Annals of Clinical and Medical Case Reports

Case Report ISSN 2639-8109 | Volume 14

Endovascular Superior Vena Cava Stenting As Effective Therapy of A Life Threatening Epistaxis in A Patient with Hereditary Hemorrhagic Telangiectasia

Pierre Yves Marcy (MD)[1,*], Lacout Alexis (MD)[2] and Mostfa El Hajjam (MD, Associate Professor)[3]

[1]Department of Radiodiagnostics and Interventional Imaging, Polyclinics ELSAN Group, PolyClinics Les Fleurs, Quartier Quiez, 83189, Ollioules, France

[2]Radiologist, Centre de diagnostic ELSAN, 15000 Aurillac, France

[3]Pluridisciplinary HHT team, Ambroise Paré Hospital, 9- Avenue Charles de GAULLE; 92100 Boulogne Billancourt France, Groupement des Hôpitaux Ile-de-France Ouest, Assistance Publique - Hôpitaux de Paris Université de Versailles Saint Quentin en Yvelines, France

[*]Corresponding author:

Pierre Yves Marcy,
Department of Radiodiagnostics and Interventional Imaging, Polyclinics ELSAN Group, PolyClinics Les Fleurs, Quartier Quiez, 83189, Ollioules, France

Received: 26 Oct 2024
Accepted: 16 Nov 2024
Published: 21 Nov 2024
J Short Name: ACMCR

Citation:

Pierre Yves Marcy (MD), Endovascular Superior Vena Cava Stenting As Effective Therapy of A Life Threatening Epistaxis in A Patient with Hereditary Hemorrhagic Telangiectasia. Ann Clin Med Case Rep. 2024; V14(9): 1-6

Keywords:

Superior vena cava stenting; Epistaxis; Hereditary hemorrhagic telangiectasia; Rendu Osler; Central venous catheter, Superior vena cava occlusion

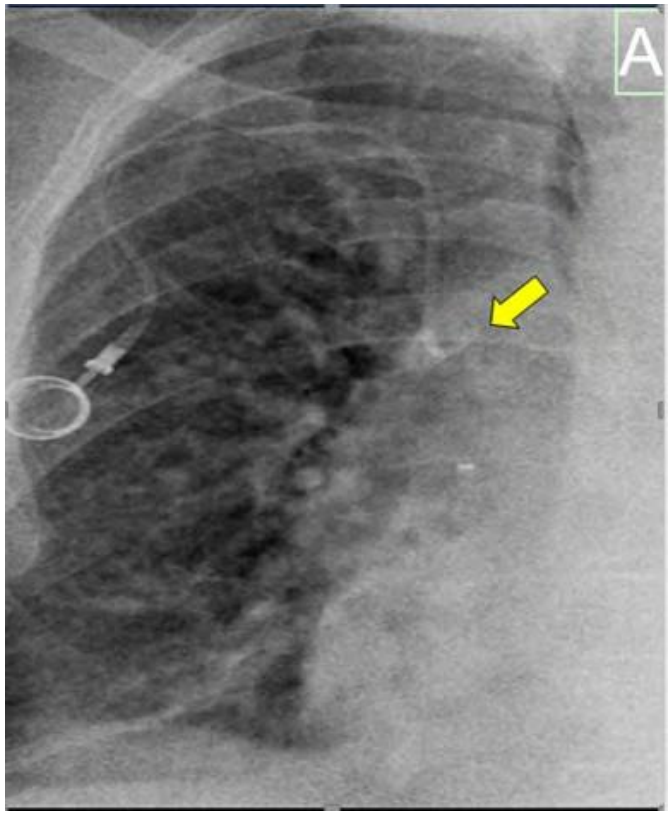
1. Abstract

Indwelling port catheters are nowadays the main etiology of benign superior vena cava (SVC) syndrome. SVC obstruction increases the venous blood pressure upstream, and subsequently into the arterio-venous sac malformations of the nasal cavity, leading to the high risk of recurrent epistaxis in hereditary hemorrhagic telangiectasia (HHT) patients. In the present case, percutaneous SVC recanalization made possible to deal successfully with epistaxis in a HHT patient presenting with complete occlusion of SVC related to central venous catheter secondary misplacement into the azygos venous arch.

