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Case Report

May-Thurner Syndrome: Case Report and Literature Review

Síndrome de May-Thurner: relato de caso e revisão de literatura

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ABSTRACT: May-Thurner Syndrome (MTS) is a rare clinical condition that is more prevalent in women and consists of compression of the left common iliac vein by the right iliac artery[1]. The objective of this study is to report a case of MTS showing the relation with thrombotic events and perform a literature review. The etiology is categorized as multifactorial and is associated with clinical manifestations related to venous hypertension: pain, edema, varicose veins in the left lower limb, superficial venous reflux, post-thrombotic syndrome and primary lymphedema[3,4].The classification occurs according to the appearance and severity of symptoms, the last stage being the appearance of deep venous thrombosis in the left lower limb, which is most frequently affected[3,6]. The diagnostic approach is initially performed through anamnesis and physical examination, with the request of complementary tests such as computed tomography, magnetic resonance imaging and venography being important[1,3,4]. The treatment does not have a specific guideline, but considering the clinical condition it can be done through anticoagulant compressive stockings, balloon angioplasty or stent placement[7].

KEYWORDS: May Thurner Syndrome; Case Study; Rare Disease; Vascular Malformation; Deep Vein Thrombosis.

RESUMO: A Síndrome de May-Thurner (SMT) é uma condição clínica rara e mais prevalente em mulheres e consiste na compressão da veia ilíaca comum esquerda pela artéria ilíaca direita[1]. O objetivo deste estudo é relatar um caso de SMT evidenciando sua relação com eventos trombóticos e realizar uma revisão de literatura. A etiologia dos sintomas é categorizada como multifatorial e está associada com manifestações clínicas relacionadas à hipertensão venosa: dor, edema, varizes em membro inferior esquerdo, refluxo venoso superficial, síndrome pós-trombótica e linfedema primário[3,4]. A classificação ocorre de acordo com o aparecimento e gravidade de sintomas, sendo o último estágio o surgimento de trombose venosa profunda de membro inferior esquerdo, o qual é mais frequentemente acometido[3,6]. A abordagem diagnóstica é realizada inicialmente por meio de anamnese e exame físico, sendo importante a solicitação de exames complementares como tomografia computadorizada, ressonância magnética e venografia[1,3,4]. O tratamento não possui uma diretriz específica, mas existe um direcionamento para o quadro clínico, assim pode ser feito por intermédio de meias compressivas anticoagulantes, angioplastia com balão ou colocação de stent.[7]

PALAVRAS-CHAVE: Síndrome de May Thurner; Estudo de Caso; Doença Rara; Malformação vascular; Trombose Venosa profunda.

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INTRODUCTION

May-Thurner Syndrome (MTS), also known as Iliac Vein Compression Syndrome (IVCS), is a rare and more prevalent clinical condition in females, especially between the third and fifth decade of life, caused by compression of the iliocaval segment. It is known that the most frequent presentation of MTS is compression of the left common iliac vein by the right iliac artery[1].

MTS was first described in 1851 by Rudolph Virchow, who visualized iliac vein compression and its signs and symptoms, which include venous claudication, venous hypertension, edema, pain, and recurrent deep vein thrombosis. Moreover, in 1956 the pathophysiology of MTS was reported by May and Thurner who showed the relation between hypertrophic changes of the venous intima layer with chronic mechanical stress, caused by the contact of the right common iliac vein over the left common iliac vein against the lumbar vertebra[2].

The etiology of the symptoms of the disease is multifactorial because it involves the expression of multiple genes, in addition to encompassing other clinical manifestations, such as superficial venous reflux, post-thrombotic syndrome and primary lymphedema[3]. In addition, some triggering factors of the syndrome may be previous surgery, prolonged immobility and pregnancy. On the clinical picture is visible the progression of the syndrome with chronic venous insufficiency, through varicose veins and varicose ulcers[4].

OBJECTIVE

The aim of this study is to report a case of May-Thurner Syndrome evidencing the relation with thrombotic episodes and to perform a literature review.

