Digestive and Liver Disease 47 (2015) 918–923

Contents lists available at ScienceDirect

Digestive and Liver Disease

journal homepage: www.elsevier.com/locate/dld

Progress Report

Portal vein aneurysm: What to know

Andrea Laurenzi[a,*], Giuseppe Maria Ettorre[a], Raffaella Lionetti[b], Roberto Luca Meniconi[a], Marco Colasanti[a], Giovanni Vennarecci[a]

[a] Division of General Surgery and Liver Transplantation, S. Camillo-Forlanini Hospital, Rome, Italy
[b] Division of Hepatology, "L. Spallanzani" National Institute for Infectious Disease, Rome, Italy

ARTICLE INFO

Article history:
Received 5 February 2015
Accepted 8 June 2015
Available online 19 June 2015

Keywords:
Portal vein aneurysm
Portal vein dilatation
Visceral venous aneurysm

ABSTRACT

Portal vein aneurysm is an unusual vascular dilatation of the portal vein, which was first described by Barzilai and Kleckner in 1956 and since then less than 200 cases have been reported.

The aim of this article is to provide an overview of the international literature to better clarify various aspects of this rare nosological entity and provide clear evidence-based summary, when available, of the clinical and surgical management.

A systematic literature search of the Pubmed database was performed for all articles related to portal vein aneurysm. All articles published from 1956 to 2014 were examined for a total of 96 reports, including 190 patients.

Portal vein aneurysm is defined as a portal vein diameter exceeding 1.9 cm in cirrhotic patients and 1.5 cm in normal livers. It can be congenital or acquired and portal hypertension represents the main cause of the acquired version. Surgical indication is considered in case of rupture, thrombosis or symptomatic aneurysms. Aneurysmectomy and aneurysmorrhaphy are considered in patients with normal liver, while shunt procedures or liver transplantation are the treatment of choice in case of portal hypertension. Being such a rare vascular entity its management should be reserved to high-volume tertiary hepato-biliary centres.

1. Introduction

Portal vein aneurysm (PVA) is an unusual vascular dilatation of the portal vein, which was firstly described by Barzilai and Kleckner [1] in 1956 and since then less than 200 cases have been reported in the literature, mainly as case reports or small surgical series. PVA discovery is progressively rising, due to the increased number of abdominal imaging procedures. Nowadays, the aetiology and management are not clearly understood and there are no clear guidelines on surgical indications.

The aim of this article is to provide an overview of the international literature to better clarify various aspects of this rare nosological entity and provide a clear evidence-based summary, when available, of the clinical and surgical management.

A systematic literature search of the Pubmed database was performed for all articles related to Portal Vein Aneurysm. All articles from 1956 to 2014 were collected. The search terms used in Pubmed consisted of: portal vein aneurysm OR portal vein dilatation OR visceral venous aneurysm AND (English [lang] OR French [lang].

All the articles regarding PVA were collected, including case reports, small series and reviews. Duplicates and articles concerning isolated splenic and/or mesenteric vein aneurysms were excluded.

2. Definition and aetiology

Overall 96 reports were identified, including 190 patients presenting a PVA [1–95].

PVA is defined as a portal vein diameter exceeding 19 mm in cirrhotic patients and 15 mm in normal livers (Figs. 1 and 2). This difference in size, according to the underlying liver status, is due to an extensive vascular study performed by Doust and Pearce in 1976. Among 53 patients examined through abdominal ultrasound, they noticed that maximum antero-posterior diameter of the portal vein was 19 mm in cirrhotic patients and 15 mm in patients with normal livers. Since then, all portal veins diameters exceeding such measures were considered pathological [96].

PVA is a rare visceral venous aneurysm with an incidence of 0.06% [10] and it represents less than 3% of all visceral aneurysms

* Corresponding author at: Division of General Surgery and Liver Transplantation, S. Camillo-Forlanini Hospital, Circ.ne Gianicolense 87, 00151 Rome, Italy. Tel.: +39 06 55170597; fax: +39 06 58704441.
E-mail address: a.laurenzi@me.com (A. Laurenzi).