2. Case Description

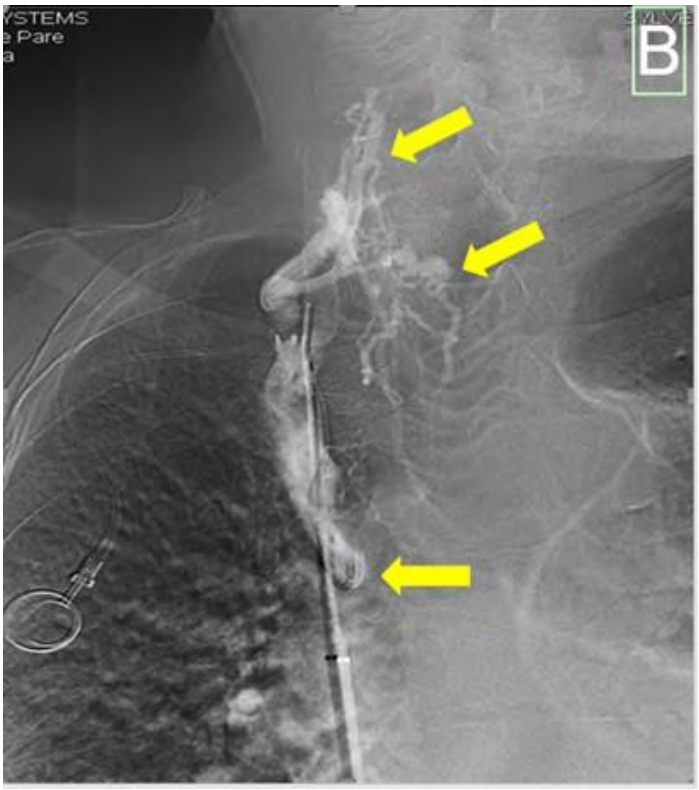
A 87-year-old man (body mass index 30) with known HHT disease presented with SVC syndrome, and worsening of epistaxis. The epistaxis had been treated by conventional therapy [1], bilateral surgical ligation of sphenopalatine arteries [2], several percutaneous arterial embolization procedures [3] and laser therapy in 1985 and 1986 respectively. In 2010, owing to the gravity of epistaxis, an implantable venous device was placed into the SVC, through the right subclavian vein, to perform blood transfusion in this elderly patient suffering from severe chronic anemia. In August 2016, the venous device occluded and recanalization was successfully achieved by using urokinase protocol. Patient subsequently presented further exacerbating episodes of epistaxis resulting in acute anemia (hemoglobin 7.7g/dL). In addition, arm and facial swelling progressively developped with concomitant orthopnea that were consistent with clinical SVC syndrome. CT scan showed SVC occlusion, staged II according to the Stanford classification [4]. CT scanning displayed a chronic and organized thrombus located at the distal tip of the venous catheter that was mislocated into the azygos vein arch (Figure 1A, B). Percutaneous SVC recanalization was successfully performed through the right common femoral vein, using a steel Wallstent endoprothesis 18 mm x 60 cm, a 11French introducer sheath, a 0.0035in guide wire. Final angiography showed SVC lumen complete reopening (Figure 1C). Venous obstruction related symptoms, particularly epistaxis, resolved immediately and durably. The port catheter was surgically explanted two days after the endovascular treatment. The SVC stent was controlled patent at 6 months CT scan follow-up (Figure 1D) and episodes of epistaxis never recurred clinically at a follow up of eight years.

Figure 1A: Pre-Treatment Findings Chest X-Ray shows catheter tip hooked into azygos vein arch.



