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Case Report

Catheter-related thrombosis of Superior Vena Cava in a patient with Superior Vena Cava Syndrome

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Abstract

Background: The superior vena cava syndrome (SVCS) is defined as the set of signs and symptoms derived from superior vena cava obstruction, both intrinsic obstruction and extrinsic compression, which causes an increase in venous pressure in the upper body region.

Methods: We present the case of a male 64 years old patient with hypotiroidism, diabetes mellitus and end stage chronic kidney disease in hemodialysis treatment who presented with cervical, facial and upper right extremity edema. The tomographic contrasted study demonstrated superior vena cava thrombosis.

Results: The patient underwent catheterization for stent placement and catheter removal. Clinical superior vena cava syndrome manifestations remitted.

Conclusions: Although thrombosis is a frequent manifestation in patients with blood coagulation alterations and patients with end stage chronic kidney disease, **catheter-related thrombosis** is a rare cause of thrombosis and superior vena cava syndrome whose must common cause is neoplastic.

Key words:
Central Venous Catheter; Catheter-Related Thrombosis; Coagulation; Superior Vena Cava Syndrome; Thrombosis; Thrombus.

Introduction

Superior vena cava syndrome (SVCS) is defined as the set of signs and symptoms derived from superior vena cava obstruction,

both intrinsic obstruction and extrinsic compression, which causes an increase in venous pressure in the upper body region [1]. Most frequent clinical manifestations are listed in table 1 **(See table 1)** It´s worth to mention that the clinical presentation of SVCS depends on the obstruction localization and the size of it. The presentation of SVCS is wide; however **catheter-related thrombosis** is a rare cause that should be suspected if the most common etiologies are ruled out. In México the first cause of SVCS is the microcytic lung cancer [2]. Around the world the first cause of SVCS as it´s in México is neoplastic, and from these ones, the most common is lung cancer [1-3]. In spite of this, **catheter-related thrombosis** is a relatively common complication of central venous catheter insertion [3]. **Central venous catheter** use is ubiquitous in the end stage chronic disease patients who require hemodialysis treatment and often in patients with multiple risk factors for venous thromboembolism [3].

Table-1 : Clinical manifestations of SVCS
Facial / Cervical edema
Collateral circulation
Dyspnea
Cough
Arm edema
Facial plethora
Hoarse voice
Visual signs
Syncope
Dizziness / Headache
Confusion
Cerebrovascular event

Since its first description in 1757, SVCS has remained as a challenge for the clinician for being an entity that endangers the life of the patient [4]. The superior vena cava (SVC) in the main conduit for venous drainage of the head, neck, upper extremities and upper thorax, its main auxiliary vessel, the azygos vein, enters in the SVC just above the pericardial reflection, other collateral systems are the internal mammary veins and the esophageal vascular plexus. Despite these collateral ways, if there is clogging the SVC almost always raises the venous pressure of the upper compartment, the flow obstruction of SVC produces venous hypertension of the head, neck and upper extremities, which in turn is responsible for the clinical presentation characteristics [5-7].

Thrombosis is defined as the formation of a blood clot within the vasculature of a person, thrombosis can result from any one or any combination of the following three causes presented in table 2 (**See table 2**) [8-10]. These three causes are better known as "Virchow´s triad" [1,11].

Catheter-related thrombosis CRT is a relatively common complication of central venous catheter insertion and manipulation, the International Society of Thrombosis and Haemostasis guidelines recommend that where possible, CVCs should be inserted on the right side, in the jugular vein with the tip located at the junction of the superior vena cava and the right atrium to minimize the risk of thrombosis [11-15]. The consequences of CRT are not insubstantial; complications can include pulmonary embolism (PE) in 10–15%, loss of venous access in 10%, infection, post-thrombotic syndrome (PTS) and delays in treatment [4, 12]. The clinical features may be fairly self-evident such as arm or neck swelling and discomfort or venous distension. Knowing the pathogenesis of SVCS and thrombosis is vital for the understanding of the disease; as well as for its identification and treatment [13]. In the following paragraphs, we present a case report of a patient with SVCS secondary to SVC thrombosis.

Case presentation

We presented a case of a 64 year old male patient from Puebla, México who presented Diabetes Mellitus type two controlled with dietary and hygienic measures, controlled hypothyroidism treated with levothyroxine 100mcgr taken in fasting, gastroesophageal reflux disease treated intermittently with proton pump inhibitors, 4 year diagnosis of end-stage chronic kidney disease receiving hemodialysis

Table-2 : Thrombosis causes	
Endothelial damage	**Inflammation of the vessel wall**: Inflammation causes cytokine release and activation of the coagulation system resulting in fibrin deposition and thrombi enlarge
	Physical trauma: due to iatrogenic stimuli (repeated venipuncture, repeated injections, caustic stimuli)
	Parasites that migrate through the vessels can damage them sufficiently to cause thrombosis
Alteration in the normal blood flow	Abnormal blood flow resulting in eddy currents, turbulence or blood stasis disrupts the laminar flow of blood, bringing platelets in close contact with the vascular wall. Also, turbulence may cause endothelial injury resulting in release of tissue factor
Hyper-coagulability of the blood	Hyper-coagulability refers to those states in which thrombosis is favored due to a change in make-up of the formed elements of the blood**.**

treatment twice a week during which nurses who manipulate catheters reported a dysfunctional Mahurkar catheter which needed great manipulation for giving a good flow for renal substitutive treatment and normal venous pressure, other disadvantage included poor liquid control and poor adherence to treatment by the patient as well as excessive intake of liquids. The patient had a history of acute myocardial infarction in 2016 with conservative treatment.

