a case report

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ABSTRACT

Superior vena cava syndrome (SVCS) is syndrome caused by mass compression, tumor invasion, and/or thrombosis of SVC. In the past, SVCS was mostly linked to infection. Nowadays, SVCS is mostly linked to malignant tumor and medical procedures. Most common malignant cause of SVCS is non small cell lung cancer. A 69-year-old man was presented with breathing difficulty. Symptom began 4 months before admission, with worsening of symptom since 2 weeks before admission. Symptom improved with sitting position, and worsened with supine position. Patient had been sleeping with 2 pillows. Patient had productive cough and hoarseness. Swelling of face and neck were present. Collateral vein distention was visible in the area of head, neck, and chest. Physical examination of lung revealed decreased vesicular breath sound at right side. Non pitting edema was found at both sides of upper extremity. SVCS in this case can be categorized into grade 2, chronic SVCS case. CT showed center right lung mass, although malignant cells were not obtained from pleural puncture and CT guided needle biopsy. Staging of mass was T4N3M1a. Mass finding at the center of right lung is in accordance with the previous findings that SVCS generally arise from lung (not mediastinum), and that right sided masses are generally more likely to cause SVCS. Management in this case was done by giving corticosteroid and diuretic. Surgery, radiotherapy, and immunotherapy were not done since malignancy diagnosis hadn't been able to be concluded.

Keywords: Superior vena cava syndrome, Malignancy, Management

INTRODUCTION

Superior vena cava syndrome (SVCS) is syndrome caused by mass compression, tumor invasion, and/or thrombosis of SVC. Clinical spectrum of SVCS ranges from asymptomatic cases to life threatening emergencies.[1] In the past, SVCS was mostly linked to infection; SVC was compressed by syphilitic aortic aneurysm or tuberculosis mediastinal lymphadenopathy. Nowadays, SVCS is mostly linked to malignant tumors and medical procedures. Compression or invasion of SVC by malignant tumor is responsible for 60% of cases; the rest are caused by thrombosis or stenosis from medical procedures or medical devices (central lines, dialysis access catheter).[2]

Spontaneous VCS thrombosis in younger patients is often associated with major thrombophilia or Behcet's disease, whereas in geriatric patients malignancy is most likely the cause.[3] SVC thrombosis is associated with pulmonary embolism, which worsens hemodynamic status and compromises gas exchange. SVC thrombosis may end up in "catastrophic SVCS".[1] Most common lung malignancy which causes SVCS is non small cell lung cancer, followed by small lung cancer. Other tumors associated with SVCS are non Hodgkin's lymphoma, germ cell tumors, adenocarcinomas, sarcomas, metastatic tumors, and local tumor formations (thymomas, thyroid masses, and esophageal carcinomas).[2,4] Common and rare causes of SVCS can be seen below.

SVCS diagnosis is based on typical clinical symptoms and signs. SVCS symptoms and signs can be categorized into neurological, laryngopharyngeal, facial, and chest wall/ upper extremity.

Symptoms typically increase in supine position. Many patients can't tolerate supine position without elevating the torso and/or head. Right sided masses are generally more likely to cause SVCS due to anatomical reasons.[4] Cardiac output may be reduced, especially in rapidly developing cases like in acute SVC thrombosis, in pre existing chronic heart failure cases, and in cases with concomitant heart and/or pulmonary artery mass compression.[5] Symptoms resulting from reduced cardiac filling may present, for example hypotension and syncope; which usually occurs during coughing or bending.[1]

Venous pressure increases in brachial, cervical, and cerebral veins lead to venous congestion of the head, neck, upper thorax, and arms. Other symptoms include plethora, respiratory distress due to laryngeal and tracheal edema, visual disturbances, impaired consciousness, and neurological abnormalities.[4]

Symptom severity is important in determining the urgency of intervention. For this purpose, a grading system was proposed to allow differentiation between life threatening and non life threatening symptoms.[1]

Table 1: Common and rare causes of SVCS.[1]

Type of cause	Malignant SVCS	Non malignant SVCS
Common causes	Non small lung cancer (NSCLC); small cell lung cancer (SCLC); non Hodgkin lymphoma (NHL); mediastinal metastatic disease	Catheter, port, or cardiac device associated thrombosis; SVC obstruction associated with upper extremity hemodialysis access; thrombophilia
Rare causes	Thymoma and other thymic neoplasms; germ cell neoplasms; mesothelioma; esophagus carcinoma; thyroid malignoma	Morbus Behcet; infectious related mediastinal granumola and fibrosing mediastinitis; aortic aneurysms

