Endovascular repair of abdominal aortic aneurysm and left common iliac artery in a patient with severe hemophilia C

Correção endovascular de aneurysma de aorta abdominal e artéria ilíaca comum esquerda em paciente com hemofilia C grave

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Abstract

Factor XI deficiency, also known as hemophilia C, is a rare hereditary blood disease that manifests with persistent bleeding after surgery, trauma, menorrhagia, and dental extractions. This article reports an endovascular repair of a patient diagnosed with an aortic and left common iliac aneurysm, with severe factor XI deficiency (factor XI activity below 20%). The procedure was successfully performed with management of the coagulation disorder by preoperative and postoperative infusion of plasma and laboratory control of the coagulation.

Keywords: factor XI deficiency; aortic aneurysm; endovascular procedures.

Resumo

A deficiência do fator XI, também conhecida como hemofilia C, é uma doença hematológica hereditária rara, que se manifesta clinicamente com hemorragia persistente após cirurgias, traumas, menorragias e extrações dentárias. Neste artigo, relatou-se a correção endovascular de um paciente com aneurisma de aorta e de artéria ilíaca comum esquerda em um paciente portador de deficiência *major* do fator XI (atividade do fator XI inferior a 20%). O procedimento foi realizado com sucesso, com o manuseio do distúrbio da coagulação por meio da infusão de plasma fresco no pré-operatório imediato e no pós-operatório, e controle laboratorial da coagulação do paciente.

Palavras-chave: deficiência do fator XI; aneurisma aórtico; procedimentos endovasculares.

Introduction

Factor XI deficiency, also known as Rosenthal syndrome or hemophilia C, is a rare hereditary blood disease that affects 1 in 100.000 people. Transmission is autosomal recessive. The deficiency is considered severe in homozygote or compound heterozygote, and mild in heterozygote. It usually manifests with persistent hemorrhage after surgeries, traumas, menorrhage and dental extractions[1]. Those who have this pathology usually present normal or prolonged prothrombin time activity and normal thrombin time. The lower the factor XI dosage, the more severe the hemorrhagic disorder. Patients with factor XI dosage lower than 20% carry a major deficiency. Levels higher than 20% describe patients with a minor deficiency, who rarely present with severe hemorrhage[2].

Thus, it is a challenge to submit patients with severe hemophilia C to major surgeries. The possibility of intense hemorrhage due to clotting disorders can be prevented or minimized with an integrated team that is prepared to use

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specific hemoderived products for such cases. In this context, a case of endovascular repair of abdominal aortic aneurysm and common iliac artery in a patient with major deficiency of the XI factor is reported.

Case

A 75-year-old asymptomatic patient, during an ultrasound examination to assess prostatic disease, was diagnosed with a saccular abdominal aortic aneurysm with maximum diameter of 4.6 cm and common left iliac artery aneurysm of 3.2 cm (Figures 1 and 2).

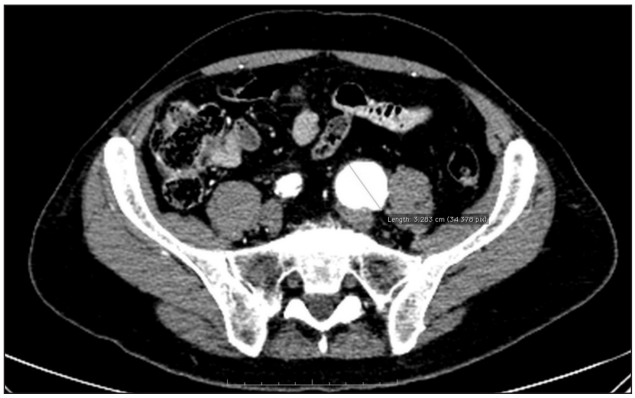

Figure 1. Angiotomography cut showing aneurysm of the common left iliac artery with 3.2 cm in diameter.

