Venous infarction secondary to congestive encephalopathy from central venous occlusive disease in a chronic hemodialysis patient: A case report

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Abstract:
Central venous occlusive disease secondary to chronic hemodialysis catheterization rarely progresses to encephalopathy, cerebral infarction, and/or hemorrhage. A 59-year-old male with 15 years of haemodialysis-dependent end-stage renal disease presented with acutely altered mental status, extensor rigidity with left hemiparesis and equal, but small and nonreactive pupils. Magnetic resonance imaging demonstrated infarction and cerebral edema. Cranial angiogram through right brachial artery injection revealed right subclavian vein opacification via a patent AV-fistula and retrograde flow to the right internal jugular vein and superior sagittal sinus secondary to occlusion of the brachiocephalic vein. All cerebral and right upper extremity venous drainage occurred via the contralateral venous outflow tract. Internal carotid artery injections revealed significant venous congestion. Despite successful angioplasty with stenting and resolution of venous flow reversal, the patient failed to recover neurologically. The devastating nature of the presented case emphasizes the need for frequent neurologic evaluation of such patients to avoid catastrophic cerebrovascular injury.

Keywords:
Central venous occlusive disease, chronic hemodialysis, venous congestive encephalopathy, venous infarction

Introduction
Central venous occlusive disease (CVD) occurs in up to 40% of patients with chronic haemodialysis central venous catheters (CVC).[1] Venous congestive encephalopathy (VCE) is a rare but devastating and potentially fatal complication.[1‑3] The presented case describes a patient with venous infarction secondary to VCE and radiographic demonstration of a cerebral vascular steal phenomenon secondary to right brachiocephalic vein (BCV) stenosis in the setting of a chronic haemodialysis CVC. This report aims to encourage the implementation of neurologically focused screening questions and a brief neurological examination at each dialysis appointment in an effort to promptly identify and treat clinically significant CVD.

Case Report
A 59-year-old male with end-stage renal disease (ESRD) on hemodialysis (>15 years) presented with altered mental status and demonstrated extensor rigidity with left hemiparesis. Magnetic resonance imaging (MRI) of the brain showed extensive subcortical signal abnormalities and restricted diffusion consistent with infarction and cerebral edema [Figure 1a-c]. Head and neck computerized tomography-angiography
demonstrated asymmetric dilation of the right superior ophthalmic vein and adjacent facial veins which prompted digital subtraction angiography (DSA) [Figure 1d]. Evaluation of the venous phase revealed stenosis of the right BCV [Figure 2f]. Cranial angiogram revealed patent filling of the right subclavian vein through the right upper extremity fistula with retrograde flow to the right internal jugular vein (IJV) and superior sagittal sinus due to stenosis of the right BCV. All cerebral and right upper extremity venous drainage occurred via left IJV. In addition, delayed contrast transit with direct internal carotid artery injection was noted, suggesting delayed flow and drainage may be secondary due to severe venous congestion [Figure 2a-d].

The patient underwent right BCV balloon angioplasty with stent placement [Figure 3a-c]. A DSA performed 3 days after stent placement demonstrated minimal

Figure 1: Magnetic resonance imaging and computed tomography angiography (CTA) at Presentation: An magnetic resonance imaging at presentation with T2-FLAIR hyperintensity (a), restriction on Diffusion-weighted imaging sequence (b) and infarction on ADC sequence (c) within the right occipital lobe. CTA demonstrates a dilated R superior ophthalmic vein (arrow) and adjacent facial veins (d)

Figure 2: Preintervention digital subtraction angiography: A preintervention digital subtraction angiography demonstrates significant vascular congestion (a-d). Occlusion of the brachiocephalic vein (arrow in e) resulted in retrograde venous flow with all cerebral and right upper extremity drainage occurs via the contralateral venous outflow tract (arrows in f-g). The right extremity arteriovenous fistula flow shows flow reversal (arrows in h-j)

Figure 3: Postintervention DSA: A postintervention digital subtraction angiography (a-d) demonstrates stable vascular congestion (c), but improved opacification of the venous sinuses and resolution of the redirected flow down the contralateral internal jugular vein (d). Successful revascularization through the endovascular stent (arrow) is noted without retrograde flow (e-g)
change in venous congestion but improved outflow through the right IJV and resolution of the described vascular steal phenomenon [Figure 3d-g]. The patient demonstrated slight neurological improvement initially but expired a month later following emergent arteriovenous fistula (AVF) ligation of a ruptured pseudoaneurysm related to chronic ESRD, complicated by multiple episodes of postoperative cardiac arrest.

**Discussion**

The presented case describes the potentially devastating nature of VCE from CVD secondary to a chronic hemodialysis CVC. The pathophysiology of this condition is best described by a “two-hit” hypothesis.[4] First, increased venous return secondary to an AVF graft results in increased venous pressure.[4] Second, endothelial trauma from multiple and/or chronic cannulization results in intimal hyperplasia resulting in resistance to cerebral venous drainage.[4] In the presented patient, the severely elevated central venous pressure from the AVF, CVD and chronic heart disease overcame the IJV valves resulting in retrograde flow into the cerebral sinuses.[2,5]

The exact pattern of retrograde flow observed on preintervention DSA has been demonstrated only once in the literature [Table 1 and Figure 2e-j].[5] Retrograde flow caused an increase in venous volume and pressure transmitted to the high-flow cerebral sinus system. This resulted in cerebral circulatory stagnation, increased cerebral venous pressure, and subsequent extravasation into cerebral parenchyma, ultimately leading to infarction [Figure 2c].