CASE REPORT

In 2023, a 30-year-old female patient was admitted to the ER of the Hospital Geral de Itapecerica da Serra (HGIS) with abdominal pain in the left hypochondrium, nausea, emesis and fever. On physical examination, she presented pain on deep palpation in the right and left hypochondrium. Regarding personal history, one patient reports being a cocaine user, smoker and about the pathological history, she had cavernous sinus thrombosis in 2014 and in 2021 she had splenic, hepatic and intestinal infarction with mesenteric claudication. She reported that she used 5 mg of warfarin.

Considering the hypothesis of splenic venous thrombosis, a computed tomography (CT) scan was requested in order to elucidate the clinical picture of the patient, which showed hepatomegaly with small cystic images and spleen with a hypodense wedge area at the hilar apex compatible with splenic infarction, in addition to a compression of the left common iliac vein by the right common iliac artery.

In view of the multiple thrombotic episodes of the patient and the CT, one of the diagnostic hypotheses can be thought of as May-Thurner syndrome. Thus, the indicated approach was to adjust the dose of warfarin according to International Normalized Ratio (INR) through outpatient follow-up. Regarding endovascular therapeutic management, stent was not indicated, since the patient did not have venous thrombosis in the left lower limb.

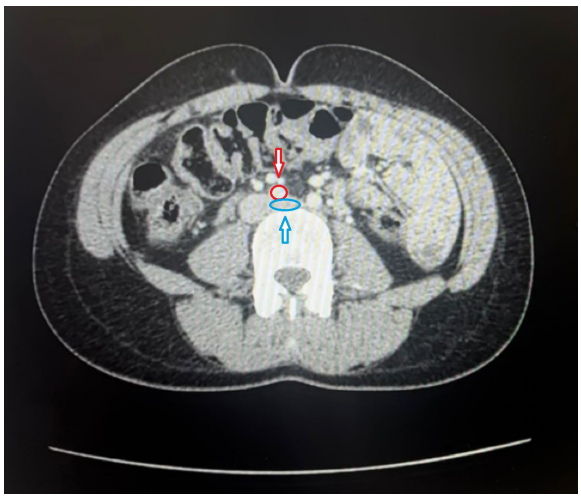

Figure 1 - CT of May-Thurner Syndrome. Compression of the left common iliac vein (blue arrow) by the right common iliac artery (red arrow).

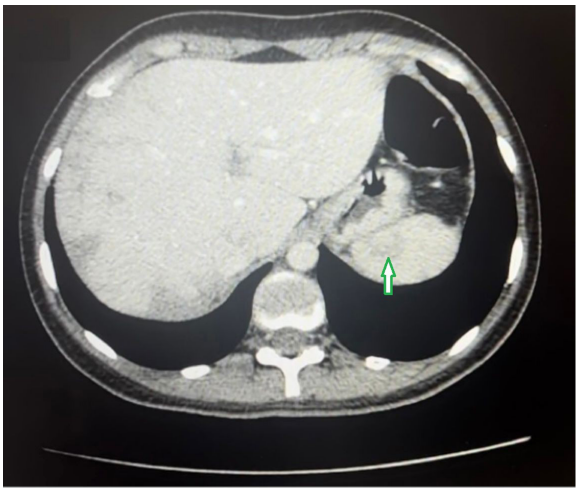

Figure 2 - CT of splenic infarction. Presenting hypodense area (green arrow) and spleen with preserved dimensions.

DISCUSSION

May-Thurner Syndrome is triggered by a compression of the left common iliac vein by the right

common iliac artery and this anatomical alteration causes chronic trauma together with pulsatile forces of the artery, which causes a pinch of the left common iliac vein between the lumbar spine and this may result in endothelial damage. This process leads to a deposition of collagen and elastin, contributing to the obstruction of the region, thrombus formation and predisposition to thrombotic events[5]. In addition, there is a narrowing of the anteroposterior diameter and transverse increase of the right common iliac vein due to compression. Under these circumstances, lead to the occurrence of a stasis of venous return within the veins of the lower limbs[6].

The pathology is classified into three clinical stages: in stage I there is a compression of the left iliac vein without repercussion of symptoms, stage II involves the gradual formation of an intraluminal venous fibrous band and stage III is characterized by a thrombosis of the left iliac vein, as well as deep venous thrombosis (DVT) of the left lower limb, which may be or may be not associated with a pulmonary embolism[3,6].