Table 2: Symptoms of SVCS.[1]

Neurological	Laryngopharyngeal	Facial	Chest wall/ upper extremities
Headache; blurred vision; confusion; decreased level of consciousness	Coughing; mucosa and tongue swelling and proptosis; stridor; dyspnoe, orthopnoe	Nasal stuffness; conjuctival and periorbital edema; facial edema; plethora, cyanosis	Neck and chest wall swelling; upper extremity swelling; distended jugular vena; neck and chest wall collateral

Table 3: Grading system of SVCS.[6]

Grade	Category	Estimated incidence (%)	Definition
0	Asymptomatic	10	Absence
1	Mild	25	Edema and vein distention in the head and neck, plethora, cyanosis
2	Moderate	50	Edema and vein distention in the head and neck, coughing, mild to moderate impairment of head, jaw or eye lid movement, eye edema with visual distubances
3	Severe	10	Mild to moderate cerebral edema causing headache and dizziness or/and mild to moderate laryngeal edema and diminished cardiac reserve with syncope after coughing or bending
4	Life threatening	5	Significant cerebral edema with confusion or apathy and/or significant laryngeal edema and/or significant hemodynamic compromise (syncope without provocation, hypotension, renal impairment)
5	Fatal	<1	Death

It can be seen from this grading system that acute fatalities in SVCS cases are rare because patients usually are in stable condition. However, it should be taken into account that patients' condition may decline rapidly in cases of acute thrombosis and/or aggressive tumor growth.[1] SVCS is sometimes confused with allergic reactions. Other differential diagnosis include right heart failure, tricuspid stenosis, tricuspid incompetence, constrictive pericarditis, pericardial effusion, mediastinal emphysema, and Quincke edema.[4] A considerable number of causes (especially in tumor cases) can be found on plain X rays.[7] X rays may reveal masses and pleural effusions suggestive of SVCS.

X rays may also reveal venous catheter and cardiac devices. However, X rays can neither rule out or determine SVCS.

Method of choice for imaging SVCS is contrast enhaced CT with multiple phase imaging. It has high sensitivity, high specificity, and reliably defines underlying pathology. Contrast enhanced CT also aids in detection of metastasis and planning of intervention procedures. Contrast enhanced MRI can be used as an alternative in case of iodine contrast media intolerance.[1]

CASE REPORT

A 69-year-old man was presented with breathing difficuly. Symptom began 4 months before admission, with worsening of symptom since 2 weeks before admission. Symptom improved with sitting position, and worsened with supine position. Patient had been sleeping with 2 pillows. Productive cough and hoarseness were present. He also experienced nausea. He lost weight more than 10 kilograms in 6 months. Classic diabetes symptoms (polyuria, polidipsia, and polyphagia) were present. History of diabetes medication was denied. Patient had hypertension, but was poorly controlled.

Patient appeared breathless during admission. Initial vital signs revealed blood pressure 160/80 mmHg, heart rate 80 beats/ minute, respiratory rate 24 breaths/ minute, body temperature 38ºC, and oxygen saturation of 98% with 9 litre oxygen per minute (administered using non rebreather mask).

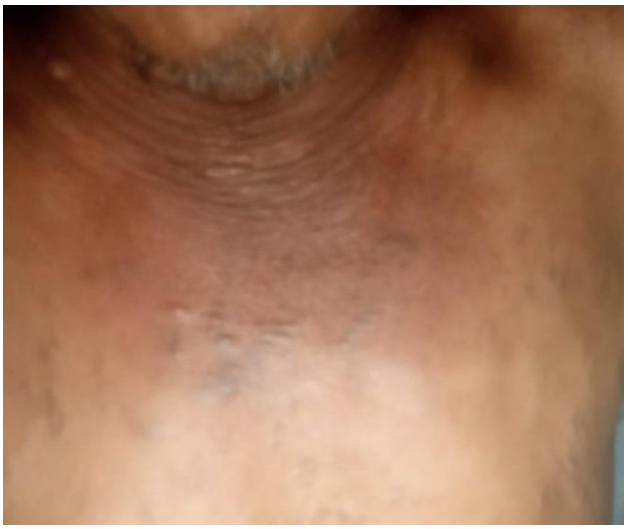

Figure 1: Collateral vein distention in upper chest.