http://dx.doi.org/10.1016/j.dld.2015.06.003
1590-8658/

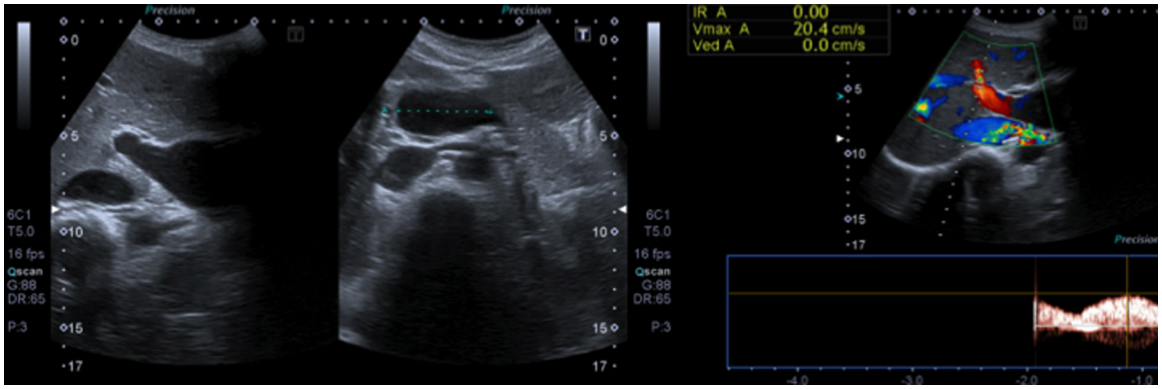

Fig. 1. Colour Doppler abdominal ultrasound of a portal vein aneurysm.

[61]; presently it is more frequently discovered due to the increased number of abdominal imaging procedures.

Aetiology of PVA is still not clearly defined, however it is considered to be either congenital or acquired. A proposed cause of the congenital variant is the incomplete regression of the right primitive distal vitelline vein.

It is well known that during the embryonic phase, the development of the portal vein is related to the involution of the interconnections that exist between the right and left vitelline veins around the duodenum. In such cases, there is an incomplete regression of the distal vitelline vein or a variant branching pattern that leads to the formation of a diverticulum which can evolve into a venous aneurysm [35,97].

The anomalies may arise from the right anterior segmental portal vein or the right anterior and posterior segmental portal veins, which originate from the umbilical portion. The presence of a PVA located at the umbilical portion or on the left portal vein may derive from a rightward deviation of the umbilical portion [41].

Moreover, the presence of vein wall defects can facilitate the development of the aneurysm. The congenital theory is supported by the cases of PVA in children not presenting portal hypertension [4,5,13,26] and from the in utero diagnosis of PVA [27].

The main cause of the acquired version is portal hypertension in liver cirrhosis [69]; in these cases the portal vein dilatation is caused by the high splanchnic flow and hyperdynamic circulation with a consequential weakening of the venous wall. Other causes of PVA are severe pancreatitis [76], trauma [43] and invasion of the portal vein by malignancy [39].

PVAs are usually localized at the level of main portal trunk, portal bifurcation and intrahepatic portal branches. Main patient characteristics reported in the literature are shown in Table 1.

3. Diagnosis

One third of patients are usually asymptomatic as PVA is an incidental finding, while approximately 50% of patients present with non-specific abdominal pain. Gastrointestinal bleeding, portal hypertension or symptoms related to the compression of adjacent organs (i.e. abdominal swelling, jaundice) occur in less than

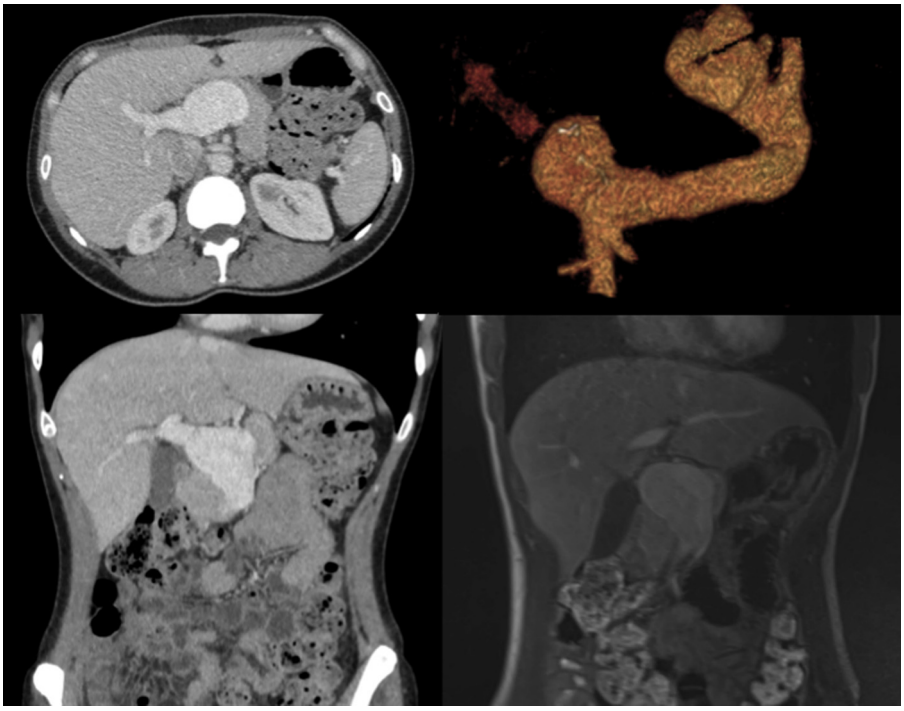

Fig. 2. Computed tomography scan of a portal vein aneurysm in an axial view (top left), 3D reconstruction (top right), coronal view on computed tomography scan (bottom left) and on magnetic resonance imaging (bottom right).