Compression can cause signs and symptoms of venous hypertension - pain, edema and varicose veins in the left lower limb, which is commonly more affected[1]. With the difficulty in venous return, skin changes related to chronic venous insufficiency arise, such as varicose ulcers, phlebitis, pelvic and/or lower limb varicose veins, which may initiate deep venous thrombosis, and is now called Cockett's Syndrome[1,4].

The etiology is correlated with primary venous insufficiency, because they are nonspecific symptoms that, associated with the rarity of the syndrome, make the etiological diagnosis difficult[4]. However, when suspecting MTS, it is possible to use complementary tests such as Doppler ultrasonography, which is of great relevance to analyze the iliac veins and any sign of compression due to the difference in venous flow velocity when comparing with the opposite lower limb[3]. There are USG findings that can help in the study of the iliac veins, such as venous flow asymmetry, that is, the difference in flow volume greater in the right common iliac vein by 40% compared to the left. In addition, another sign would be the disproportion between the pressure of the inferior vena cava and the iliac veins, which demonstrates the presence of stenosis[4]. However, even though Doppler USG is a good test for analyzing signs of DVT or venous insufficiency, has limitation regarding the observation of stenosis of the pelvic veins, due to the deep anatomical location[1,4].

For this reason, when there is a suspicion of MTS in the complementary examination, either because a region with alteration in turbulent blood flow, stenosis or even the presence of collateral veins was analyzed, it is possible to use computed tomography venography or magnetic resonance venography, since they are imaging exams that allow the identification of some significant findings such as DVT and vascular compression[1,3]. In addition to these options, the gold standard used as an imaging diagnostic method is phlebography with measurement of venous pressures, which provides a morphological study of the stenosis and the degree of hemodynamic compromise[4,3].

In relation to the present case under study, the patient had important hemodynamic repercussions such as cavernous sinus thrombosis, splenic, hepatic and intestinal infarction with mesenteric claudication. And on the CT scan of the patient, it was possible to observe compression of the left common iliac vein by the right common iliac artery, which encompasses the diagnostic hypothesis of MTS.

The treatment aims at resolving the acute thrombotic event and vascular stenosis, as well as preserving valve function. In the past, the use of isolated anticoagulants was recommended, however this practice was flawed because it does not involve the reversal of existing thrombi, that is, the obstructive component. Furthermore, the patient would be at risk of the consequences of the post-thrombotic syndrome, such as persistent edema, pain in the lower limbs and the possibility of recurrence of thrombotic events[3].

Currently, the therapeutic management of SCVI does not contain a specific guideline, however there is a direction of treatment through the appearance of symptoms, presence of DVT, severity and classification of the clinical condition. Thus, in stage I, the conservative method is used with compression stockings; in stages II and III, the need for mechanical or open surgical thrombectomy, balloon angioplasty and stent implantation is evaluated[7]. Endovascular treatment in patients with the occurrence of thrombotic events associated with this pathology began in the last decade with the performance of balloon angioplasty and subsequent placement of the stent, this is considered the first option because it is a minimally invasive procedure, reestablishes blood flow, chemically dissolves thrombi and provides symptomatic improvement to the patient[8].

CONCLUSION

May-Thurner Syndrome is a relevant clinical condition, due to its significant prevalence in women. This pathology causes compression of the left common iliac vein, capable of developing clinical conditions that range from asymptomatic patients to severe hemodynamic repercussions, therefore, early diagnosis and treatment are necessary in the face of significant alterations.

It is reiterated that the diagnosis is made through the anamnesis, physical examination and imaging tests mentioned above to confirm the diagnostic hypothesis. There is still no specific protocol for therapeutic management, however there is a direction according to the classification of the disease, with the possibility of conservative treatment and minimally invasive procedures,

the main endovascular treatment being angioplasty with the use of a balloon and placement of a stent.

In view of the present limitations, such as the lack of a guideline and an image exam with a higher degree of specificity, it is of paramount importance to carry out more scientific studies and research that address this theme, with the aim of improving the quality of patients' lives.

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