



Superior Vena Caval Thrombosis in Haemodialysis Patient

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ABSTRACT

Obstruction of blood flow in the superior vena cava (SVC) results in symptoms and signs of SVC syndrome. SVC obstruction can be caused either by invasion or external compression of the SVC by contagious pathologic processes involving the right lung, lymph nodes, and other mediastinal structures, or by thrombosis of blood within the SVC. Occasionally, both mechanisms co-exist. We hereby report a case of a 28 year old Saudi male patient who was diagnosed with End Stage Renal Disease (ESRD) and was maintained on regular hemodialysis via right jugular vein dual lumen catheter for 10 months. Three years later, the patient presented with signs and symptoms suggestive of SVC obstruction that was successfully managed with SVC stenting.

Key words: Hemodialysis, superior vena cava, stenosis

Hemodiyaliz Hastasında Süperiyör Vena Kava Trombozu

Süperiyör vena kava (SVK)'daki kan akımının engellenmesi SVK sendromunun belirti ve bulguları ile sonuçlanır. SVK obstrüksiyonu, sağ kalbi, lenf nodlarını ve diğer mediasten yapılarını tutan bulaşıcı bir patolojik sürecin SVK'ya dıştan basısı veya invazyonu ile veya SVK içindeki kanın trombozu nedeniyle oluşabilir. Nadiren her iki mekanizma birlikte olabilir. Bu vakada 28 yaşında son dönem böbrek yetmezliği tanılı ve 10 aydır sağ jugular ven dual lümen katateri ile düzenli hemodiyaliz uygulanan bir Suudi erkek olguyu sunduk. Üç yıl sonra hasta SVK tıkanıklığını düşündüren belirti ve bulgularla başvurdu ve SVK stenti ile başarıyla tedavi edildi.

Anahtar kelimeler: Hemodiyaliz, süperiyör vena kava, stenoz

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INTRODUCTION

The superior vena cava is a thin-walled blood vessel with low intravascular pressure, enclosed in a tight compartment. It can be compressed easily because the chest has no room for expansion. Extra luminal compression of the superior vena cava by tumors or enlarged lymph nodes can occur acutely or gradually and obstruction can be complete or partial. Intraluminal obstruction of the SVC can be caused by infiltration by tumor, although thrombosis is a more common cause. Acute and complete obstruction of the SVC is caused more often by thrombosis than by compression or tumor infiltration. Syphilitic thoracic aortic aneurysms, fibrosing mediastinitis, and other complications due to untreated infections were frequent causes of superior vena cava syndrome (SVCS) in the pre-antibiotic era (1).

CASE

A twenty eight year old Saudi male patient presented in 2003 with ESRD secondary to hypertension. Renal biopsy revealed totally sclerosed glomeruli. Hemodialysis was started on November 2004 via right jugular dual lumen polyurethane permicath and continued for approximately ten months. On September 2005 we started to use left Brachial-Cephalic Arterio-Venous Fistula (A.V.F.) for hemodialysis.

In August 2008, the patient started to complain of facial swelling, dyspnea and visible dilated collateral veins over the chest wall. A chest computed tomography (CT) scan revealed occluded SVC with many dilated collaterals (Figure 1 A,B). Coagulation study showed anti-thrombin III at 40% (normal levels being 50-150%), Protein C activity at 19% (normal levels being 50-150%) and Protein S activity at 47% (normal levels being 58-127%). The diagnosis of SVC thrombosis was established and the patient was maintained on oral anticoagulant (warfarin), aiming for a target INR between 2-2.5.

In October 2008, the patient was transferred to ER because of increased neck and facial swelling that was associated with dysphagia and orthopnea. There was no history of vomiting or fever. An ENT surgeon was consulted and flexible laryngoscopy was carried out which was negative for upper airway obstruction. Ultrasound revealed sluggish flow in both internal jugular veins, with obscure subclavian veins. A chest and neck CT scan was requested, unfortunately, the patient was severely

orthopnic and could not tolerate to lie down to perform the procedure. SVC venography was done which revealed obstruction of the SVC (Figure 2). The patient was treating with anticoagulant therapy which was not enough to prevent the SVC obstruction and because of the chronic onset and sever complaining it was very late to use thrombolytic therapy ,so a radiologist intervention was consulted and a stent was placed to restore the blood flow. This was followed by marked improvement of the patient's clinical condition. The patient was discharged in good condition, maintained on Warfarin treatment with target INR between 2-2.5. Currently, the patient continues to receive haemodialysis 3 times weekly using the left A.V.F.

