Noncirrhotic Portal Hypertension Secondary to Portal Vein Aneurysm

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September 16, 2022

Abstract

We present a case of symptomatic, non cirrhotic portal hypertension secondary to Portal Vein Aneurysm of 3.7 cm diameter. It presented as dyspepsia. It was diagnosed during an ultrasound colour doppler. Medical therapy was prescribed with close follow up of the patient.

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Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

Introduction

Portal vein aneurysm (PVA) is a rare, vascular malformation defined as a saccular of fusiform dilatation that exceeds the normal calibers. PVA is defined as a portal vein diameter greater than 1.9 cm in cirrhotic patients and 1.5 cm in patients without liver disease. The reported incidence of 0.43% makes PVAs exceedingly rare, making up less than 3% of all venous aneurysms. The low incidence presents clinicians with problems of forming management guidelines acceptable for use. The etiology of disease is not well defined in literature and is presumed to be congenital or acquired. Complications of PVA include portal hypertension, vein rupture, and thrombosis.

Case Report

A 40-year-old female was admitted to our clinic with 3 months of dyspepsia. Other complaints included reflux symptoms and intermittent epigastric pain. The patient previously underwent upper endoscopy with findings of grade 2 esophageal varices (Figure 1) and erythematous pangastritis. Biopsy of the antrum showed active chronic gastritis with no atrophy or metaplasia, and a positive *H. pylori* for which she was treated. On admission, the spleen was palpable 2 finger breadths under the costal margin. Other signs of chronic liver disease and portal hypertension were absent on physical examination. Family history was negative and no offending medications were found. The laboratory findings on admission are shown in Table 1.

Additional lab tests and imaging were ordered to investigate a cause of her possible cirrhosis. Extensive evaluation at our center was completed to rule out any possible chronic liver diseases. Hepatitis A, B, C serologies, ANA, ASMA, anti–LKM, AMA, MPO serologies were all negative. SPEP and UA were normal. Transient elastography result was 5.5 kPa.

Initial abdominal US with Doppler in an outside hospital showed normal shape, size and consistency of liver; an enlarged spleen measuring 18.4 cm with a dilated splenic vein radius of 1.3 cm with an open lumen, normal flow and no thrombosis. Normal portal vasculature, flow and no ascites were seen. In our institution, Color Doppler showed turbulent flow in a PVA, without thrombosis (Figure 2A). Liver parenchyma was normal (Figure 2B). Abdominal CT angiography with contrast displayed a main portal vein diameter of 14 mm, and a 3.7 cm aneurysmal dilatation was detected in the 5 cm section of the posterior segment of the right portal vein (Figure 3A). The anterior segment of the right portal vein was also dilated with a radius of 1.2cm. Vascular collaterals were seen in perisplenic and peripancreatic areas. There were no thromboses seen in vascular structures. Abdominal MRI and CT showed the same 3.7 cm aneurysm detected in the posterior segment of the right portal vein (Figures 3B, 4A, 4B). The patient was followed up clinically and prescribed propranolol, nonselective beta-blocker therapy, with the diagnosis of non-cirrhotic portal hypertension secondary to portal vein aneurysm.

Table 1: Laboratory Results of the patient.

Blood Tests	**Results**	**Reference Range**
White Blood Cell (per $10^3\mu$l)	5.9	4.1-11.2
Hemoglobin (g/dl)	13.9	11.7-15.5
Platelet count (per $10^3\mu$l)	125	160-390
ESR (mm/h)	7	0-20
C-reactive protein (mg/L)	2.4	0-5
Glucose (mg/dL)	98	70-100
Creatinine (mg/dL)	0.8	0.7-1.4
Alanine aminotransferase (U/L)	8	5-45
Aspartate aminotransferase (U/L)	17	5-42
Alkaline phosphatase (U/L)	42	35-105
LDH (U/L)	172	135-250
GGT (U/L)	9	5-85
Albumin (g/dL)	4.8	3.2-5.5
Total Bilirubin (mg/dL)	0.96	0.2-1.2
Direct Bilirubin (mg/dL)	0.34	0-0.3
Indirect Bilirubin (mg/dL)	0.62	0.1-0.5
Ferritin (ng/ml)	25.7	13-150
Ceruloplasmin (mg/dL)	40	16-45
Alpha-1-Antitrypsin (g/L)	1.89	0.9-2
Vitamin B12 (pg./ml)	334	191-663
Folate (ng/ml)	7.8	3.1-17.5
Thyroid stimulating hormone (mIU/L)	3.2	0.27-4.2

Blood Tests	Results	Reference Range
Free-T4	16.5	12-22
Gamma globulin (%) (g/dl)	15.3	9.69-18.9
International Normalized Ratio	1.11	0.85-1.2

Discussion

PVA is an extremely rare finding divided into intrahepatic and extrahepatic location. Of the 190 cases of PVA reviewed in literature, only 38% were classified as intrahepatic [1], seen in our patient. The values required for PVA diagnosis are a diameter measuring greater than 0.7cm in normal patients and 0.85cm in cirrhotic patients [2]. The average diameters of extrahepatic and intrahepatic PVA are described as 30mm and 25.5mm, respectively [3]. A 37mm intrahepatic PVA diameter was noted in our patient, making it one of the largest intrahepatic PVA described in literature.

