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CASE REPORT

Facial edema associated to central vein stenosis in a chronic kidney disease patient

Edema facial associado a estenose de veia central em paciente com doença renal crônica

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Abstract

Introduction: central vein stenosis (CVS) is a common complication in chronic kidney disease (CKD) patients, leading to vascular complications and higher mortality. Exposure to catheters is an important risk factor, and there is a wide spectrum of clinical presentation, including edema of the upper arm, neck, upper chest, and face, impairment of dialysis efficiency, and reduction of vascular accesses. **Case report**: a 41-year-old woman with CKD on dialysis and a history of multiple vascular accesses was admitted with a 3-month history of cervical and left hemiface edema. An angiotomography revealed marked stenosis in the proximal segment of the left brachiocephalic vein, and a venography confirmed the diagnosis of CVS, with subocclusive stenosis of the venous trunk and significant collateral veins. An endovascular treatment was performed with balloon angioplasty. She presented considerable improvement in the edema after the procedure. **Conclusion**: to avoid the development of central vein obstruction, it is important to enhance our knowledge about the main risk factors associated with this pathology. Although it is a treatable condition, recurrence of symptoms may occur, and subsequent vascular interventions may be needed. Therefore, prevention strategies are the most efficient approach.

Keywords: central venous stenosis; chronic kidney disease; central catheter; hemodialysis.

Resumo

Introdução: a estenose venosa central é uma complicação comum em pacientes com doença renal crônica (DRC), levando a complicações vasculares e maior mortalidade. A exposição a cateteres é um importante fator de risco, e a sua apresentação clínica envolve um amplo espectro de sinais e sintomas, incluindo edema em membros superiores, região cervical, tórax e face, comprometimento da eficiência da diálise e redução de sítios viáveis para confecção de acessos vasculares. **Relato de caso:** paciente do sexo feminino, 41 anos, com DRC dialítica e história prévia de múltiplos acessos vasculares, foi admitida com história de edema em região cervical e hemiface esquerda há três meses. A angiotomografia revelou estenose acentuada no segmento proximal da veia braquiocefálica esquerda, e a flebografia confirmou o diagnóstico de estenose venosa central, com estenose suboclusiva do tronco venoso e veias colaterais significativas. Foi realizado tratamento endovascular com angioplastia com balão, apresentando melhora considerável do edema após o procedimento. **Conclusão**: para evitar o desenvolvimento de obstrução venosa central, é importante aprofundar o conhecimento a respeito dos principais fatores de risco associados a esta patologia. Embora corresponda a uma condição tratável, o paciente pode evoluir com recorrência dos sintomas, e intervenções vasculares subsequentes podem ser necessárias. Portanto, as estratégias de prevenção são a abordagem mais eficiente.

Palavras-Chave: estenose venosa central; doença renal crônica; cateter central; hemodiálise.

INTRODUCTION

The acknowledgment of central and peripheral venous systems in chronic kidney disease (CKD) patients is essential to guarantee long-term vascular preservation. Venous obstruction due to catheter-related thrombosis is a common complication in CKD patients on dialysis[1]. The obstruction of a central vein has a major impact on the survival of those patients since the impairment of its patency has repercussions on the surgical creation of vascular accesses (VAs), whether arteriovenous fistula (AVF) or a prosthetic arteriovenous graft (AVG).

High exposure to catheters is a risk factor for central venous stenosis (CVS)[1,2] and the real incidence and prevalence of CVS in the hemodialysis patients is unknown since most studies are usually limited to the evaluation of symptomatic patients. Some patients may remain asymptomatic, developing signs and symptoms of CVS only after an AVF or AVG creation in the upper limb ipsilateral to the obstruction. The increase in blood flow after a VA creation leads to the symptoms of venous hypertension[3,4].

We describe the case of a patient with CKD on dialysis and a history of multiple vascular accesses who developed CVS and was admitted to a tertiary hospital.