DISCUSSION

Nowadays, malignancy is the most common cause of the SVCS. It accounts for more than 78% of the cases, and bronchogenic carcinoma is the most common malignancy (2,3). Non-malignant causes of SVCS include granulomatous infections secondary to tuberculosis, actinomycosis, aspergillosis, blastomycosis, nocardiosis, goiter, aortic aneurysms, and histoplasmosis-related mediastinal fibrosis (4,6). Other non-malignant causes include sclerosing cholangitis and sarcoidosis. The incidence of SVC syndrome arising from benign etiologies is increasing. It is now more commonly associated with indwelling central venous devices which create a nidus for SVC thrombosis (2,7,8). Iatrogenic causes of SVCS include venous thrombosis as a consequence of central venous catheters or pacemaker catheters and also fibrosis, caused by radiation therapy of the mediastinum (8). Thrombi in the SVC were detected by trans-esophageal echocardiography in 30% of patients who had single lumen silicone rubber hemodialysis catheters (9).

Polyvinyl chloride, polyethylene and teflon catheters are associated with increased thrombogenicity, as compared to silicone rubber (10,11). Furthermore, the SVC stenosis may be induced by persistent trauma of the endothelium by the catheter tip and the higher blood flow during dialysis hours. The risk of thrombus formation, either in the vessel or in the catheter, is increased by devices inserted into the vascular system.

Patients with hypercoagulable disorders who need central venous access may require a daily low dose of warfarin to help prevent thrombus formation (12).

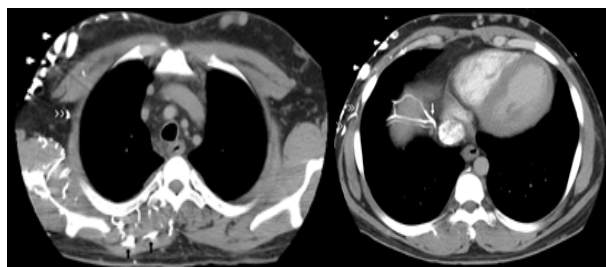


Figure 1. A: CT scan at the level of the chest shows subcutaneous venous collaterals (short arrows) and a Para vertebral collaterals (long black arrows), Note also enhancement within a thoracic vein (head arrows). B: CT scan at the level of the diaphragm shows anterior thoracoabdominal wall venous collaterals (short arrows) and a large diaphragmatic collaterals (long arrows), Note also enhancement within an intercostal vein (head arrows).

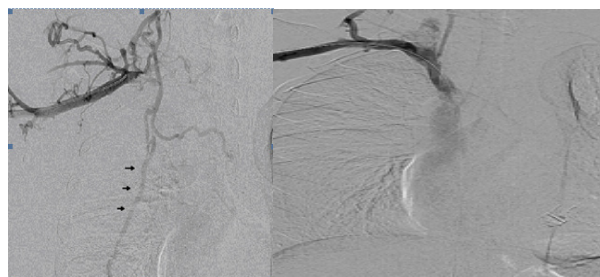


Figure 2. A: Venography shows collaterals venous circulation due to SVC Stenosis, Note enhancement the internal mammary vein (long black arrows). B: Venography after SVC Stenting

The Azygos vein is a major tributary of the superior vena cava and joins it at the level of the second thoracic vertebra (13). Impaired venous drainage above the level of the Azygos vein causes less venous pressure and less pronounced SVCS. This is because the venous return from the upper body can be redirected from the subclavian vein to the Azygos vein, the proximal vena cava and the right atrium (7,14). Impaired venous drainage below the Azygos vein is a more complex problem and causes more symptoms as the shunted blood must return to the right atrium by way of the upper abdominal veins and the inferior vena cava, which requires higher venous pressure. When venous circulation through the superior vena cava is impaired, venous hypertension, venous stasis, and decreased cardiac output result. If untreated, these will progress to thrombosis, laryngeal and cerebral edema, stupor, coma, pulmonary complications and death (14).

