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Celiac Trunk Thrombosis as a Presentation of Takayasu Arteritis☆

Trombosis del tronco celíaco como presentación de la arteritis de Takayasu

Takayasu arteritis (TA) is a granulomatous vasculitis that affects large arteries, primarily the aorta and supra-aortic trunks, although medium-sized arteries may also be involved.[1]

We present the case of a 35-year-old woman, with no family history of interest or known cardiovascular risk factors except obesity and oral contraception, who came to the emergency department due to exacerbation of postprandial epigastric abdominal pain that had been progressing for months. Lab work showed evidence of leukocytosis with neutrophilia, and abdominal ultrasound revealed no significant findings. The patient was hospitalized with suspected erosive gastropathy. Gastroscopy demonstrated changes in coloration of the gastric mucosa suggestive of ischemic disease. Subsequently, emergency abdominal CT scan identified complete thrombosis of the celiac trunk, hepatic hypoperfusion and splenic infarction. Diagnostic arteriography was performed and the findings were compatible with the CT scan (Fig. 1a). Due to the high risk of thrombus migration and signs of hypoperfusion of the abdominal organs, endovascular treatment was ruled out, and we opted to perform surgical treatment.

During emergency exploratory laparotomy, we observed generalized hepatic hypoperfusion and splenic infarction, with no other alterations. Thrombectomy was performed (Fig. 2), followed by angioplasty through the splenic artery to subsequently perform splenectomy. Hepatic reperfusion and restoration of flow to the celiac trunk were confirmed by intraoperative Doppler ultrasound.

On the second day of the postoperative period, a follow-up Doppler ultrasound was performed, which showed the absence of flow in the celiac trunk but no clinical repercussions. CT scan confirmed these findings but showed no alterations in the perfusion of the abdominal organs. On postoperative day 15, the patient was discharged from the hospital with home anticoagulation therapy using low molecular-weight heparin after hypercoagulability studies had shown no alterations.

One month later, the patient came to the emergency department for cyanosis of the fingertips of the right hand; corticosteroid and antihypertensive treatment were prescribed. After one month of treatment and persistence of pain along with claudication of the upper right limb, the patient was hospitalized. CT angiography of the aorta and supra-aortic trunks with 3D reconstruction showed wall thickening at the origin of the right subclavian causing a small stenosis in the segment between the clavicle and the first right rib (Fig. 1b).

Given the symptoms, claudication of the upper right limb, and the difference in systolic blood pressure between both arms greater than 10 mmHg, the diagnosis of Takayasu disease was established. Treatment with prednisone and methotrexate was initiated, and the patient's progress was good.

TA is a chronic, idiopathic inflammatory disease that mainly affects women under the age of 40 and children. The annual incidence is estimated at 1.2–2.6 cases per million inhabitants per year in the western population, although it is much higher in Southeast Asia.[1]

Histopathology revealed adventitial thickening, areas of leukocyte infiltration of the tunica media and intimal hyperplasia. This response predisposes patients to the development of stenoses or arterial occlusion.[2]

The spectrum of presentation, severity and rate of progression of the disease can often lead to an inaccurate evaluation and a delay in diagnosis.[3] Clinically, it presents with general symptoms and specific symptoms of vascular involvement (arterial hypertension, carotidynia and vascular murmurs)[1] related to involvement of the celiac trunk in 18% of cases, according to the literature.[3,4]

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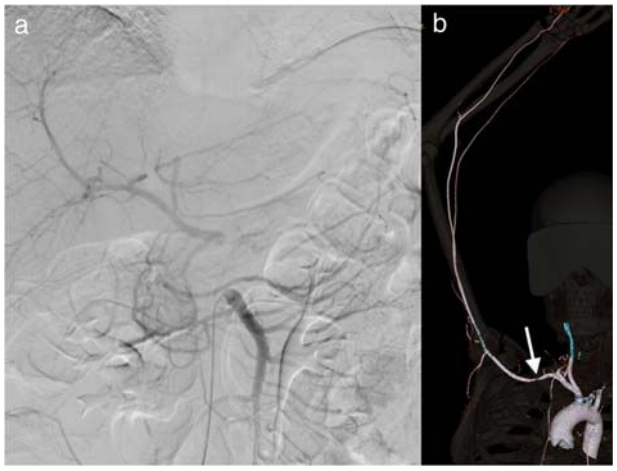

Fig. 1 – (a) Diagnostic angiogram of thrombosis of the celiac trunk and (b) CT angiography with 3D reconstruction showing stenosis of the right subclavian (white arrow).

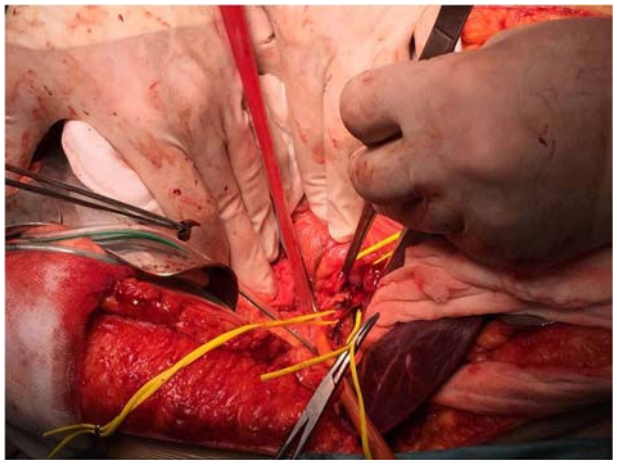

Fig. 2 – Intraoperative image of thrombectomy with the thrombus exiting the splenic artery.