Both conjuctiva appeared normal. Sclera wasn't icteric. Swelling of face and neck were present, including eye edema. Collateral vein distention was visible in the area of head, neck, and chest. There wasn't lymph node enlargement at facial, coli, supraclavicular, and axillary areas. Physical examination of lung revealed decreased vesicular breath sound at right side, with normal vesicular sound at left side. No additional breath sounds, such as stridor or wheezing, were found. Physical examination of heart and abdomen was normal. Non pitting edema of both sides of upper extremity was present. Edema of lower extremity was absent. Initial laboratory test showed hyperglycemia (326 mg/dl). HbA1c was high (7.1%).

Chest X ray showed homogenous opacities on right hemithorax covering right sinus, right diaphragm, and right border of heart suggesting massive right pleural effusion. Effusion size was difficult to estimate. Aorta was dilated and calcified.

Chest X ray result prompts pleural puncture. 70 ml of reddish yellow fluid was extracted. Laboratory analysis of pleural fluid showed positive rivalta test, high glucose, high protein, and low albumin content. Pleural cell count was monoctye dominant. Anatomical pathology analysis of pleural fluid showed neither specific process nor malignancy.

Patient then was given intravenous beta lactam antibiotic, intravenous diuretic, intravenous corticosteroid, intravenous proton pump inhibitor, nebulized bronchodilator, oral mucolytic, oral calcium channel blocker, oral angiotensin receptor blocker, and subcutaneous insulin. Patient was initially treated with 9 litre oxygen per minute (administered using non rebreather mask), but then was titrated down in the next days. Oxygen saturation never fell below 97% after down titration.

Patient management was continued with contrast enhanced CT. CT result showed center right lung mass and right hilar mass, with enlargement of hilar and mediastinal lymph nodes; right pleural effusion. Staging of mass was T4N3M1a.

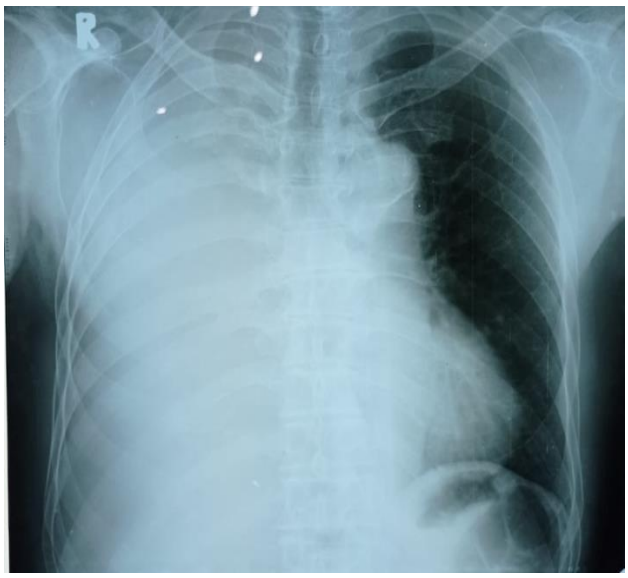

Figure 2: Chest X ray showing homogenous opacities on right hemothorax covering right sinus, right diaphragm, and right border of heart.

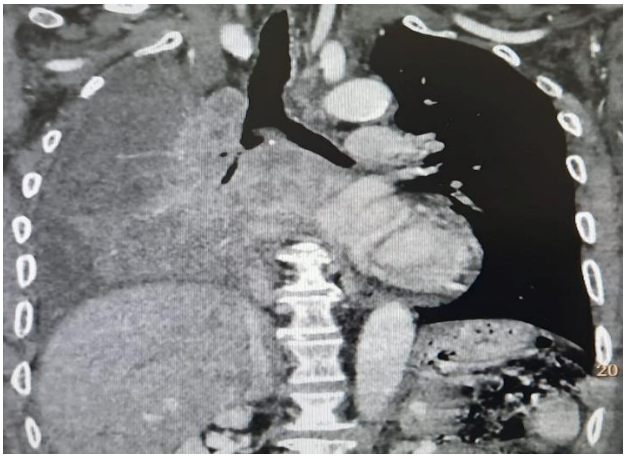

Figure 3: Contrast enhanced computed tomography showing mass with center right lung mass.

Trans thoracal biopsy (TTB) with CT guiding was performed to obtain malignant cells, but anatomical pathology analysis showed neither specific process nor malignancy. Patient was referred to central hospital afterwards for further diagnosis and therapy.