The development of clinical manifestations of SVCS depends on the amount of venous hypertension, the delay in circulation time, the development of collateral pathways of circulation, degree and rapidity of obstruction of the superior vena cava and the clinical signs and symptoms of the underlying causative patho-physiologic process. If onset of SVCS is gradual, symptoms may be subtle. However, rapid onset of SVCS, in the absence of collateral circulation, will cause a more dramatic and life-threatening presentation, often with neurologic and

respiratory sequelae resulting from cerebral and laryngeal edema.

Early symptoms of SVCS include; swelling of the face, arms, fingers, or neck; often the first symptom, dyspnea; which is the most common symptom and a non-productive cough (7-8). Also feelings of fullness of the head, difficulty buttoning shirt collars (a stoke sign) and in women; breast swelling, dysphagia, hoarseness and chest pain. Late symptoms of SVCS include, Life-threatening symptoms of respiratory distress e.g. orthopnea, headache, visual disturbances, dizziness, syncope, lethargy, irritability, mental status changes and the formation of esophageal varices (15). The choice of diagnostic procedures with suspected SVCS depends upon the patients' status.

The preferred diagnostic tools to confirm the diagnosis of SVCS are a chest CT scan with IV contrast and a chest MRI scan (7,16,17). CT and MRI scans are non-invasive, accurate in distinguishing between tumor mass and thrombosis as causes of SVCS, and able to document the extent and location of involvement. Chest x-ray films also may be of diagnostic value. Chest x-ray results that are associated with SVCS include a lung or mediastinal mass, pleural effusion and superior mediastinal widening (8,18). In SVCS, lung masses are frequently seen on the right on chest x-ray films because the superior vena cava enters from the right (14).

In malignancy, accurate, definitive histologic diagnosis by biopsy or cytology specimen is necessary to provide appropriate treatment of SVCS. This is because the modality of treatment is usually based on the histologic diagnosis of the underlying malignancy. Emergency treatment without histologic diagnosis is reserved for patients who demonstrate brain edema with mental status changes, decreased cardiac output with hemodynamic compromise, or laryngeal edema with respiratory compromise and impending loss of airway (16). If the development of SVCS is gradual, as occurs more commonly, the diagnostic workup should be completed first to confirm a definitive diagnosis before treatment is initiated.

The four main treatment modalities for SVCS are radiation therapy (non-small cell lung cancer), Chemotherapy (small cell lung cancer and lymphoma), Pharmacologic therapy (steroids, diuretics and thrombolytic therapy) and surgery. Thrombolytic therapy may be used when SVCS is caused by catheter-induced intraluminal thrombosis. Thrombolytic therapy or tissue plasminogen activators are used to treat catheter-induced thrombosis and can effectively lyse clots (7,19). It should be noted that treatment with thrombolytics should be initiated within five to seven days of the onset of symptoms for maximum effectiveness (6). However catheter removal may be necessary.

Anticoagulants may be used to help relieve venous obstruction by preventing thrombus formation where SVCS is caused by a tumor. However, pharmacologic management of SVCS with anticoagulant therapy is controversial (16,20). The risk of hemorrhage with anticoagulant therapy must be weighed against the possible benefits. One potential preventive measure for catheter-induced thrombosis is prophylactic administration of warfarin (7,21).

Surgical intervention for SVCS includes stent placement or superior vena cava bypass and is used occasionally when SVCS is chronic or recurrent (14). Surgical intervention in patients with malignancy-induced SVCS should be reserved for patients who have failed other therapeutic treatments such as radiation therapy and chemotherapy. Surgery to relieve the obstruction may be beneficial in patients with retrosternal goiter or aortic aneurysm (8).

In conclusion, central venous catheters in haemodialysis patients may result in superior vena cava (SVC) syndrome. With the increasing use of these catheters,

the SVC syndrome is likely to become common among hemodialysis patients. The syndrome is serious and requires early recognition and treatment, although the symptoms may be delayed, and alternate approaches for renal replacement therapy should be considered. Thrombolysis constitutes the cornerstone of therapy. If insufficient, endovascular interventions such as percutaneous transluminal balloon angioplasty or intravascular stent placement, should be considered.

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