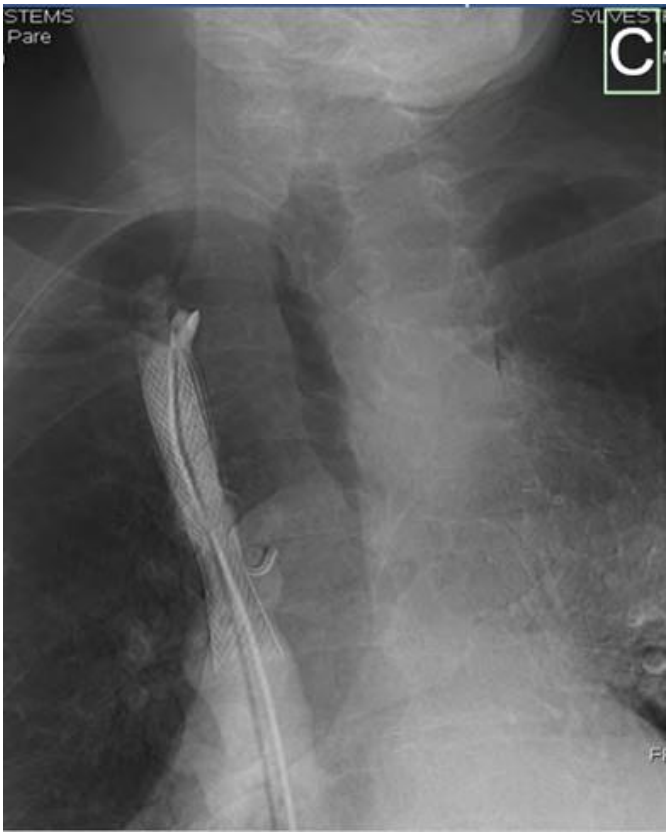
Chest front x- ray shows subclavian vein (SCV) port catheter tip mislocation into azygos arch. Key feature displays a retracted catheter tip inclined medially to the SVC long axis (arrow). Secondary catheter tip displacement was likely due to severe coughing or vomiting efforts.

Figure 1B: Caval angiography displays SVC occlusion (Azygos Catheter tip, arrow) and dilated venous cervical collateral pathways (oblique arrows).



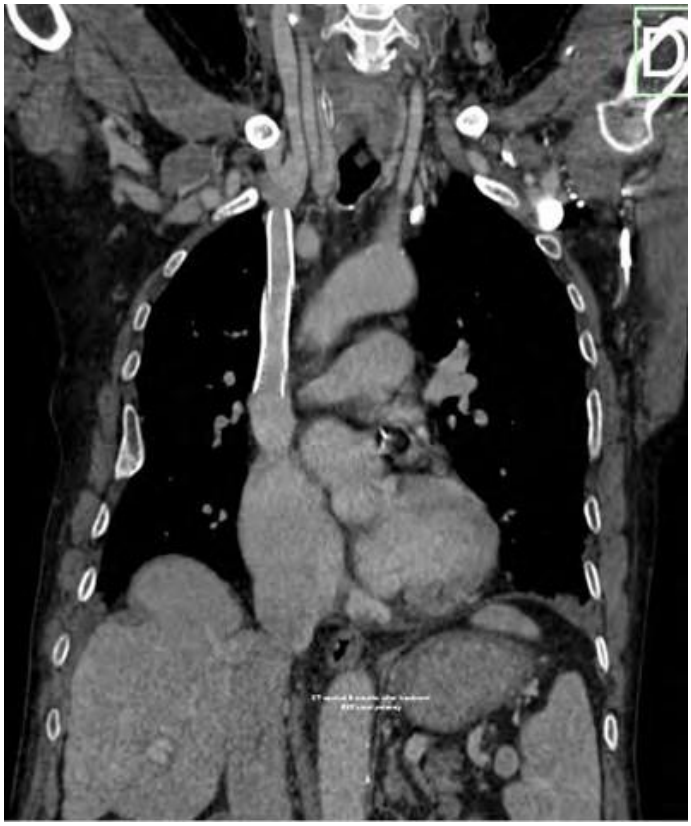
Percutaneous SVC recanalization was successfully performed through the right common femoral vein, using a 18mmx 60mm steel Wallstent endoprothesis, a 11French introducer sheath, a 0.0035in-guide wire.

Figure 1C: Caval angiogram displays antegrade SVC flow, disappearance of collaterals after SVC stent placement.



Post- SVC stenting phlebography shows complete reopening of the SVC while disappearance of the collateral veins. SCV catheter will be retrieved few days after the SVC stenting procedure.

Figure 1D: Enhanced – Chest CT control at 6months follow-up. SVC stent is patent



Chest CT scan follow-up at six months confirms durable SVC patency, without collateral vein opacification. SVC clot has completely disappeared.