CASE REPORT

A 41-year-old female patient, with hypertension for over ten (10) years and chronic kidney disease (CKD) on dialysis for eight (8)

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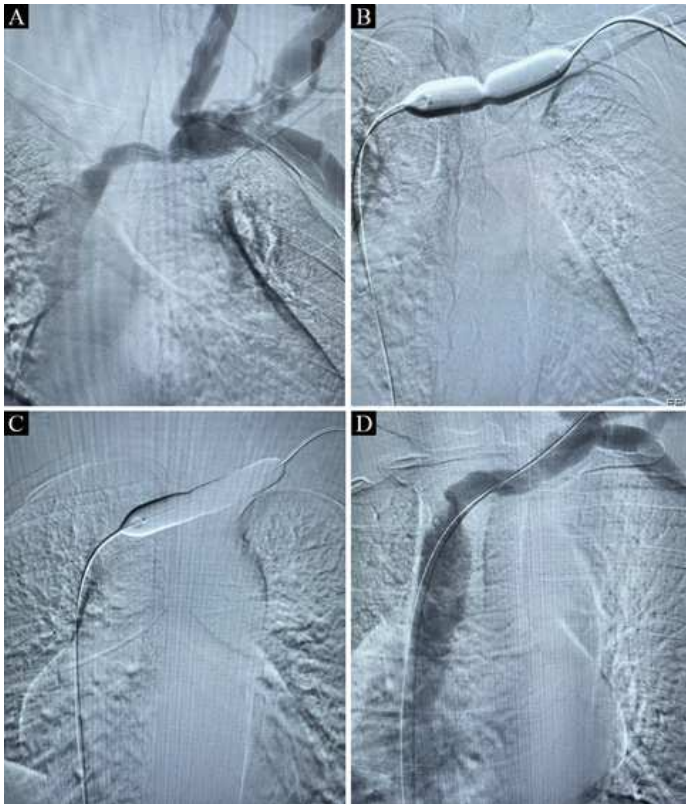
years, was admitted to the emergency department of a tertiary hospital complaining about cervical and left hemiface edema, which had been progressive for three (3) months. Her blood pressure was 208 x 121 mmHg, her heart rate was 59 bpm, and her temperature was 36.2ºC. Physical examination revealed cervical, left hemiface (figure 1A), left upper limb edema, tearing and ipsilateral conjunctival hyperemia, and significant collateral circulation in the cervical region and left hemithorax. She had a brachiocephalic arteriovenous fistula (AVF) in her left upper limb. Cardiopulmonary auscultation and examination of the abdomen and lower limbs showed no alterations.

The history of vascular accesses included several double-lumen catheters (in the right and left internal jugular veins and in the left femoral vein) and a radiocephalic AVF in the left upper limb, which had been used for around six (6) years. The patient brought an angiotomography, which showed marked stenosis in the proximal segment of the left brachiocephalic vein, with no obvious current thrombosis, leading to dilation and tortuosity of the upstream homolateral cervical and subclavian veins. The patient underwent a phlebography, which confirmed subocclusive stenosis of the venous trunk and significant collateral veins (figure 2A). An angioplasty of the central venous stenosis was performed using a balloon catheter (figure 2B, 2C, and 2D) without complications. After the procedure, she presented considerable improvement in the edema in the left hemiface (figure 1B), cervical region, and upper limb.

Figure 1. (A) Left hemiface edema preoperatively. (B) Significant reduction of edema 24 hours after the vascular procedure.

Figure 2. Phlebography images. (A) Subocclusive stenosis of the venous trunk and presence of collateral veins. (B) Balloon showing the area of stenosis. (C) Fully inflated balloon. (D) The final result with a reduction in collaterals after correction of the stenosis.



DISCUSSION

Venous obstruction is defined as a pathological narrowing of the vascular lumen that causes an impairment of blood flow. It can be partial (stenosis) or complete (occlusion). Stenosis or occlusion of the central veins can lead to severe venous hypertension and impaired VA blood flow, which may increase morbidity and mortality in CKD patients[3].