The patient went to the hemodialysis unit to his usual treatment with clinical manifestations of SVCS; he referred dysphagia, inability to fall asleep in supine decubitus for shortness of breath, dyspnea and acute edema of the upper right extremity. Due to the patient´s evident symptomatology, extension studies where requested. The presented image (***See Fig 1***) was

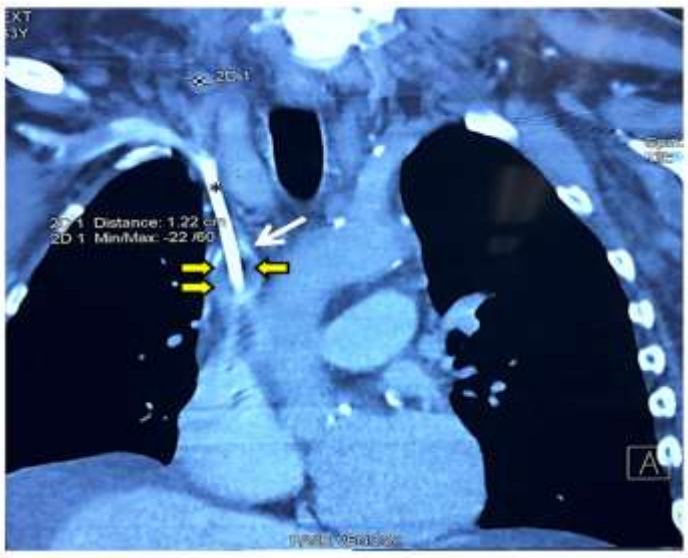

Fig-1: Thrombosis of SVC surrounding catheter

Yellow arrows mark the thrombus periphery, white arrow shows the thrombus per se and ***central venous catheter*** *is marked by an asterisk*

the request of tomographic contrasted study in this patient the time he presented with cervical, facial and upper right extremity edema. The subclavian **central venous catheter** is seen on the left side of the image marked with an asterisk. As the yellow arrows mark in the conferred image, the thrombus surrounds almost completely the lumen of the superior vena cava. The image in its venous phase shows the superior vena cava thrombosis, the white arrow demonstrate the contrast enhancement in a haloform as the periphery of the thrombus. Also, it is noticed a concentric left ventricle hypertrophy and aortic calcification. General blood studies and coagulation studies where performed reported as normal.

Our patient was referred to the Cardiology department where new blood studies were performed demonstrating normal coagulation values. Conservative treatment was established with a daily subcutaneous 60 mg Enoxaparin injection, daily 75mg Clopidrogel, continuous O2 by nasal tips at 3lt per minute and 100mg acetylsalicylic acid at night. Despite the conservative treatment one week later patient remains with the symptomatology and presented worseness of the facial and upper right extremity edema, the reason why invasive treatment was indicated. No diuretic therapy was established since the patient had diuretic-resistance. The patient underwent catheterization for stent placement and catheter removal. A new catheter was placed in left femoral for the continuation of hemodialysis treatment. Clinical **superior vena cava syndrome** manifestations remitted.

Although thrombosis is a frequent manifestation in patients with blood coagulation alterations and patients with end-stage chronic kidney disease, this case was a rare presentation of a vena cava thrombosis since our patient had normal coagulation studies. A few weeks later, patient deceased by cardiac arrest and respiratory insufficiency secondary to massive pleural leak, there was no evidence of stent collapse or death associated with the thrombosis history. The patient died due to a complication of his renal chronic disease by water retention and lung edema, his clinical record highlighted a previous history of the pleural leak for poor control of liquids and hydro electrolytic imbalance.

Discussion

In this study case and review, we describe the risk factors for developing CRT in our patient. In the clinical practice despite being relatively common, there are limited evidence-based guidelines for the best diagnostic approach. The clinical history combined with CT allows differentiating between SVC thrombosis and extrinsic compression. Among its advantages, CT not only allows the identification of the cause of the obstruction (extrinsic versus intrinsic) and its degree but also level and extent of blockade and documentation of the collateral circulation, which is especially important if a surgical bypass is expected to be performed [16].