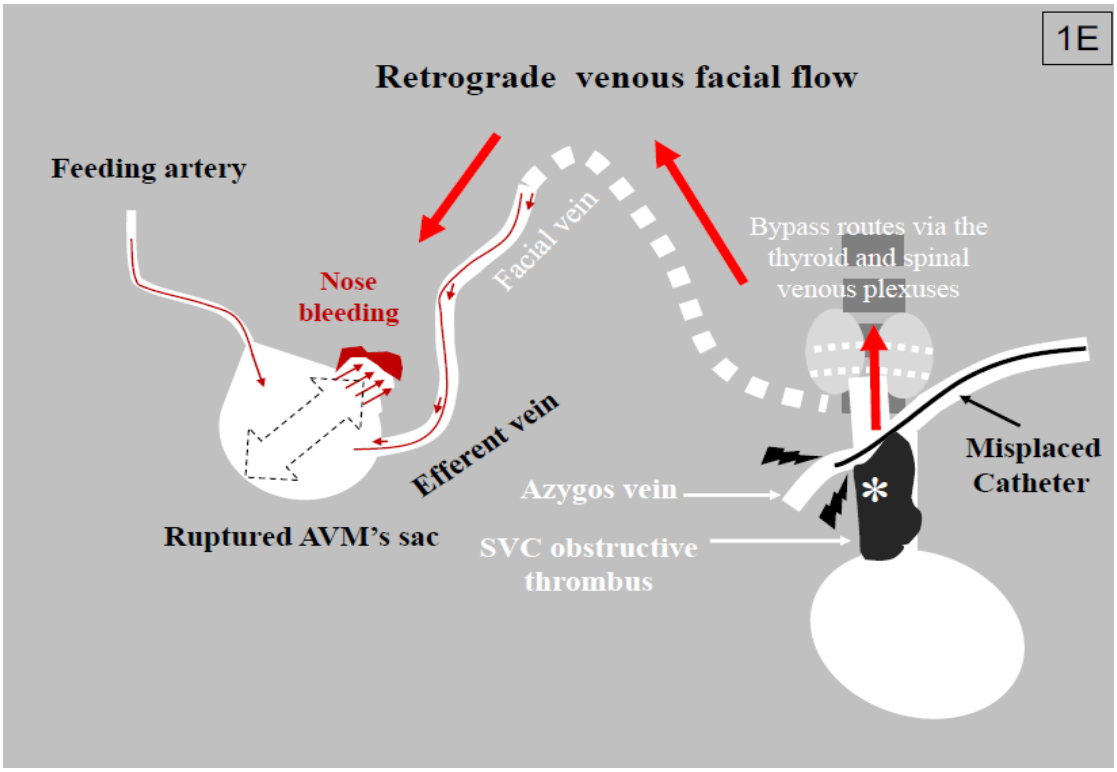


Figure 1E: Scheme of arteriovenous sac malformation (AVM) sac, before SVC stenting. The dilation of the nasal AVM sac is due to increased blood pressure and retrograde flow (red arrows) into the SVC, internal jugular veins, facial veins and the AVM's sac dilated efferent vein. SVC clot (*) explains the SVC occlusion related to secondary catheter tip misplacement. Double black arrow stigmatizes the rupture of the dilated AVM's sac, and subsequent nose bleeding.