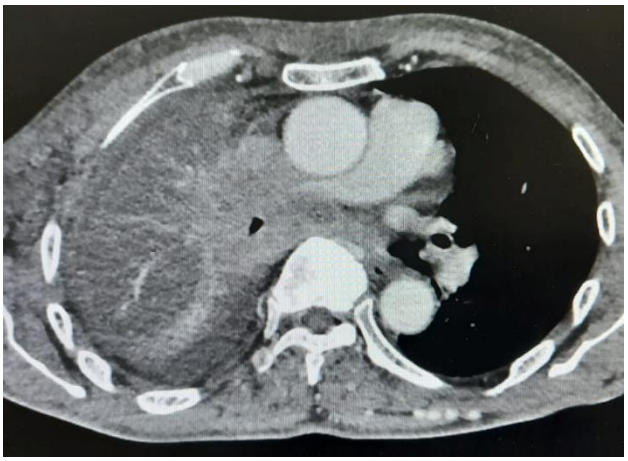

Figure 4: Axial view of mass causing SVCS in this case.

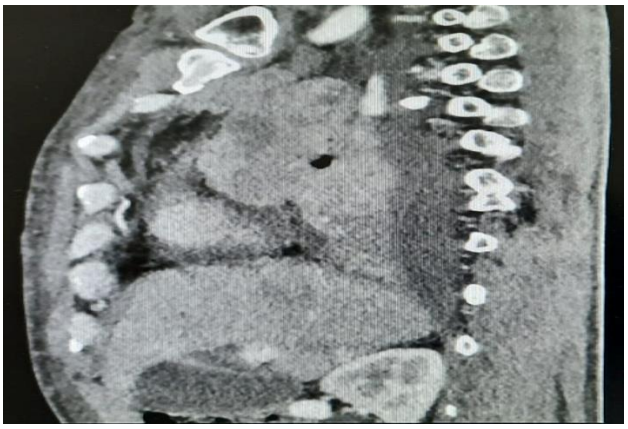

Figure 5: Sagital view of mass causing SVCS in this case.

DISCUSSION

Symptoms and signs of SVCS in this case were clear. Categorization of SVCS symptoms can be seen in Table 2. From laryngopharyngeal category, patient had dyspnoe and orthopnoe. Patient had been sleeping with 2 pillows. Symptoms improved by sitting, and worsened by lying down. Patient also had productive cough and hoarseness. Hoarseness is caused by edema of larynx.[8] Dysphagia also may present as a result from larynx swelling, but was absent in this case.[8] Chronic SVCS may present as distention of collateral veins, which may be observed in upper chest.[9] Collateral vein distention in upper chest was present in this case, hence this case is a chronic SVCS case. This is consistent with patient's history. Symptom began 4 months before admission, with worsening of symptom since 2 weeks before admission. Stridor was absent, but decreased breath sound was present on right side of chest. This is consistent with previous findings that right sided masses are generally more likely to cause SVCS. Symptoms and signs from neurological category were absent. Patient didn't experience headache, blurred vision, confusion, and decreased level of consciousness. Upper extremity, neck, and face swelling were apparent. Eye edema was present, but conjuctival edema wasn't. According to SVCS grading system, case in this report can be categorized into moderate category (grade 2).

Several procedures were conducted since malignancy was suspected. Contrast enhanced CT showed center right lung mass and right hilar mass, with enlargement of hilar and mediastinal lymph nodes. Mass finding at the center of right lung is in accordance with the previous findings that SVCS generally arise from lung, and not mediastinum. Most common malignant cause of SVCS is non small cell lung cancer (~50%), followed by small cell lung cancer (~25%).[10] However, malignancy may also arise from mediastinum.[11] Pleural puncture with fluid cytology and CT guided needle biopsy of intrathoracic mass were done in this case, although neither confirmed presence of malignant cells. Other modalities which may be done to confirm malignancy include sputum cytology and CT guided needle biopsy of lymph nodes. These other modalities were not performed in this case.

Mass staging was T4N3M1a. Mass obliterated SVC and infiltrated right mediastinum, hence this tumor can be staged into T4 tumor. T4 tumor is defined as tumor with >7 cm in greatest dimension or any tumor with invasion of mediastinum, diaphragm, heart, great vessels, recurrent laryngeal nerve, carina, trachea, oesophagus, spine, or separate tumor in different lobe of ipsilateral lung. There was contralateral lymph nodes involvement, hence N3 tumor. N3 is defined as contralateral mediastinal or hilar; ipsilateral/contralateral scalene/supraclavicular. Staging for metastasis was M1a since there were contralateral pulmonary nodules. M1a is defined as tumor in contralateral lung or pleural/pericardial nodule/malignant effusion.