The veins of the thoracic central venous system include the intrathoracic segments of the internal jugular vein, subclavian veins, brachiocephalic veins, and superior vena cava. There are three predominant mechanisms of venous obstruction: venous wall thickening, endoluminal obstruction, and extrinsic compression[5]. Venous wall thickening is the most common mechanism, whose pathophysiology is related to the inflammatory response of the endothelium and oxidative stress, which may be a result of trauma from central venous cannulation, presence of a foreign body, as well as increased flow and turbulence due to long-term catheters used[3].

Catheters are often associated with an increased risk of thrombosis. There is activation of coagulation factors in response to endothelial damage, which can lead to endoluminal obstruction. In addition, infections associated with those catheters may exacerbate inflammation and predispose to the

development of CVS[4]. Placement of multiple catheters, longer duration, location in the subclavian vein, and placement on the left side of the neck also seem to contribute to an increase the incidence of CVS[6]. The presented patient had a history of multiple VAs, including several double-lumen catheters, as well as a radiocephalic AVF in the upper left limb (ipsilateral to the obstruction), which may have contributed to the development of symptomatic CVS.

A stenosis becomes symptomatic in hemodialysis patients if increased venous pressure and blood flow after a VA creation overwhelms the collateral venous and lymphatic drainage[7]. The clinical findings vary according to the obstruction site and the associated mechanism, including dilated and tortuous collateral veins over the ipsilateral upper arm, neck, upper chest, and face, as well as serious arm edema and disabling pain[7,8]. Pleural effusion, laryngeal edema, and restrictive pulmonary involvement can occur in severe cases. Respiratory symptoms can be seen in patients who develop superior vena cava syndrome[4,5].

The signs and symptoms related to VA dysfunction during hemodialysis include the presence of tortuosity and aneurysmal dilation of the veins, collateral formation, prolonged bleeding after dialysis, increased venous pressure, and vascular access thrombosis. Increased venous pressure can even reduce the efficiency of dialysis in these patients[4].

Besides careful history and clinical examination, duplex venous ultrasound can be useful to confirm the diagnosis of CVS, which may reveal an absence of respiratory variation in vessel diameter and the presence of collateral veins. However, assessment using duplex ultrasonography may be impaired in patients with significant muscle mass and/or obesity. Central angiography is the gold standard for the diagnosis of CVS[4,6]. The diagnosis of the presented patient was confirmed by angiography, which showed subocclusive stenosis of the venous trunk and significant collateral veins.

A conservative approach may be an option in the management of patients with chronic obstruction and adequately developed collaterals[4,6]. Based on two retrospective studies that evaluated the patency of central veins in patients with asymptomatic versus symptomatic CVS, with results showing a more rapid progression of stenosis in asymptomatic patients undergoing angioplasty, current KDOQI guidelines do not recommend endovascular intervention unless there is a clinical indication[9]. If hemodialysis efficiency is compromised or if the patient presents persistent moderate to severe clinical signs and symptoms, the intervention is indicated. In such cases, the endovascular approach is the cornerstone of treatment[9,10,11].

The first line of treatment for symptomatic patients is balloon angioplasty, with intraluminal stenting being reserved for cases of therapeutic failure[9]. The presented patient underwent angioplasty of CVS using a balloon catheter, with considerable improvement in the edema after the procedure. Although short-term results of endovascular treatment are encouraging, with success rates reaching 90%, recurrence is frequent due to the elastic nature of CVS, and repeated percutaneous transluminal angioplasties (PTAs) may be required. Studies show that long-term patency of central veins may be as low as 25% at 12 months[4,12]. Open surgical approaches with bypass are usually reserved for refractory cases[9].

In summary, it is important to prioritize the prevention of CVS by controlling the main risk factors related to this condition. The most important measure for CVS prevention is avoidance of central venous catheters[12] to ensure vascular preservation in patients with CKD.

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