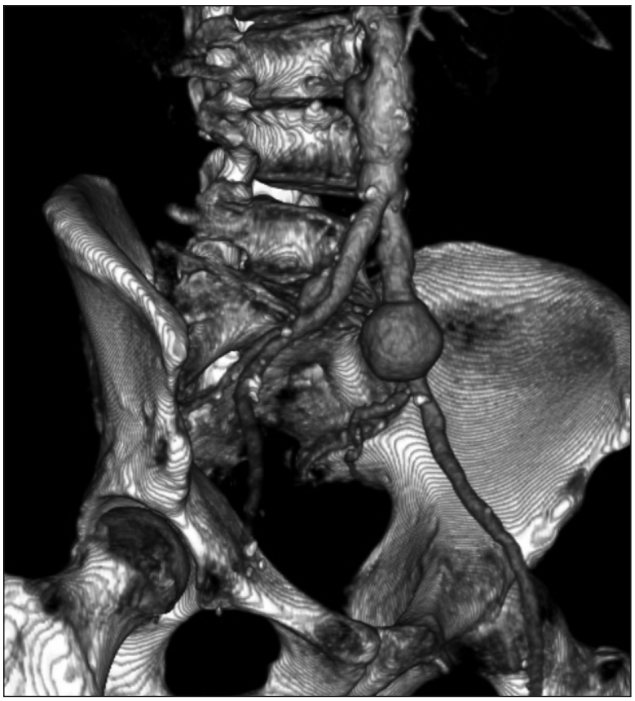

Figure 2. Reconstruction of angiotomography showing aneurysm of the common left iliac artery.

The patient had undergone carotid angioplasty and presented incessant and excessive bleeding at the puncture site. He was referred to hematology to search for clotting disorders, and lab tests showed the XI factor dosage lower than 1%, thus characterizing that the patient had a major factor XI deficiency.

In the presented case, the choice was for the endovascular therapeutic of the aortic and common iliac aneurysms. Since the patient had severe hemophilia C, he was submitted to specific preparation for this clotting disorder. The patient received infusion of four units (880 mL) of fresh plasma immediately before the surgery.

The procedure was performed under general anesthesia, invasive pressure monitoring and femoral cutdown. Full anticoagulation was performed with 5,000 U of intravenous heparin. Considering the characteristics of the aneurysm and the results, a Zenith Flex® graft with 24 mm diameter body and 82 mm in length was chosen. The main body endoprosthesis was implanted through the right access, and a 12x105 mm graft was implanted to treat the common iliac aneurysm through the left femoral. The implantation of a 12x54 mm graft was necessary to complete fixation of the endoprothesis at the right side. Control arteriography showed graft patency and endoleak absence (Figure 3). Heparinization was reversed with the infusion of protamine sulphate (50 mg). Estimated blood loss was 150 mL.

The patient was referred to the Intensive Care Unit (ICU) for post-operative care. During the first 24 hours, he received 200 mL of FFP every 6 hours and had no intercurrences. He was discharged on the third postoperative day, with no hematomas on the surgical sites. On the 13th postoperative day, the patient was asymptomatic and underwent a control angiotomography, which did not show any abnormalities.

Discussion

No previous literary evidence was found concerning the endovascular treatment of aortic aneurysm in patients with severe hemophilia C. There are reports of conventional treatment for abdominal aortic aneurysm in three patients with hemophilia A[3-5] and one with B[6]. There is one case report of a patient with severe hemophilia B (factor IX activity <1%) who was successfully submitted to endovascular treatment[7]. This patient had a proper preoperative preparation with the infusion of the isolated factor IX.