Infarction associated with CVD has been reported twice previously [Table 1].[1,6] However, neither case described such severe retrograde flow into the superior sagittal sinus and/or contralateral sinus drainage.[1,6] The cortical

| Table 1: A review of previous cases describing neurologic sequela secondary to haemodialysis catheter associated central venous occlusive disease |
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| Case | Age/ gender | HD catheter (time) | Presentation | CVD location | Treatment | Outcome |
| Presented case | 59/ male | Yes (15 years) | AMS, ischemic infarct, left hemiparesis | Left BCV | Angioplasty with stent | Minimal improvement, modified rankin score 5 |
| Prasad et al.[1] | 47/ male | Yes | AMS, SDH | Left BCV | Angioplasty with stent | Improved right extremity strength, decreased left upper extremity edema and hypertonia |
| Samaniego et al.[3] | 50/ female | Yes (11 years) | 2 weeks headache, AMS | Right BCV | AVF ligation | Return to baseline 1 week postoperative |
| Herzig et al. case 1[3] | 73/ male | Yes+pacemaker | AMS, headache, papilledema | Left BCV | Failed recanalization, AVF ligation | Return to baseline POD2 |
| Herzig et al. case 2[3] | 67/ female | Yes | 2 weeks AMS, Rupper extremity weakness and myoclonic epilepsy | Left BCV | Angioplasty with stent, repeated 7 months after initial procedure | 1st stent thrombosed. Seizure free at 4 months follow 2nd operation |
| Simon et al.[6] | 65/ male | Yes | Headache, tinnitus, vision change | Right subclavian vein | Angioplasty | POD1 resolution of headache, 5 months postoperative return of visual acuity |
| Hartmann et al.[5] | 59/ female | Yes (3 years, 2 years) + pacemaker (18 years) | 3d progressive occipital headache | Left BCV | AVF ligation | Return to baseline 1 week postoperative |
| Nishijima et al.[6] | 47/ female | Yes (5 years) | AMS, seizure, R hemiplegia | Left BCV | AVF ligation | Immediate improvement of AMS, right hemiplegia. Irreversible venous infarction on MRI |
| Saha et al.[7] | 53/ female | Yes (3 years) | 2 h headache, vomiting, AMS | Left IJV | AVF ligation | Immediate return to baseline |
| Mackay and Biourse[5] | 60/ female | Yes | Headache, papilledema | Right subclavian vein | AVF ligation | Improved visual function and resolution of papilledema at 4 weeks postoperative |
| Nishimoto et al.[8] | 62/ female | Yes (9 years) | Dyspnea followed by POD2 headache and generalized seizure | Left BCV | 1st—angioplasty with stent; 2nd AVF ligation | Immediate resolution of headache and seizures |
| Lal et al.[10] | 62/ male | Yes (3 years) | Headache, papilledema, vision changes | Right BCV | AVF ligation | Return to baseline at 6 weeks |
| Molina et al.[11] | 74/ male | Yes (5 years) | Headache, papilledema, vision changes | Right/left BCV | AVF ligation | Improved papilledema, vision improved from 20/400 to 20/200, no further headaches |

HD: Hemodialysis, CVD: Central venous occlusive disease, AMS: Altered mental status, SDH: Subdural hematoma, BCV: Brachiocephalic vein, IJV: Internal jugular vein, AVF: Arteriovenous fistula, POD: Postoperative day, MRI: Magnetic resonance imaging
venous distribution of the infarcts on imaging [Figure 1] was likely due to the coexistence of a hypoplastic right transverse sinus to coupled with increased cortical reflux from impeded venous drainage secondary to BCV stenosis. Despite successful vascular intervention and resolution of the vascular steal phenomenon, return to neurologic baseline in our patient did not occur, as has been described in the majority of similar case reports, due to his death from ESRD complications 1 month after discharge.[1,6] Reversal of the congested capillary phase [Figure 2c and 3c] was unable to be evaluated in our patient given his expiration secondary to complications from ESRD prior to being able to repeat DSA.

To our knowledge, 12 previous cases [Table 1] report clinical findings consistent with VCE secondary to CVD similar to the presented case. The location of CVD included nine within the BCV, two within the subclavian vein and one within the IJV.[1-11] Management of these patients varied. Relief of CVD was achieved by either angioplasty with or without stenting at the site of occlusion, ligation of the AVF, or a combination of these interventions.[1-11] Unlike the presented patient, who passed 1 month after discharge, the vast majority (83%, 10 of 12 cases) of previously reported patients returned to neurological baseline within 6 months and immediate return was noted in a few.[1-11]

Currently, there are no recommendations regarding screening for clinically significant CVD. Previous cases were evaluated following symptom development and in some, intervention occurred weeks afterward.[1-11] While this seems sufficient in most cases, our patient highlights the potential for severe morbidity and mortality caused by VCE from CVD. We recommend simple, cost-effective screening by review for new-onset neurologic symptoms and neurologic exam to evaluate for changes in neurologic baseline at each dialysis appointment. While symptoms such as headache can be nonspecific, serial evaluation of the patient is advisable to monitor for the progression of symptoms that may indicate symptomatic CVD and the need for prompt evaluation with appropriate cranial imaging.[12] It is important to note that universal radiographic screening for CVD in asymptomatic patients is not likely to be beneficial as most asymptomatic patients do not experience complications. One study suggested prophylactic intervention may result in the progression of stenosis, though unlike our patient, the study patients did not have CVCs.[12]

Due to increasing placement of AVF for chronic hemodialysis patients and growing awareness of the potential for neurovascular complications, it is possible that the incidence of CVD cases may increase. The complex nature of this pathology and potential for devastating neurologic consequence necessitates further interdisciplinary discussion regarding early symptomatic CVD detection and intervention. Further study to evaluate the clinical sensitivity of the described screening method in the prevention of catastrophic neurologic injury secondary to VCE from CVD is necessary.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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