Conventional and endovascular treatment for a rare combination of diseases: superior vena cava syndrome and aortoiliac aneurysm – control after 12 months

Tratamento convencional e endovascular para rara associação de doenças: síndrome de veia cava superior e aneurisma aorto-ilíaco – controle após 12 meses

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Abstract

The association between superior vena cava syndrome and an aorto-iliac aneurysm is not common. The approach to each of theses diseases can be either by the conventional way with open surgery or by endovascular techniques. We report the two methods of surgical intervention and discuss their indications and results in this particular case.

Keywords: superior vena cava syndrome; iliac aneurysm; blood vessel prosthesis..

Resumo

A associação entre a síndrome de veia cava superior e uma dilatação aneurismática das artérias aorta e ilíacas não é comum. A abordagem de cada uma destas patologias pode ser efetuada através do modo convencional, com cirurgia aberta ou pela técnica endovascular. Neste trabalho, relatamos as duas modalidades de intervenção cirúrgica executadas e discutimos suas indicações e os resultados deste caso em particular.

Palavras-chave: síndrome de veia cava superior; aneurisma ilíaco; prótese vascular.

Introduction

Superior vena cava and other central veins thrombosis are not rare entities, but benign causes are uncommon, accounting for not more than 15 to 22% of such thrombosis¹. Central venous catheter-induced thrombosis has become the leading benign cause of central veins occlusion due to the increasing number of invasive procedures, such as central venous access for dialysis, invasive monitoring and implantation of pacemakers²⁻⁴. The association of superior vena cava syndrome with an aorto-iliac aneurysm makes the case to be presented rarer still. In this paper, we report the conventional and endovascular approaches for the management of a patient with this association, discussing indications and the sequence of procedures, as well as the outcome after 12 months of follow-up.

Case report

A 56-year-old male patient was admitted at the emergency room complaining of shortness of breath and flushed red face. He had undergone permanent pacemaker implantation

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on the right infraclavicular region after episodes of bradycardia and syncope, 11 years earlier. Inframammary migration had led to pulse generator replacement and repositioning of the pacemaker wires 3 years earlier. Six months after the latter procedure, the patient presented with episodes of intermittent feeling of chest oppression, dyspnea, facial edema and plethora. He reported progressive increase in intensity and frequency of such episodes. Physical examination revealed dyspnea at rest, neck, chest and upper-extremity edema, intense facial plethora and a pulsatile mass in the hypogastrium extending to the left inguinal region. The results of routine laboratory exams were normal and chest radiography showed mediastinal enlargement caused by previous congestive heart failure. Thyroid ultrasonography and coronariography were normal. Upper GI endoscopy showed varices in the upper third of the esophagus. Transesophageal echocardiogram showed images suggestive of thrombus around the pacemaker wire. Computed tomography (CT) scan of the chest did not disclose perihilar adenopathy nor mediastinal or pulmonary mass, confirming the benign origin of the syndrome. Fibrosis was observed in the juxta-atrial segment of the superior vena cava (Figure 1). Abdominal and pelvic CT scan showed a 3 cm in diameter aneurysm in the distal third of the abdominal aorta and a bilateral common iliac artery aneurysms, measuring 2 cm in the right and 6 cm in the left (Figure 2). Upper extremity phlebography showed slow flow in the left brachiocephalic trunk, reversed blood flow in the azygous vein and severe stenosis of the superior vena cava at its intra-pericardial portion. Those findings were compatible with type III superior vena cava syndrome (Figure 3).



Figure 2. Iliac artery aneurysms.







Figure 1. Fibrosis of the superior vena cava just above the atrium.



Figure 4. Anastomosis of spiral saphenous vein graft with left brachiocephalic trunk.



Figure 5. Anastomosis of the vein-to-right atrial bypass.



Figure 6. Outcome after flow liberation by grafting.

