EXTRA HEPATIC PORTAL VEIN ANEURYSM –
A RARE CASE REPORT

RAMEGOWDA.R.B¹, PHANI KONIDE², JAGANNATHA KARNALLI³, VINOD REDDY BAIRI⁴,
SINDHU BR⁵, VIMALA SHESHADRI IYENGAR⁶.

¹Assistant professor, Department of Medicine, ²³rd year Resident, Department of Medicine,
³Professor, Department of Medicine, ⁴1ST year Resident, Department of Medicine,
⁵1ST year Resident, Department of Medicine, ⁶Professor, Department of Medicine,
Adichunchanagiri Institute of Medical Sciences Balagangadharanatha Nagara,
Nagamangala, Mandya-571448, Karnataka, India.

Introduction
Portal venous aneurysm (PVA) is a term used to describe aneurysms of the portal vein (PV),
superior mesenteric vein (SMV), and splenic vein (SV) in the region of spleno-portal junction. They are rare with less
than 200 cases reported in the literature since it was first described in 1956. Although it is being identified more
frequently, the aetiology, natural history and management choices are still relatively unclear.¹ Herein we
present a case of an incidental portal venous aneurysm.

Case report –
A 70 year old elderly male patient who is not a known diabetic or hypertensive, ex smoker not an alcoholic
presented with the history of easy fatigability and palpitations for 15 days. He did not have any history of jaundice,
fever, chills, liver disease, gastrointestinal bleeding.

On examination patient was anemic and there were no signs of chronic liver disease or portal hypertension.
Abdominal examination was unremarkable.
Investigations revealed microcytic hypochromic anemia of HB 6.8gm % and metabolic panel including liver function tests were within normal limits. There was no occult blood in the stool and upper GI endoscopy was normal. USG abdomen showed liver normal in size measuring 14.4 cm normal in shape and echotexture, with dilated main portal vein measuring 18.7mm showing turbulent flow on applying Doppler. There was no evidence of thrombosis in portal veins. F/S/O of aneurysmal dilatation of main portal vein and its right and left branches at the porta-hepatis. CBD was normal, mild splenomegaly and right renal cortical cyst. CECT abdomen revealed liver unremarkable and prominent portal vein measures 17.3 mm in diameter with prominent right and left branches. No evidence of any thrombosis or collaterals formation. (Fig 1 &2)

3. Discussion

Primary venous aneurysms are much less common compared to arterial aneurysms. However, reported incidence of PVA has increased in recent years due, most likely, to the increased availability of advanced imaging techniques in clinical practice. They represent approximately 3% of all venous aneurysms with a reported prevalence of 0.43%. First reported by Barzilai and Kleckner, there are now over 170 cases described in the literature. The majority of these are located in the main extra-hepatic PV with aneurysms of the SV-SMV confluence. PVAs may be either congenital or acquired but the exact etiology still remains controversial and often difficult to determine. Portal hypertension secondary to chronic liver disease is considered the most
common cause of acquired portal vein aneurysms. Portal hypertension leads to intimal thickening with compensatory medial hypertrophy. Progressive replacement of the medial hypertrophy by fibrous tissue weakens the tensile strength of the venous wall making it more susceptible to aneurysmal dilatation. Other causes described in the literature include pancreatitis, trauma and previous surgical intervention. However, for many cases, there is no obvious underlying disease process attributable as the cause of PVA. Embryologically, the portal venous system develops from the vitelline and umbilical veins, which are responsible for venous drainage of the intestine. Supporters of a congenital etiology of portal vein aneurysms suggest that failure of a complete regression of the right vitelline vein may leave a diverticular remnant that can subsequently enlarge to form a saccular aneurysm later in life. A report of an in-utero diagnosis of portal vein diverticulum lends support to this theory. Other hypotheses attribute formation of portal vein aneurysms to an inherent weakness in the venous wall.

Several US and autopsy studies have shown that PV size varies considerably in normal population. A study by Doust and Pearce reported a maximum diameter of 1.5 cm in normal patients rising to 1.9 cm in cirrhotic patients. An anechoic cyst-like lesion near the portahepatis is the characteristic finding on B-mode ultrasonography with Doppler or colour flow studies helping to confirm the diagnosis by showing blood flow through the lesion, thus differentiating it from a liver cyst. Contrast enhanced CT and Magnetic Resonance Angiography (MRA) are useful adjuncts in patients with equivocal US findings or for anatomical characterization of the aneurysm when planning for surgical interventions.

The most common presentation of portal vein aneurysms is abdominal pain (44.7%), followed by incidental detection on CT and US (38.2%), with a minority of patients presenting with gastrointestinal bleeding (7.3%). Complications of PVA include thrombosis, biliary tract obstruction, inferior vena cava obstruction, and duodenal compression. On the whole PVAs are stable and have a low risk of complications with 88% of patients showing no progression of aneurysm size or complications on subsequent follow up scans. Indeed, there is a recent report of a 6 cm saccular aneurysm undergoing spontaneous complete regression over many years of follow up. Rupture of a PVA has been reported in three cases, of which one patient died. Symptomatic or expanding aneurysms are generally considered indications for surgical intervention. The type of surgical intervention depends on associated features. Patients with portal hypertension and portal vein thrombosis have usually undergone shunt surgery. This surgery aims primarily to decompress the portal venous system rather than treat the aneurysm itself although reduced pressures could possibly prevent progressive dilatation of the aneurysm. For patients without portal hypertension, the operation of choice is aneurysmorrhaphy. By resecting out redundant parts of the venous wall and re-suturing, portal circulation is preserved and laminar blood flow restored thus reducing the risk of future thrombosis.

The natural history of PVAs is not fully understood and there is no consensus for their management. PVAs should initially be assessed by colour Doppler ultrasonography with CT scans reserved for indeterminate ultrasound scans and symptomatic patients. CT scans would serve a dual purpose to both rule-out any other concurrent pathology and to help delineate anatomy. Patients that are asymptomatic could be managed conservatively with serial US imaging. Symptomatic patients with severe abdominal pain, symptoms of pressure effect, or with expanding aneurysms and/or complications such as thrombosis or rupture would require surgical intervention. If the aneurysm is associated with portal hypertension, portal-venous shunts should be considered in the first instance. For all other aneurysms without concomitant portal hypertension or cirrhosis, primary aneurysmorrhaphy should be attempted. Patients with thrombosis that extends from the main portal vein into the splenic or superior mesenteric veins should also undergo thrombectomy if possible. The management of large aneurysms in asymptomatic patients is still controversial and should be decided on a
case-by-case basis with discussion with the patient about the risks and benefits. Further studies are required to assess the long-term risk of complications in this population group.¹

In summary, extra-hepatic portal vein aneurysms are a rare occurrence but one that is becoming more widely recognised with the increased use of CT and MRI in clinical practice. The portal vein aneurysm in our patient is likely to be congenital in origin, as he had no signs of liver disease or portal hypertension. Our management choice of expectant treatment is in keeping with the evidence in the literature.

References