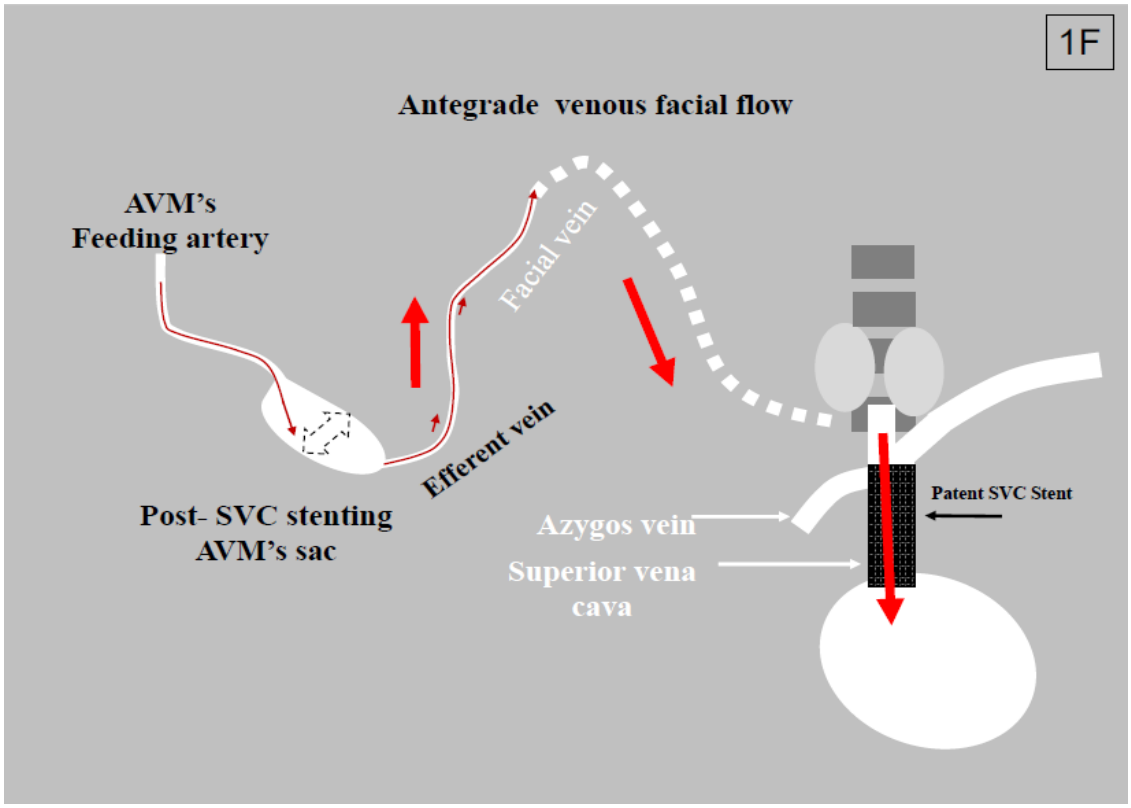


Figure 1F: Scheme of arteriovenous sac malformation, after SVC stenting. The AVM efferent vein flow has become antegrade thanks to the SVC reopening (vertical arrow). AVM's sac diameter has dramatically decreased, active nose bleeding definitely will stop.

3. Discussion

SVC syndrome was first described in 1795 and is well documented in the scientific literature. More than 80% of reported cases are due to malignant obstruction. Indwelling port catheters are nowadays the main cause of benign SVC occlusion [5, 6]. Dedicated studies have shown that the risk of SVC occlusion is higher when the implanted catheter is too short, particularly when located in front of the azygos vein arch or at the confluence of the innominate veins, and in case of left sided insertion [7, 8]. As initial chest x- ray showed satisfactory implantation of the central venous catheter tip 2cm above the atrial caval junction, we assume that secondary catheter malposition into the azygos vein occurred for intrathoracic hyperpressure reason. Thus we strongly advocate for 2cm – overtrimming the central venous catheter when left-sided inserting or in case of obese patients. Overweighted patients often present with perivenous loose connective tissue and potential venous catheter loops giving a higher elasticity reserve to the inserted catheter, thus a higher risk of secondary catheter displacement.

Patients with benign SVC syndrome have a normal life expectancy when compared to SVC patients presenting with malignant disease. Percutaneous SVC endoprothesis insertion is a safe and efficient percutaneous procedure and definitely is the treatment of choice of benign SVC syndrome, allowing a long-term relief of the clinical symptoms with a primary patency rate > 90% [5,6]. However some life threatening SVC stenting complications have been reported to date, including the following: acute recurrence of SVC, cardiac tamponade, stent migration, and overload syndrome [7,8]. SVC recurrence is commonly due to acute clot thrombosis of the SVC stent lumen, namely in case of stent migration or tilting, persistence of severe SVC stenosis (incomplete reopening of SVC lumen), and/or absence of anticoagulant therapy during and after the percutaneous procedure. Another cause of central vein reocclusion is due to too early insertion of the central venous catheter. Indeed, performing concomitantly to the SVC stenting procedure, the placement of a central venous port catheter device is at high risk of acute clot formation within the SVC stent lumen. As a matter of fact, the SVC metallic stent lumen usually requires 48hours to optimal opening, and a minimum of two weeks to become endothelialized. We thus strongly recommend in case of absolute indication of central venous line insertion to plan the procedure under anticoagulant therapy and when the implanted SVC stent is completely open. It is preferable to implant the device via the femoral route. This is of particularly importance in young patients in whom preservation of the thoracic network is of prime importance. Right femoral vein access is strongly recommended as this provides a straightforward and safe access to catheterize the SVC occlusion in a podo cranial way. The angioplasty balloon catheter is thus easily safely and progressively inflated along the transcardiac vertical guidewire to compress the blood clot against the SVC wall. Then, the SVC stenting can easily be performed by using a metallic 18 mm x 60mm Wallstent endoprothesis. Downsizing risk is secondary cardiac embolization [9]. Patients with concerning symptoms should have their stent surgically removed or retrieved by endovascular methods [10]. In the present case, SVC obstruction promoted facial and intracranial venous hyperpressure, thus increasing pressure into the efferent veins of the nasal mucosae HHT malformation, secondary nose arterio-venous malformation sac dilation and rupture, which explains the occurrence of the epistaxis. As arterio-venous malformation sac pressure dropped dramatically after successful SVC stenting, nosebleeds stopped eventually, and SVC clinical symptoms completely resolved.