Thrombosis and stenosis were not suspected as cause of SVCS in this case because the symptoms and signs don't suggest those conditions. Patient didn't have history of central line insertion, dialysis access cathether, or medical devices (including cardiac ones) use. Duplex ultrasound of upper extremities is useful in identifying thrombus in jugular, subclavian, and axillary veins. This procedure is also useful in identifying upper extremity access site for venography and endovascular intervention. Digital subtraction venography is the gold standard is the gold standard for SVCS evaluation.

Venography also enables identification of collateral venous pathways and assessment of obstruction severity. This further enables assessment of hemodynamic significance of SVC blockage. Limitation of invasive venography is inability to evaluate extrinsic SVC compression causes.[10] Bone marrow needle biopsy and aspiration may be performed to patients with suspected lymphoma. Invasive procedures may be indicated when diagnosis can't be established with minimal invasive procedures. These invasive procedures include bronchoscopy, mediastinocopy, video assisted thoracoscopy, and thoracotomy.

Management of SVCS in this case includes corticosteroid (dexamethasone) and diuretic (furosemide). Corticosteroid was given to decrease swelling and dyspnea.[9] Corticosteroid may also reduce tumor load. Administration of corticosteroid relieves respiratory distress and reverses swellings on the face within 48 hours.[12] High dose corticosteroid may also reduce inflammation.[13] Another indication for corticosteroid administration includes edema formation prevention, especially in the setting of radiotherapy.[14] Another management of SVCS include seated position, or at least head elevation. Goal of this manuever is to reduce hydrostatic pressure in upper half of body.[9]

Diuretic was given to normalize blood pressure, and to decrease swelling and dyspnea. Diuretic use for SVCS management is still a matter of debate. Diuretic may be recommended in SVCS case, although it is unclear whether changes in right atrial pressure affect venous pressure distal to the obstruction. Diuretic is commonly well tolerated and has good safety profile; hence diuretic use in SVCS case is justified. On the other, diuretic should be stopped if its administration doesn't relieve symptoms.[14]

Most common lung malignancy which causes SVCS is non small cell lung cancer, followed by small lung cancer.[10] Surgery is standard treatment for patients with stage I non small cell lung cancer.[15] Non small cell lung cancer of stage I to IIIA is considered resectable, and non small cell lung cancer of stage IIIB is potentially resectable.[16] Other treatment modalities for non small lung cancer include radiotherapy and immunotherapy. As many as 77% of all lung cancer patients have indication for radiotherapy, although it is often underutilized. Radiotherapy can be used as curative or palliative modalities across all disease stages.[17] Other possible treatment choice includes immunotherapy. In patients with resectable NSCLC stage, immunotherapy can be considered as safe and feasible modality. Immunotherapy may improve pathological response rates with acceptable toxicity.[16] These modalities hadn't been given in this case since malignancy diagnosis hadn't been able to be concluded.

