Absence of supra-renal inferior vena cava: case report

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Abstract

The congenital absence of the inferior vena cava is a rare and occasional finding at complementary exams or abdominal surgeries. As the majority of patients without other malformations are asymptomatic, it is hard to estimate the prevalence of these anomalies. Our goal was to report a case of a 28-year-old patient, male, referred to thorax X-ray due to high fever state (39°C), nonresponder to medication. The X-ray suggested azygos vein dilatation. The interview about cardiovascular symptoms revealed thoracic pain during both physical exercises and at rest, and occasional dyspnea during sleep related to the period of intensification of physical exercises. Computadorized tomography with and without contrast did not solve the case, what explain the request of an angiotomography, that showed absence of thoracic inferior vena cava.

Keywords: Vena cava, inferior/abnormalities; azygos vein/abnormalities; venous thrombosis/etiology.

Introduction

Inferior vena cava (IVC) is a large-caliber vessel that drains blood from the lower limbs, from most of the trunk, besides abdominal and pelvic organs. It is formed by the junction of the common iliac veins and lies right of the abdominal aorta, draining into the right atrium. The azygous system connects the superior and inferior vena cavae directly or indirectly¹,². Congenital absence of IVC is an occasional finding in imaging exams. Because most patients without other malformations are asymptomatic, it is difficult to estimate the prevalence of such anomaly.

Case report

A 28-year-old male patient was admitted to the Hospital São Sebastião, Maceió (AL), Brazil, with high fever (39°C), not responding to medication, being referred for radiography of the sinus cavities and chest. He had no relevant past medical history. Chest radiography was suggestive of azygos vein dilatation. The fever came down spontaneously after a week, and no infection or other cause was found. After hospital discharge, he was referred to an angiologist in order to investigate the chest radiograph finding. At the Clínica de Especialidades (MedAngio), the patient reported chest pain...
at exercise and at rest, and occasional dyspnea associated with a period of intense physical exercise. Computed tomography (CT scan), first performed without a contrast agent and, later on, with a contrast agent, was inconclusive. Spiral CT angiography was indicated and showed:
1) continuous IVC azygous (Figure 1);
2) no aneurysms or thrombi;
3) retroaortic renal vein.

The patient was then oriented to discontinue heavy physical activities due to the high risk of venous thrombosis. He has been asymptomatic for six months.

Discussion

The IVC is formed between the sixth and eighth weeks of embryonic life in a complex process involving three primitive vein pairs (posterior cardinal, subcardinal and supracardinal veins), which are extensively anastomosed. Such anastomosis recede during the embryologic process, thus originating the IVC. When abnormalities occur during this development, anomalies of IVC such as partial or total agenesis, may occur. The azygous system tends to dilate as a compensatory mechanism.

As these anomalies may be asymptomatic, their frequency is hard to measure. However, their prevalence in the general population is estimated to be 0.5%. About 90% of the cases are related to partial absence of the supra-hepatic segment of the IVC. Total absence of the renal and supra-renal IVC segments is very rare.

IVC anomalies may be accompanied by vascular, cardiac, bowel, portal and liver malformations, and polysplenia. Most cases are asymptomatic and occasionally found by routine exams not related to the malformation. However, in case of insufficiency of collateral veins, blood drainage from the lower limbs and pelvis becomes inadequate, causing venous stasis and a predisposition to venous thrombosis. Bilateral and recurrent deep vein thrombosis (DVT) in the proximal vein of young patients without history of coagulopathies may imply IVC anomaly, so imaging methods are required. About 5 to 7% of the patients with DVT present some IVC anomaly; the incidence is higher in young patients and those with bilateral DVT.

There is no indication for interventional treatment in case of partial IVC absence. However, knowing this entity is important in order to avoid unnecessary surgical procedures. Since this condition may predispose the patient to DVT, some authors recommend anticoagulant therapy, but this is no consensus. In the present case, oral anticoagulation therapy and thrombophilia investigation were not necessary because the patient had an

Figure 1 – (A) Abdominal inferior vena cava; (B) IVC with azygous continuation; (C) azygous vein; (D) cava-azygous transition; (E) azygous vein; (F) hemiazygous vein; (G) azygous vein draining into the right atrium.
anatomical congenital alteration, not an acquired functional one; the only recommendation was to restrict heavy physical activities. Anticoagulant therapy is quite expensive for the patients and, when the disease can be controlled with behavioral measures, patients accept the treatment more easily, besides having their quality of life improved. The patient has been followed for one year and has been asymptomatic, under the cardiovascular point of view.

References


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