4. Conclusion

The use of central venous catheters is a common iatrogenic cause of SVC syndrome. In HHT patients, central venous occlusion not only leads to a series of clinical complaints but also causes dysfunction of the central venous catheter and jeopardizes the future of suitable vascular access in fragile patients who often require blood transfusion in case of severe and chronic epistaxis. Standard rules should be undertaken to prevent risks of catheter-related complications into the central venous system, particularly optimal positioning of the catheter tip, namely when left sided (i.e. at or even below the cavo-atrial junction).

5. Funding

The authors declare that this study received funding from Association BonSens.org to cover the publication fees.

References

1. Robard L, Michel J, Prulière Escabasse V, Bequignon E, Vérillaud B, Malard O, Crampette L; SFORL Work-Group. Guidelines of the French Society of Otorhinolaryngology (SFORL) (short version). Specific treatment of epistaxis in Rendu-Osler-Weber disease. Eur Ann Otorhinolaryngol Head Neck Dis. 2017; 134(1): 37-41.
2. Kitamura T, Takenaka Y, Takeda K, Oya R, Ashida N, Shimizu K, Takemura K, Yamamoto Y, Uno A. Sphenopalatine artery surgery for refractory idiopathic epistaxis: Systematic review and meta-analysis. Laryngoscope. 2019; 129(8): 1731-1736.
3. De Bonnecaze G, Gallois Y, Bonneville F, Vergez S, Chaput B, Serrano E. Transnasal Endoscopic Sphenopalatine Artery Ligation Compared with Embolization for Intractable Epistaxis: A Long-term Analysis. Am J Rhinol Allergy. 2018; 32(3): 188-193.
4. Lacout A, Marcy PY, Thariat J, Lacombe P, El Hajjam M. Ra- dioanatomy of the superior vena cava syndrome and therapeutic orientations. Diagn Interv Imaging 2012; 93(7-8): 569-77.
5. Qanadli SD, El Hajjam M, Mignon F, de Kerviler E, Rocha P, Barré O, Chagnon S, Lacombe P. Subacute and chronic benign superior vena cava obstructions: endovascular treatment with self-expanding metallic stents. AJR Am J Roentgenol.1999; 173(1): 159-64.
6. Breault S, Doenz F, Jouannic AM, Qanadli SD. Percutaneous endovascular management of chronic superior vena cava syndrome of benign causes : long-term follow-up. Eur Radiol. 2017; 27(1): 97-104.

7. Puel V, Caudry M, Le Métayer P, Baste JC, Midy D, Marsault C, Demeaux H, Maire JP. Superior vena cava thrombosis related to catheter malposition in cancer chemotherapy given through implanted ports. Cancer. 1993; 72(7): 2248-52.
8. Zhao X, Wang K. Hemorrhagic pericardial tamponade in a hemodialysis patient with catheter-related superior vena cava syndrome: a case report. J Cardiothorac Surg. 2024;19(1): 151.
9. Marcy PY, Magné N, Bruneton JN. Strecker stent migration to the pulmonary artery: long-term result of a “wait-and-see attitude”. Eur Radiol. 2001; 11(5): 767-70.
10. Zvavanjanja RC. Percutaneous retrieval of migrated Viabahn stent from a segmental pulmonary artery. CVIR Endovasc. 2018; 1(1): 1.