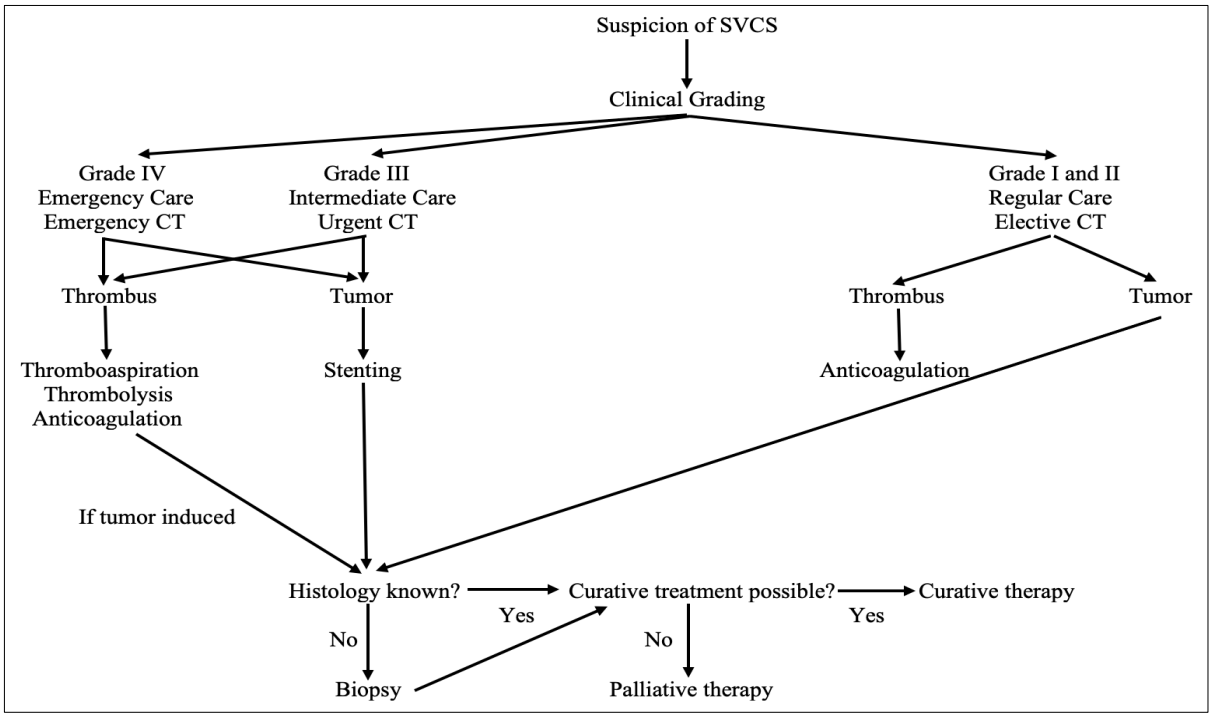

Figure 6: SVCS management algorithm.[1]

Algorithm for SVCS management can be seen in Figure 6. SVCS management is determined by etiology (tumor, thrombus, others), onset, symptoms severity, and availability of therapeutic modalities. SVCS grading can be seen in Table 3. Most patients present with relatively stable condition requiring regular care, although in rare cases patients may require emergency care.

Grade 4 SVCS indicates life threatening condition which is characterized by confusion and obtundation indicating cerebral edema, and/or stridor indicating respiratory distress (due to laryngeal edema), and/or syncope indicating hemodynamic compromise, and/or hypotension. Grade 4 patients require emergency contrast enhanced CT and emergency care. Emergency care required depends on SVCS cause. Tumor compression prompts stent placement, whereas thrombosis prompts thromboaspiration and/or thrombolytic therapy.

Grade 3 SVCS is characterized by headache, dizziness, mild to moderate laryngeal edema, and diminished cardiac output with syncope after coughing or bending (but otherwise stable hemodynamics). Grade 3 patients should be monitored closely in intermediate care unit . These patients require urgent contrast enhanced CT for further assessment. In thrombosis cases, grade 3 patients are given anticoagulant. Thrombosis and/or thrombolytic therapy may be considered in these patients. In tumor cases, histologic diagnosis is essential to determine optimal treatment (although this suggests some time delay). Stent therapy in tumor cases provide faster relief of symptoms compared to radiotherapy and chemotherapy. Moreover, stent therapy doesn't interfere biopsy process, hence stent therapy is the preferred option for grade 3 patients with unknwon histologic diagnosis. On the other hand, grade 3 patients with known radiotherapy or chemotherapy sensitive tumor (such as lymphoma) are treated with such therapies. In grade 1 and 2 SVCS, patients are usually stable. This allows time for elective diagnosis and interdisciplinary treatment planning. Management are not different from grade 3 and 4 SVCS therapy modalities. What differs is the urgency of management; therapy should be delivered faster in grade 3 and 4 SVCS cases.

As catheter and device associated thrombosis begins to predominate as non malignant SVCS cause, preventive measures should be taken. This includes more strict indication for central line and catheter insertion. Ports should be removed after succesful tumor treatment, and not left for years. Devices that are not actively used should be removed after first thrombotic event.

CONCLUSION

We presented a case report of a patient with SVCS. Symptoms and signs of SVCS in this case were clear. SVCS in this case can be categorized into grade 2, chronic SVCS case. CT showed center right lung mass, although malignant cells were not obtained from pleural puncture and CT guided needle biopsy. Staging of mass was T4N3M1a. Mass finding at the center of right lung is in accordance with the previous findings that SVCS generally arise from lung (not mediastinum), and that right sided masses are generally more likely to cause SVCS. Management in this case was done by giving corticosteroid and diuretic. These were given to decrease swelling and dyspnea. Surgery, radiotherapy, and immunotherapy were not done since malignancy diagnosis hadn't been able to be concluded.

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