Vena Cava Superior Syndrome Six Years after Central Venous Catheter Removal in a Patient on Hemodialysis

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Keywords
Vena cava superior syndrome · Central venous catheter · Hemodialysis

Abstract
Vena cava superior (VCS) syndrome is rarely seen as a complication of central-venous-catheter placement. Usually, the syndrome appears when the presence of the catheter causes intraluminal obstruction or thrombosis. In this case report, however, we describe a patient on intermittent hemodialysis who had been free of any venous central line for over 6 years, presented with a VCS syndrome. The CT scan showed an absent VCS without extravascular compression. Previous catheter placement was diagnosed as the case of the VCS syndrome. It is important to realize that VCS syndrome can occur late after removal of central venous catheters, and thus, clinicians should be aware of its symptoms in any patient who has had an upper central line in the past medical history.

Introduction
Vena cava superior (VCS) syndrome is the collection of symptoms resulting from obstruction of the VCS. Symptoms of VCS syndrome include facial and upper extremity edema, shortness of breath, headache, and distended neck and chest veins [1]. It is by far most frequently caused by external compression due to malignancy, mostly lung cancer or lymphoma. Other possible causes include hygroma, mediastinitis, or an (intra-)vascular problem such as an aneurysm.
or thrombosis. Also, central venous catheters can cause VCS syndrome by both intraluminal obstruction and endovascular damage [1–3].

The pathogenesis of VCS in relation to catheter placement is not yet completely understood. The underlying mechanism will most likely be based on vascular wall stress by both direct trauma to the endothelium by the catheter and also movements by respiration and a high flow through the vein during hemodialysis [4, 5]. Catheter placement in the right internal jugular vein has a lower risk of VCS syndrome compared to left-sided placement for multiple reasons which are mainly dependent on its anatomy (see Fig. 1) [6]. The left internal jugular vein has a smaller cross-sectional area than the right-sided jugular vein. Also, it has a longer and more indirect route to the right atrium. Finally, the left subclavian vein crosses the brachiocephalic artery and aorta which gives repetitive compression and mechanical trauma through pulsation [7–10].

Since central venous catheters are frequently used as vascular access for hemodialysis, it is important to consider this diagnosis in patients with signs and symptoms compatible with VCS syndrome and an indwelling catheter [11]. However, also years after removal of a central venous catheter the syndrome can occur. To highlight this possibility, we present a case of new onset VCS syndrome more than 6 years after central-venous-catheter removal.

Fig. 1. Anatomy of the central chest veins.
Case Presentation

A 72-year-old female patient who has been hemodialysis dependent since 7 years due to hypertension presented at the weekly routine visit with her nephrologist with distended veins on the upper chest and swelling of the supraclavicular fossa on both sides. Precise timing of first occurrence of these symptoms was unclear but was considered to be some weeks previous. Due to these present clinical features, the patient was suspected of having a VCS syndrome. A CT scan was therefore performed that showed an obliterated VCS (Fig. 2), thereby confirming the diagnosis of VCS syndrome. Regarding the cause of the VCS syndrome, no external compression of the VCS by any tumor was seen on the CT scan. The presence of extensive collateral veins formation implicated an already long-existing situation. Therefore, we reviewed her past medical history.

Approximately 7 years earlier, 2.5 months after the start of hemodialysis treatment, a nontunneled venous catheter (Gamcatch Jugcath, 11 French, 15 cm) was placed in the right jugular vein because of fistula malfunctioning for which a vascular procedure was performed (Fig. 3a).
Two months later, because of continuing cannulation problems, a new brachiocephalic fistula was created on the left side. During the same procedure, a tunneled central venous catheter (Retro long-term HD catheter, 14.5 French, 23 cm) was placed with ultrasound guidance in the left jugular vein for hemodialysis access during fistula maturation (Fig. 3b). Unfortunately, also with this fistula there were maturation issues at first and cannulation problems for which a percutaneous balloon angioplasty was performed approximately 8 months after fistula creation. After this procedure, cannulation of the fistula was without problems and the tunneled catheter was removed. As per protocol, flow measurements were performed periodically afterward. During following years, the patient was seen by her nephrologist weekly for routine controls. Dialysis efficacy was adequate with urea reduction ratios continuously above 0.74 and weekly eKt/V ranging from 3.6 to 3.9. No interventions were necessary in these years until occurrence of the VCS syndrome.

After the diagnosis of the VCS syndrome, the patient was referred to a vascular expertise center where a percutaneous recanalization of the VCS and left brachiocephalic vein was performed and a self-expanding stent (14 × 60 mm) was placed. She was prescribed oral anticoagulant medication for 6 months after the procedure. Symptoms of the VCS syndrome subsided after this procedure.

Discussion/Conclusion

The VCS syndrome is a description of symptoms which appear when the VCS is either compressed or narrowed causing increased venous pressure. Thrombosis in the VCS mostly occurs due to the presence of a central venous catheter, causing intraluminal obstruction and triggering thrombosis [1–5]. In our patient, it is however remarkable that there has been a 6-year period free of the central-venous-catheter presence before development of the VCS syndrome. This suggests she has either been asymptomatic for a relatively long period or there has been ongoing vascular inflammation after catheter removal which eventually resulted in occlusion of the VCS.

Two recent studies on central venous stenosis in hemodialysis patients with a tunneled catheter reported a remarkably high incidence of 4.3–9.4% [12, 13]. Notably, both reported that only the minority of patients were symptomatic: 18–30%. Risk factors for development of venous stenosis were similar in both studies: longer duration of catheter carriage increased the incidence of venous stenosis (relative risk [RR] 1.47 [12] vs. RR 1.40 [13]), whereas older age seemed to have a slightly protective effect (RR 0.96 [12] and RR 0.70 [13]). One study also reported the number of catheters (RR 2.2 per additional catheter) and the presence of a left-sided catheter (RR 2.6) to increase stenosis risk [13]. Moreover, venous thrombosis can be a sign of hypercoagulability such as deficits in protein C or S [14]. Our patient was not checked for this due to an evident different underlying mechanism of her VCS syndrome.

Not many data on VCS syndrome post-central venous catheter are available. Previous case reports describe either development of acute VCS syndrome during presence of a catheter [15, 16] or short after its removal (2006 [17]: VCS syndrome 2 weeks after catheter extraction). To the best of our knowledge, this is the first case report describing occurrence of VCS syndrome several years after catheter removal.

Treatment options for VCS stenosis include surgical bypass or endovascular therapy such as stenting or recanalization. In specific cases, catheter-based thrombus removal can be considered, depending on longevity [18]. Endovascular therapy has high rates of success (83–100%) and low complication rates (0–7.5%), including restenosis [18, 19].

In conclusion, occlusion of VCS syndrome as a complication of central venous catheter is rare but should be recognized. Even years after removal of the catheter, one should consider
the diagnosis when symptoms of VCS syndrome occur in a patient who had central-venous-catheter placement in the past medical history.

**Statement of Ethics**

This research work does not contain human or animal subject research material. This review of patient data did not require ethical approval in accordance with local/national guidelines. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

**Conflict of Interest Statement**

The authors have no conflicts of interest to declare.

**Funding Sources**

This research was performed without any funding.

**Author Contributions**

Michelle Janssen and Susan Logtenberg both had substantial contribution to the conception of this work. Both wrote the main of the paper and interpreted and analyzed the data. The work was critically revised together and then finally approved. Both are accountable for all aspects of this work.

**Data Availability Statement**

All data that support the findings of this study are included in this article. Research data are not publicly available as it is contained within a confidential medical record. Further inquiries can be directed to the corresponding author.

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