

Pyloric Stenosis Revealing Agenesis of the Retrohepatic Inferior Vena Cava: About A Rare Case

A. Jallouli^{1*}, H. Ouaya¹, M. Jarti¹, M. Aouroud¹, A. Ait Errami¹, S. Oubaha², Z. Samlani¹, K. Krati¹

¹Hepato-Gastroenterology Department, University hospital of Mohammed VI, Marrakesh, Morocco

²Physiology Laboratory, Faculty of Medicine and Pharmacy, Cadi Ayyad University, Marrakesh, Morocco

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*Corresponding author: A. Jallouli

Hepato-gastroenterology department, University hospital of Mohammed VI, Marrakesh, Morocco

Abstract

Case Report

Agenesis of the retrohepatic inferior vena cava (IVC) is a rare vascular malformation characterized by agenesis of the retrohepatic segment of the IVC and a dilated azygos vein draining venous blood from the caudal segments. Most patients are asymptomatic if the anomaly is isolated. Congenital heart diseases and asplenia or polysplenia syndromes have been reported in association with this entity. We report through this observation the case of agenesis of the retrohepatic inferior vena cava revealed by a pyloric stenosis.

Keywords: Agenesis of the retrohepatic inferior vena cava, Pyloric stenosis, Azygos continuation.

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INTRODUCTION

Agenesis of the retrohepatic inferior vena cava (IVC) is a rare vascular malformation characterized by agenesis of the retrohepatic segment of the IVC and a dilated azygos vein draining venous blood from the caudal segments.

We report the case of a 46-year-old patient, admitted for etiological workup of a pyloric stenosis with incidental discovery of agenesis of the retrohepatic IVC with azygos continuation.

CASE REPORT

A 46 year old male patient, without any particular pathological history, complains of atypical chronic epigastralgia of moderate intensity, fixed without irradiation, without any aggravating or relieving factor, evolving for 2 years, associated with late postprandial food vomiting of intermittent evolution, without any other digestive or extradigestive manifestations, in particular no dyspnea, no chest pain, evolving in a context of apyrexia and alteration of the general state made of anorexia, asthenia and weight loss.

Clinical examination found a conscious patient, hemodynamically and respiratory stable, with epigastric tenderness, no palpable mass or impaction,

no clinically detectable hepatomegaly or splenomegaly, no flank dullness, cardiovascular examination was also without particularities. The rest of the somatic examination was unremarkable.

A thoraco-abdominal computed tomography (CT) scan showed significant gastric distension reaching the hypogastrum upstream of a very tight pyloric stenosis, associated with a venous return anomaly such as agenesis of the retrohepatic IVC with azygos continuation via a very dilated hemi-azygos vein (Figure 1 and 2).

An upper endoscopy showed a fibrous pyloric stenosis with a small opening that could not be crossed by the endoscope.

The patient was transferred to the surgical department for surgical management of the pyloric stenosis. The procedure consisted of resection of the pyloric stenosis with anastomosis. The post-operative follow-up was without incident,

The patient was then referred to the cardiology consultation for management of the agenesis of the retrohepatic IVC.

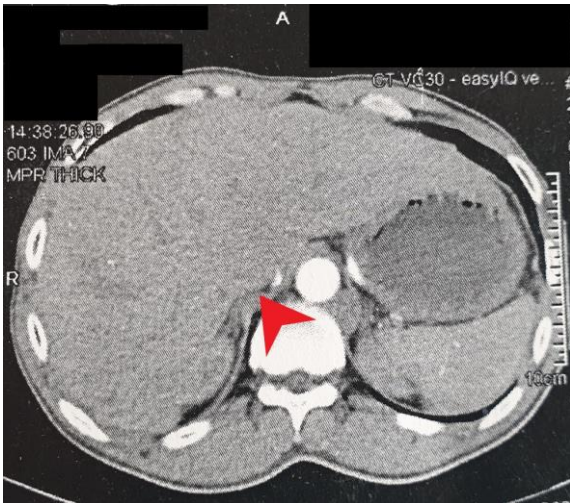


Figure 1: Agenesis of the retrohepatic segment of the IVC

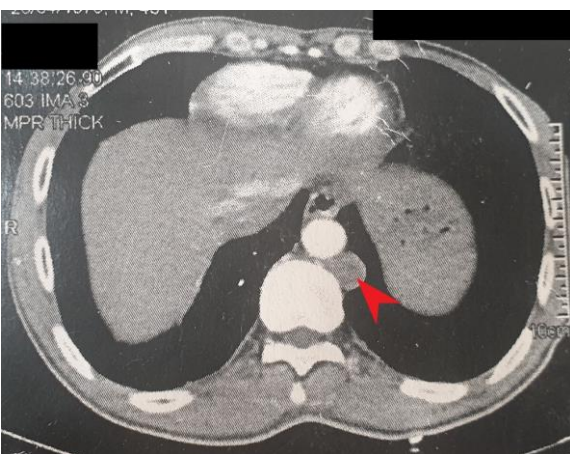


Figure 2: Dilated hemi-azygos vein

DISCUSSION

Agenesis of the retrohepatic segment of the inferior vena cava is the absence of fusion of hepatic and prehepatic segments. It is a rare congenital malformation representing about 0.6% of the general population (Minniti *et al.*, 2002).

In this situation, there are 3 possible routes for venous return to the right atrium (Bass *et al.*, 2000): via the azygos vein to the superior vena cava (SVC), via the left accessory hemizygos and superior intercostal veins to the left brachiocephalic vein, or via the accessory hemizygos and a persistent left SVC in the coronary sinus.

There is an association of this entity with other cardiac and visceral anomalies, such as dextrocardia, atrial septal defect (ASD), atrio - ventricular canal (AVC), situs inversus, polysplenia or asplenia (Weiner *et al.*, 1993).

Although this anomaly may be discovered incidentally in asymptomatic individuals, it may also be diagnosed in the setting of deep vein thrombosis (Schneider *et al.*, 2002).

The diagnosis of IVC agenesis can be confirmed by computed tomography (CT) or magnetic resonance imaging (MRI). Detection of collateral circulation can also be done by angiography (Sneed *et al.*, 2005).

Magnetic resonance phlebography is a safe and reliable contrast-enhanced alternative to conventional phlebography or CT. The procedure is simple and rapid and can be combined with phase-contrast phlebography and flow direction studies (Sneed *et al.*, 2005).

CONCLUSION

Cases of agenesis of the IVC are most often detected incidentally. Recognition of these anomalies is very important and can avoid misdiagnosis as well as problems when planning interventional procedures (e.g. placement of IVC filters) or abdominal surgery.

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