The rising incidence is likely due to the availability of diagnostic imaging techniques [1]. It was first described in 1956 by Barzilai and Kleckner [2]. Since then less than 200 cases have been seen in the literature [1]. Incidence values of PVAs were found to be 0.43% in a retrospective study of more than 4000 patients undergoing routine abdominal contrast – enhanced CT, of which 28% of patients were found to be symptomatic [3]. Diagnosis of a PVA is often incidental with the preferred imaging modality being a Color Doppler, likely due to the lack of radiation and low cost. Abdominal CT and MRI are rarely used for initial diagnosis, rather for precise localization of the lesion. To our knowledge, all reported PVA's have been visualized using Doppler US, regardless of imaging modality used for diagnosis. None of the cases reported mention absence of PVA on Doppler US, which was initially seen in our patient during her first Color Doppler US conducted at an outside hospital. The second Color Doppler US done in our institution showed a 3.7cm Portal Vein Aneurysm. (Fig X) The results of Doppler US may vary depending on the experience of the person and the technical features of the device. This stresses the importance of evaluating patients with portal hypertension using CT angiography with contrast, or MRI, confirming the diagnosis.

The etiology of PVA is not clearly defined, likely due to the small amount of cases, although it is categorized into either congenital or acquired. Supporting the congenital cause of PVA is the diagnosis of an in-utero PVA via ultrasonography [4], as well as the presence of PVAs in children without portal hypertension [5]. The proposed mechanism is due to the incomplete regression of the right primitive distal vitelline vein [1], which leads to the development of a diverticulum subject to enlargement as portal vein pressures increase [6]. Other suggested congenital factors include an inherent portal vein wall weakness due to developmental defects [7], which in pair with increasing portal pressures leads to dilatation. The acquired form of PVA mainly result from portal hypertension, seen in cirrhosis, is the most common cause [6], with other causative factors being inflammatory processes such as pancreatitis and invasive malignancies. In our patient, the transient elastography that was evaluated using the FibroScan device was completely normal, therefore the liver parenchyma of our patient was normal.

Presenting features of PVA are relatively nonspecific. Abdominal pain is the most common symptom, which was noted in our patient, and is present in approximately half of the patients [1]. Asymptomatic cases are seen in approximately 1/3 patients. Reported complications range from the consequences of portal hypertension (varices, ascites, splenomegaly), compression of adjacent structures (duodenum, inferior vena cava, common bile duct) due to large extrahepatic aneurysms, as well as aneurysmal rupture and thrombosis [1].

The management of PVA varies in regards to the size, location (intrahepatic or extrahepatic), presence/absence of compressive features, thrombosis, and presence of symptoms. Due to the limited number of published cases, no algorithms or guidelines have been established. Current evidence suggests that asymptomatic PVA without thrombosis or the other aforementioned complications be treated conservatively with serial ultrasound imaging every 6-12 months in order to monitor progression [8, 9, 10, 11, 12]. The majority of

PVA are stable and have a low risk of complications with studies showing 88% of patients with no progression [10,13]. Surgical interventions are commonly indicated for complicated cases of PVA: symptomatic patients, progressing aneurysms, rupture and complete thrombosis [14]. PVA with acute PVT has been managed with anticoagulation therapy in non – cirrhotic patients [1,9], although if no clinical improvement is seen, thrombolysis and suction thrombectomy are indicated [15]. Moreno et al. considers a surgical approach to treatment for patients with PVA >3cm without thrombosis, [16] in-order to prevent further complications. Our patient with a symptomatic PVA larger than 3.7cm was managed medically due to being a poor surgical candidate, and due to high post-operative mortality rates (17.5%) in previous reports. Follow up of patients undergoing PVA surgery due to complications was poor resulting in further issues of management [1].

Conclusion

PVA are rare vascular malformations with many uncertainties in management. Incidence of disease is rising; thus, clinicians and radiologists must be able to recognize and identify this disease. PVA can mimic solid, cystic and hypervascular lesions which can lead to incorrect therapy [9]. Duplex ultrasonography is the diagnostic of choice, although CT and MRI should be considered as supplementary in all cases, to determine the extent and location as well as confirm the presence of PVA in cases where US is inconclusive. Evidence based recommendations are absent due to the scarcity of cases and the majority of them being small series or isolated cases. Therefore, the majority of cases are treated on a case-by-case basis, due to comorbidities, risk factors and symptoms. Conservative management is used in the majority of patients, with surgery being reserved for select cases with severe complications. PVA in non-cirrhotic patients with portal hypertension can be managed with nonselective beta-blocker therapy and endoscopically if necessary, preventing the complications of portal-hypertension on PVA.

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