Refractory rhinorrhoea secondary to superior vena cava syndrome

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Abstract

Superior vena cava (SVC) syndrome is a collection of clinical signs and symptoms resulting from either partial or complete obstruction of blood flow through the SVC. This obstruction is most commonly a result of thrombus formation or tumour infiltration of the vessel wall. Incidence of SVC syndrome, post haemodialysis catheter insertion, is about 5% to 19 %, however, with regards to refractory rhinorrhoea secondary to SVC syndrome, this is the first case report in medical literature. Common symptoms of SVC syndrome are facial puffiness, upper limb oedema, hoarseness of voice, cough and rarely, bilateral reversible hearing loss and cerebrospinal fluid (CSF) rhinorrhoea. Here, we present a case of refractory rhinorrhoea and bilateral conductive hearing loss in a patient with SVC syndrome which resolved immediately after relieving the SVC obstruction. There is extreme paucity of data regarding refractory rhinorrhoea as manifestation of SVC syndrome and hence should always be considered in differential diagnosis, with predisposing thrombotic background. This case also highlights the limitations of radiological procedure and timely surgical intervention in SVC syndrome.

Keywords: SVC syndrome, rhinorrhoea, Maharashtra, India

Introduction

Case report

A 56-year diabetic male with chronic renal disease for last 5 years, was admitted to the hospital with severe cellulitis of right leg since 3-4 days. He also had complaints of facial puffiness, hoarseness of voice, headache, upper limb oedema, dry cough, bilateral hearing loss and rhinorrhea for 1 year. He had history of central venous catheter insertion for hemodialysis on the right side 18 months back. However, after 6 months of insertion, there was thrombosis of right internal jugular vein (IJV) and left sided central venous catheterization of IJV was done. Unfortunately, after 3-4 months, even left sided catheter got thrombosed and he started developing complaints of facial puffiness, headache and upper limb edema. In the interim period, right arterio-venous fistula was made for dialysis. Over another couple of months, he developed hoarseness of voice, dry cough, bilateral hearing loss and rhinorrhea. In view of above progressive symptoms, computed tomographic (CT) angiography was performed which revealed significant stenosis of right IJV with stenosis of proximal subclavian vein. Left brachiocephalic vein also showed significant stenosis with multiple collaterals, leading to SVC syndrome. In view of thrombotic occlusion of left brachiocephalic vein, ballon angioplasty was done using 8 French catheter. Following the procedure, his symptoms improved for a transient period of 3-4 months and then reappeared. For his constant and excessive running nose, patient was treated with multiple antihistamines and anti-inflammatory medications by various subspecialties, but had no relief. During this admission, he presented with sepsis. Examination of right lower limb revealed severe cellulitis. He also had facial puffiness and hoarseness of voice. Otolaryngological examination revealed normal appearing nasal cavity with bilateral conductive hearing loss. Biochemical markers showed neutrophilic leucocytosis and deranged renal parameters. Cellulitis was managed conservatively. In view of past history and recurrence of SVC syndrome like symptoms, CT angiography of upper limb was done which showed long segment chronic thrombotic occlusion of right distal jugular vein for length 5 cm, proximal segment of right subclavian vein and complete short segment thrombotic occlusion of left brachiocephalic vein (Fig. 1). In view of ongoing sepsis, surgical intervention was deferred by vascular surgeon and was planned for balloon angioplasty. Balloon angioplasty was performed by interventional radiologist and his symptoms of hearing loss and rhinorrhea improved instantaneously (Fig. 2). Remaining symptoms like puffiness of face, limb edema and hoarseness of voice improved over next 48 hours. Subsequently, patient was discharged. On follow up at 3 months, he had partial recurrence of symptoms of SVC syndrome. He was then planned for

surgical intervention and underwent bypass graft from left IJV to SVC. At 10 months of follow up, patient is comfortable with well controlled symptoms.

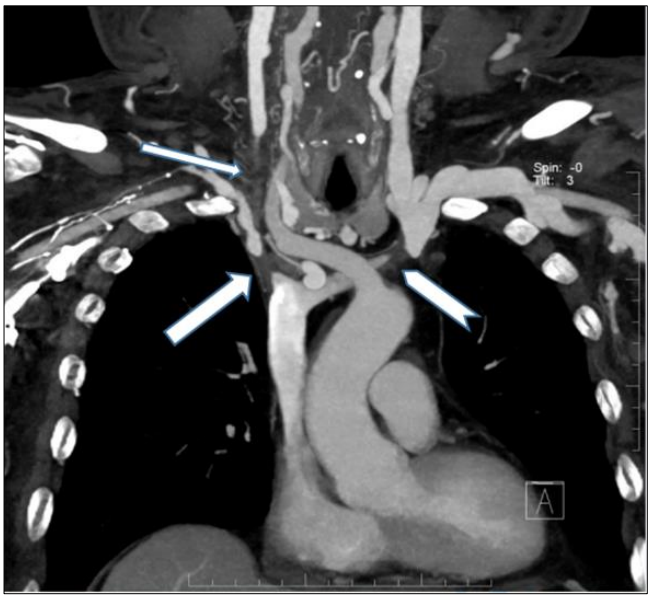

Fig 1: CT angiography of upper limb showing long segment occlusion of right distal jugular vein (thin arrow), proximal segment of right subclavian vein (thick arrow) and complete short segment occlusion of left brachiocephalic vein (chevron)