Literature review highlights that in suspicion of SVCS and while a definitive treatment is defined it´s necessary to grant treatment with diuretics to reduce intravascular volume and initiate a short cycle of parental steroids to reduce edema, still, these therapies have not been studied in depth [17-18]. Our patient didn´t receive either diuretic because of diuretic-resistance nor parental steroids.

Our patient had a dysfunctional Mahurkar catheter which promoted excessive manipulation of the central line to obtain an adequate flow during hemodialysis treatment, if we consider the thrombosis causes previously mentioned (**See table 2**) we can assess that, these micro trauma generated by the arduous manipulation could lead to a small thrombus which became larger as more platelets, fibrin and erythrocytes accumulate until complete obstruction of the vessel lumen triggered the clinical manifestations presented by our patient. Bibliography stipulates that pathology analysis is needed to confirm the presence of malignant disease4. Despite 87%-97% of the patients with SVCS have malignant tumors, this case report a benign cause of SVCS, the causes of this syndrome are listed in table 3 (**See table 3**) [4-15].

Table-3 : Superior vena cava synrdrome causes

Benign causes	Malign causes
Intrathoracic Goiter	Microcytic Lung Cancer
Behçet Syndrome	Squamous Cell Lung Cancer
Lymphadenopathy	Lung Adenocarcinoma
Aneurysms	Non-Hodgkin Lymphoma
Mediastinitis	Big Cell Lung Cancer
Sarcoidosis	Metastatic Tumors
Pacemarker	Hodgkin´s Lymphoma
Catheter associated Thrombosis	Thyroid Cancer

SVCS may be life-threatening in select cases and may require emergent treatment. If laryngeal edema causing laryngeal constriction or cerebral edema is present, these medical emergencies require prompt management and rapid treatment of the underlying cause of SVCS [1, 16]. In this case, the SVC thrombosis led to invasive bypass treatment with stent placement to release the obstruction caused by the thrombus. If this had not been carried out the clinical SVCS sequelae might have caused long-term morbidity or mortality if left untreated. Considering the neoplastic disease as the first cause of SVCS an empiric treatment with radiation, stenting, and/or chemotherapy may be indicated even before biopsy results become available in patients with malignant SVCS [1, 18-20]. Similarly, if clinical and radiographic evidence reveals a rapidly growing tumor with a high likelihood of invading other critical thoracic structures, then prompt treatment to retard cancer growth is indicated [16-20]. Since our patient had a non-neoplastic acquired SVCS this treatment was not considered.

Central venous catheter insertion causes endothelial trauma and inflammation, which can lead to venous thrombosis [16-20]. Central venous lines are indispensable components of therapy in many cancer patients and in those undergoing hemodialysis, parenteral feeding, plasmapheresis, or administration of certain drugs [17-19]. However, there is considerable uncertainty about the risks, treatment, and prevention of **catheter-related thrombosis** (CRT) because of substantial study heterogeneity and a paucity of rigorous clinical trials on its management [20-25].

Catheter-related thrombosis can be classified into 3 types: pericatheter sheath ("fibrin sleeve"), thrombotic occlusion of the catheter lumen, and mural thrombosis, either superficial (SVT) or deep vein thrombosis (DVT) [20-25]. As mentioned before, the main risk factor in our patient was the great catheter manipulation; we could hypothetically establish that his CRT was secondary to this extrinsic factor. Thence, the clinical manifestations and extension studies for discarding and establishing the SVCS etiology are the main determinants for a quick approach and individualized treatment. In this case, despite invasive intervention with stent placement, patient deceased for other causes.

Conclusion

Although patients with end-stage chronic disease have an increased risk of thrombosis secondary to catheter manipulation, this case was a rare presentation of an SVC thrombosis since as mentioned before our patient had normal coagulation studies. However great manipulation of the central line and microtrauma generated by this manipulation was the main risk factor for presenting this complication. Clinical

examination gives the necessary data for SVCS diagnosis; the cause remains unknown until extension studies are performed.

CRT is an increasingly most frequent complication due to central venous catheter use becoming a risk factor for thrombosis that could present itself as an SVCS which is considered a medical and surgical emergency, it´s necessary a quick and certain diagnosis define treatment guidelines that nowadays tend to be more and more individualized.

The most frequent etiology of the SVCS is lung cancer; however, the thrombosis of the SVC is an increasing cause due to the increasingly frequent use of minimally invasive procedures and the use of central venous accesses. The most frequent clinical manifestation in facial edema, followed by upper limb edema, respiratory symptoms are frequent and when the condition is severe, imaging studies provide useful information to classify compression into extrinsic or intrinsic compression, and often define the origin of the obstruction.

Catheter-related thrombosis could be prevented if patients with higher risk of emboli formation and thrombosis are well-managed. Also, prophylactic measures could be taken into accounts such as inhibitors of platelet aggregation or anticoagulants. Although each patient must receive an individualized approach and treatment considering the risk factors, comorbidities or previous pharmacological treatment to avoid drug interactions and improve the long-term prognosis.

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