The diagnostic criteria of the American College of Rheumatology are: age of onset before age 40, claudication of one of the extremities, decrease in the pulse of a brachial artery, difference in systolic blood pressure between both arms greater than 10 mmHg, vascular murmur and alterations on imaging tests. Three of these 6 criteria are necessary to establish the diagnosis.[5]

In its treatment, corticosteroids, immunosuppressants, biological agents, antiplatelet/ anticoagulants or vasodilators can be used.[1] The main surgical indications are renovascular hypertension, cerebrovascular diseases, coronary ischemia or vascular claudication.[2]

Chronic thrombosis of the celiac trunk may lead to different clinical manifestations, such as an ischemic syndrome of the celiac region, or it may remain asymptomatic due to the development of collateral circulation.[6] In case of acute occlusion, ischemic necrosis will occur, which will affect to a greater extent those organs with terminal vascularization, such as the spleen in this case, an event described in the literature by Kelekis et al.,[7] among others. Treatment involves revascularization, which can be either endovascular or by surgical bypass.[6]

Splenic infarction is a relatively uncommon diagnosis, and only 10% are diagnosed pre-mortem.[8] The main causes include: thromboembolism, acute infection and hematological disease.[9] There are hardly any cases associated with vasculitis, with a frequency of 2% in the O'Keefe et al. study, and no specifically described events due to TA in the literature.[8,9] Genc et al. report a similar case of splenic infarction associated with thrombosis of the celiac trunk in a context of acute abdomen, although without being associated with vasculitis.[10]

In conclusion, despite the fact that the involvement of the celiac trunk in TA has been described in the literature,[3,4] the case we present is an extremely rare event given its complete occlusion, causing splenic infarction.

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Leiomyosarcoma of the Inferior Vena Cava☆

Leiomiosarcoma de vena cava inferior

Primary leiomyosarcoma of the inferior vena cava (IVC) is a rare mesenchymal tumor with a poor prognosis.[1] Only 300 cases have been reported,[1,2] so the data available to guide us in their treatment are very limited. Detailed case reports case can help make appropriate decisions for the proper management of future patients.

Our patient is a 63-year-old woman with multiple previous abdominal surgeries due to colon adenocarcinoma with liver metastases. The follow-up computed tomography (CT) scan detected a new 27-mm retroperitoneal nodule that was dependent on the posterior wall of the IVC (Fig. 1). After fine-needle aspiration (FNA) biopsy, she was diagnosed with primary leiomyosarcoma of the IVC. The patient was asymptomatic.

Through exploratory laparotomy and after complex adhesiolysis, a solid tumor was evident from the infrahepatic vena cava to the origin of the left renal vein and attached to the right adrenal gland. No invasion of other intra-abdominal structures was observed.

After clamping the retrohepatic vena cava, left renal vein and the confluence of the right renal vein (Fig. 2A), *en bloc* cavectomy was performed with the right adrenal gland. The flow was reconstructed with a 20-mm ringed polytetrafluoroethylene (PTFE) graft, with direct re-implantation of the left renal vein (Fig. 2B). The definitive pathology results confirmed the diagnosis of primary vascular leiomyosarcoma. Resection margins were tumor-free. After 24 h in the intensive care unit, the patient was moved to the floor with no signs of vascular compromise.

Due to intestinal perforation, re-operation was necessary on the third postoperative day. The subsequent patient progress was satisfactory, and she was discharged on the tenth postoperative day with no signs of infection or graft thrombosis on follow-up CT. Seven months later, there are no data of tumor recurrence, thrombosis or prosthetic infection.

Inferior vena cava leiomyosarcoma is a rare, locally very aggressive, malignant tumor with slow growth, which means that they can remain asymptomatic until advanced stages of the disease.[1] The most frequent location is infrarenal.[3]

Currently, *en bloc* surgical resection is the only potential cure. Complete exeresis with free margins increases long-term survival.[4–6]

Different grafts can be used for the reconstruction of the inferior vena cava. However, given the limited number of case reports, there is no consensus about which technique or material is most appropriate.

Some authors advocate venous grafts or primary suture with the aim of reducing the risk of infections or thrombosis.[3,7] However, authors like Michael et al.[6] highlight several technical factors that, even today, are the subject of debate.

When PTFE grafts are used, some suggest the use of ringed grafts to prevent collapse. In contrast, others argue that this type of grafts have worse tissue integration and may predispose patients to the formation of entero-prosthetic fistulae to the duodenum.[1–6] In the case of our patient, we used a ringed PTFE graft as described, which is the most widely used surgical technique, in association with coverage by the greater omentum to prevent the formation of fistula tracts.

In patients with involvement of the middle segment, we must also consider whether to reconstruct the renal veins. In the case of the right renal vein, the need for restoration of flow is inexorable; however, on the left side, the renal vein could be ligated due to the presence of collateral circulation.

In our patient, the tumor reached the origin of the left renal vein. In this case, we opted for direct re-implantation to the PTFE prosthesis in order to reduce the risk of subsequent renal disease that would further increase the morbidity of the procedure.

Recent studies advocate neoadjuvant radiotherapy (RTx) for the management of inferior vena cava leiomyosarcoma. It

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