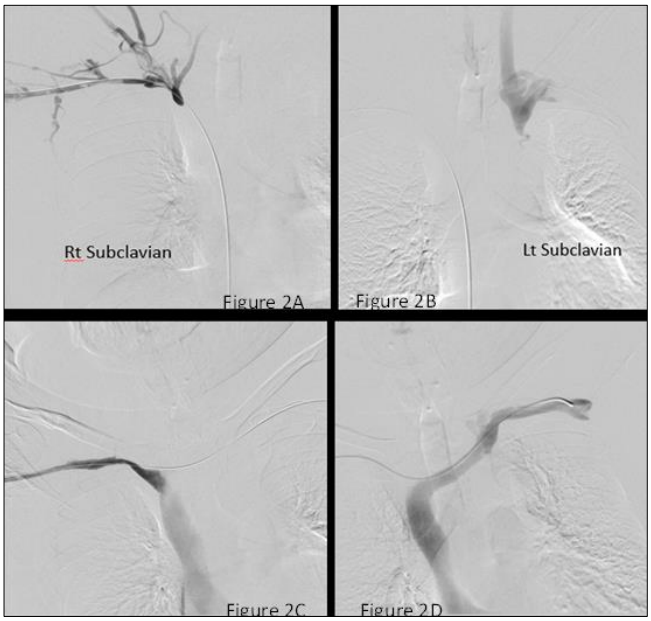

Fig 2: A/B: Digital subtraction images showing complete occlusion of bilateral subclavian veins. C/D: Digital subtraction imaging showing patency of right and left subclavian veins post balloon angioplasty

Discussion

Most common cause of SVC syndrome, after malignancy, is intravascular device for haemodialysis, cardiac procedures etc. [1]. The incidence of SVC syndrome is increasing as the population for the need of dialysis is increasing. About 60-85% of SVC syndrome are due to mediastinal malignancies like mediastinal lymphoma, thymoma and thyroid malignancies. Rest of 15-40% of cases are due to non-malignant tumours and central venous catheters induced thrombosis [2]. However, data on central venous catheterization related thrombosis are inconsistent. The true incidence may be underestimated because many patients are asymptomatic. Even in the presence of symptoms the diagnosis is misunderstood. In indwelling venous devices, incidence of thrombosis varies from 1.5 to 13% [3]. SVC syndrome can be classified anatomically and clinically.

Table 1: Anatomical classification: Doty and standford's classification [4]

Type 1	Stenosis up to 90% of the supra-azygos SVC
Type 2	Stenosis more than 90% of the supra-azygos SVC
Type 3	Complete occlusion of SVC and reverse blood flow in Azygous vein
Type 4	Complete occlusion of SVC with involvement of major tributaries and azygous vein

Pathogenesis of CSF rhinorrhoea in SVC syndrome can be understood by knowing that, any disruption in venous flow proximal to the right heart chambers could potentially result in elevated intracranial venous pressures, which in turn leads to CSF leak through the cribriform plate, presenting as rhinorrhoea. The clinical manifestations of SVC syndrome depend on acuteness and severity of thrombosis. If the SVC syndrome is gradual in onset, the symptoms may be mild. But in case of rapidly progressive SVC syndrome and in absence of collaterals, symptoms are more life threatening, as in our case. Early symptoms may be swelling of face, arms and neck, mild dry cough, hoarseness of voice, chest pain and dysphagia. Late symptoms may include orthopnoea, headache, syncope, and oesophageal varices. In rare cases patient present with bilateral reversible hearing loss and CSF rhinorrhoea, as noticed in our patient, the exact incidence of bilateral hearing loss and CSF rhinorrhoea is not known due its extreme rare presentation and reporting. In our case, beta-2 transferrin test confirmed the diagnosis of CSF rhinorrhoea. In imaging modalities high resolution CT and magnetic resonance imaging remains gold standard due to their ability to define the exact location and extent of thrombus, collaterals and tumour, thus facilitating, intervention, if required in the same sitting. Treating the underlying cause for SVC syndrome is of paramount importance. Conservative management for mild cases, SVC syndrome due to clot or acute thrombus is treated with low molecular weight heparin, warfarin and catheter directed thrombolysis. In malignant cause symptoms of SVC syndrome improve after irradiation or chemotherapy up to 80%. Interventional radiology management includes use of balloon angioplasty and stenting, but these are helpful in benign cause or short segment of thrombotic lesion [4, 6]. Surgical management is needed only in severe cases, which cannot be managed with medical or radiological intervention. Saphenous and femoral vein graft are best autologous conduits whereas externally supported polytetrafluoroethylene graft [PTFE] is the least thrombogenic prosthetic graft for SVC reconstruction [5]. Conclusion As the incidence of central venous line, being used for various procedures is increasing, thrombosis of SVC should a part of differential diagnosis for refractory rhinorrhoea. Emphasis should be laid upon the rarer symptoms and to be addressed early during their course, for a better and improved long-term outcome.

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