The association of aortic aneurysm and severe hemophilia C is very unusual, and is owed not only to the

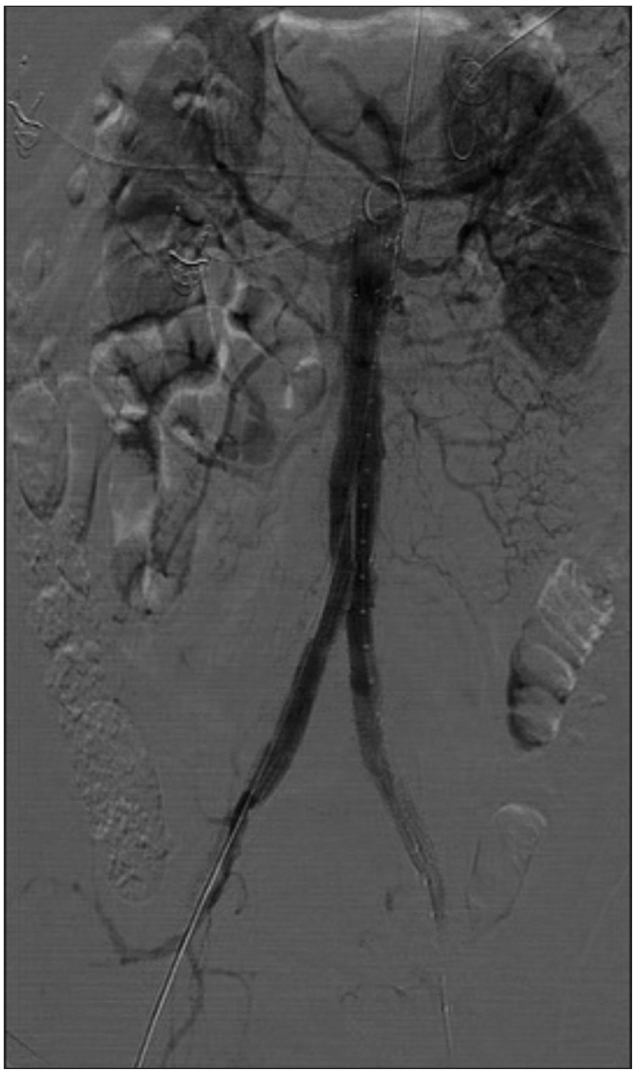

Figure 3. Intraoperative arteriography showing patency of the prosthesis and absence of endoleaks.

rarity of hemophilia C, but also to the higher mortality rate of hemophilic patients, with mean life expectancy of 63 years[8].

Conventional surgeries for aortic aneurysm repair in patients with no coagulopathy have estimated blood loss of 750 to 1,700 mL[9], which tends to be higher among patients with unfavorable aneurysm anatomy and longer postoperative period[9]. This patient had a favorable anatomy aneurysm, and the Zenith Flex® graft was chosen because of the available measures, the space given for the contralateral limb catheterization and the team's experience.

Compared to conventional treatment, endovascular repair is associated with significant blood loss reduction and the need for transfusions[10,11]. Thus, if anatomy allows, endovascular repair is considered as the method of choice for patients with significant clotting disorders.

Surgical repair in this case was based on iliac artery diameter and on the saccular form of the aortic aneurysm. There is also the fact that hemophilic patients would hardly survive the rupture of an aortic aneurysm, even if tamponade.

The blood disorder in the presented case was controlled by the preoperative infusion of fresh plasma, when the patient presented factor XI activity <1%, and by controlling the clotting with lab tests. With no need for new infusions of fresh plasma, the prothrombin time remained in accordance with the international normalized ratio (INR), between 0.9 and 1.2, and the factor XI activity was from 50 to 100% until the fourth postoperative day, when the patient was discharged with no hemorrhagic or thrombotic problems.

The present conduct proved to be a good option for major surgeries in patients with severe hemophilia, since the coagulation behavior of the patient was practically normal with the use of fresh plasma and with the strict control of coagulogram.

Conclusion

This case report showed that endovascular repair of abdominal aortic aneurysms can be safely performed in patients with severe hemophilia C. An adequate evaluation and preoperative preparation, as well as a careful perioperative multidisciplinary approach, are necessary to prevent these patients from having hemorrhagic complications.

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Statistical analysis: **N/A**
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