After 12 days of thorough investigation, during which the patient was kept in bed rest (with the head elevated at 60°) and received clinical and nutritional support, the decision was made to repair first the aorto-iliac aneurysm. With the patient under general anesthesia and orotracheal intubation, a Zenith endograft was deployed, the ,left iliac aneurysm was embolized and a PTFE femoro-femoral crossover bypass was performed . The procedure went uneventfully. Postoperative period was also uneventful and the lower extremities pulses were bilaterally palpable. The patient was discharged on the 2nd postoperative day, in stable condition. He was scheduled for readmission in two weeks.

On readmission, a left brachiocephalic vein to right atrial bypass was performed with a spiral saphenous vein graft (Figures 4, 5 and 6). The patient developed sternal wound dehiscence in the 7th postoperative day, and was reoperated for sternal re-suture. The patient was placed on oral anticoagulation and discharged from the ICU 14 days after the first procedure, presenting complete regression of signs and symptoms. Since hospital discharge, he has been followed up at the outpatient clinic. ,He has had no with symptoms or complaints, and has been able to perform his daily activities normally. CT angiography at 12 months of follow-up confirmed patency of the thoracic vein graft (Figure 7) and exclusion of the aneuryms, with reduction of the left common iliac artery aneurysm diameter to 4.8 cm in (Figure 8), with no signs of endoleak (Figure 9).

Discussion

Superior vena cava syndrome of benign origin is a rare condition, although increasingly prevalent due to the rising number of endovascular procedures. As shown in this



Figure 7. Follow-up at 12 months - graft patency.



Figure 8. Follow-up at 12 months - resected aorto-iliac aneurysm.



Figure 9. Follow-up at 12 months – reduction in the aneurysm diameter on the left common iliac artery.

paper, the presence of an associated aorto-iliac aneurysm makes this case even more unusual. Regarding the treatment, both conditions may be approached by either conventional surgery or endovascular technique. In cases of malignant origin, endovascular recanalization is clearly the treatment of choice⁵⁻⁹, and surgical repair should be reserved for resectable tumors¹⁰⁻¹³. In cases of superior vena casa syndrome of benign cause, the open surgical approach remains the treatment of choice¹⁴⁻¹⁷, although some authors sustain their preference for the endovascular technique, regardless of the costs of additional interventions and the need for longer follow-up¹⁸⁻²⁰.

In this case, we had a young patient with extensive type III chronic venous thrombosis who was not a candidate to endovascular reconstruction due to the juxta-atrial location of the occlusive lesion. Open surgery is also indicated in patients with less extensive lesions (type I or II), who have not benefited from an attempted endovascular treatment¹.

On the other hand, we realized that the conventional open treatment of the aorto-iliac aneurysm would dramatically increase the surgical risk in this patient. The sequence of interventions in this case is another important issue to be discussed. It is well known that patients with aneurysmal dilatation of the aorta are at an increased risk of rupture during or after an unrelated surgical procedure. Based on this, we decided to treat the aneurysm first. The endovascular technique was chosen in this case aiming at a lesser surgical trauma and faster recovery of the patient, who would be submitted to another intervention shortly. Both procedures were successful and, 12 months later, the patient remains asymptomatic on follow-up.

Conclusion

Based on what has been presented, we conclude that the surgical techniques for the treatment of superior vena cava syndrome of benign origin associated with aorto-iliac aneurysm should be thoroughly discussed, before patient's management aiming at better outcomes. The association of these entities requires deciding the best sequence of interventions. Since this a rare case, a longer follow-up is necessary. One can conclude, however, that, in some situations, the association of endovascular and conventional techniques may clearly benefit the patient.

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Author's contribution

Conception and design: GPS, PEOR Analysis and interpretation: GPS, PEOR, LCRA, RAVM Data collection: GPS, MCTV, DQN, MHL, SLA Writing the article: GPS Critical revision of the article: PEOR, DQN Final approval of the article*: GPS, MCTC, MHL, DQN, LCRA, RAVM, SLA, PEOR Statistical analysis: non applicable

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