Open ligation of a right portohepato venous shunt: A case report

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SUMMARY

Intrahepatic portosystemic venous shunts (IPSVS) are incidentally diagnosed and generally asymptomatic. Treatment is usually indicated if patient develop symptoms. We present a case report on a patient with symptomatic IPSVS and our surgical approach on the management of IPSVS. The highlight of this discussion is to demonstrate the benefits open surgical approach to treat IPSVS when IR facility is not available.

INTRODUCTION

Intrahepatic portosystemic venous shunt (IPSVS) are rare abnormal blood vessel connections of the intrahepatic vein with branches of the portal vein or inferior vena cava (IVC) (1). The shunts are described in five morphological types (2). A single large vessel of constant diameter connecting the right portal vein directly to the (IVC) is regarded as Type 1. Type 2 is a localised peripheral shunt with single or multiple communications found between peripheral branches of portal and hepatic veins in one hepatic segment. For Type 3, an aneurysmal connection is formed between the peripheral portal and hepatic veins. Type 4 is regarded as multiple communications between peripheral portal and hepatic veins diffusely in both lobes. Finally, Type 5 is described as patent ductus venosus. In this case report, we present a case of IPSVS type 3 treated via an open surgical ligation method.

CASE PRESENTATION

We have a 40-year-old Indian female with a background of dyslipidaemia, and chronic Hepatitis B, presented with right hypochondrium pain for two weeks. Patient's condition worsened four months later, developed cardiac failure symptoms, reduced effort tolerance, bilateral lower limb oedema, New York Heart Association (NYHA) Class II and sleep disturbance, West Haven Grade I. These symptoms have affected her work. Clinically, she was afebrile, with a blood pressure of 110/ 72 mmHg, heart rate of 80 bpm and examination showed mild right hypochondrium tenderness. There were bilateral lower limbs pedal oedema up to mid shin.

INVESTIGATIONS

Blood investigations showed total bilirubin level of 18 umol/L, Alkaline Phosphatase (ALP) 80 U/L, Alanine Amino Transferase (ALT) 55 U/L and Aspartate Amino Transferase (AST) 43 U/L. Serum ammonia was 40 umol/L. Other blood parameters such as full blood count, renal profile, coagulation profile and serum amylase were normal. Ultrasound of the hepatobiliary system showed a portal vein aneurysm. Contrasted Tomography (CT) multiphase imaging of the liver demonstrated a finding of fatty liver, and communication between the right anterior sectoral branch of portal vein and distal branch of middle hepatic vein (MHV) (Figure 1). Echocardiogram showed left ventricular ejection fraction of 65%, the size of all heart chambers normal, with mild mitral regurgitation. Oesophagogastroduodenoscopy (OGDS) were normal as well.

We have decided for an open ligation of the shunt instead of interventional radiographic approach due to financial reasons. The patient was explained regarding the procedure, risk and benefits followed by an informed consent.

TREATMENT

Intra-operative ultrasound color doppler flow was carried out to identify the fistulous communication of the middle hepatic vein and anterior sectoral branch of the right portal vein. The site of fistulous communication was marked with diathermy on the liver surface. The segment VII was dissected with Cavitron Ultrasonic Surgical Aspirator (CUSA) until the fistulous communication was identified (Figure 2). The fistula is subsequently ligated with Hem-o-lok clips and divided between ties and prolene 4/0 sutures reinforcement. Post ligation, ultrasound doppler pressure wave forms were absent over portal vein branches and hepatic vein tributaries confirmed dissociated system.

OUTCOME AND FOLLOW-UP

Patient recovered well post operation. There were no peri-operative complications. There was transient rise in ALT (379 U/L) and AST (415 U/L), however ALP, bilirubin, renal profile, full blood count, and coagulation profile remained normal. She was discharged well three days post surgery. Follow-up two months later, patient's symptoms markedly improved. Her ALT was 82 U/L, AST was 48 U/L, with normal ALP and bilirubin levels. A repeat CT assessment showed complete resolution of the fistula (Figure 3).

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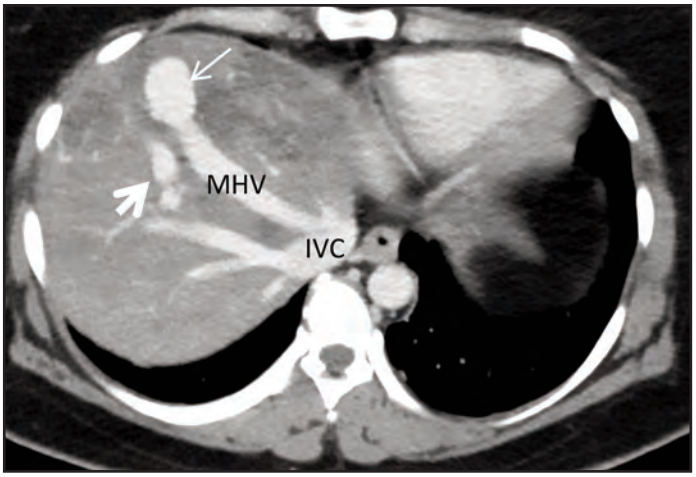

Fig. 1: CT Axial view. Presence of the shunt (thin arrow) connecting the MHV and the anterior sectoral branch of right portal vein (thick arrow). MHV - middle hepatic vein, IVC - inferior vena cava

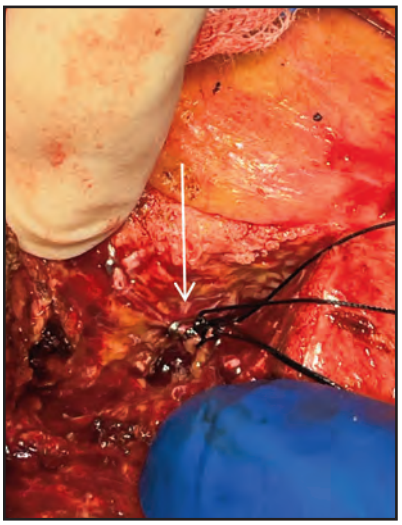

Fig. 2: Ligation of the IPSVS (arrow) using Hem-o-lok clips and Prolene 4/0 suture ties

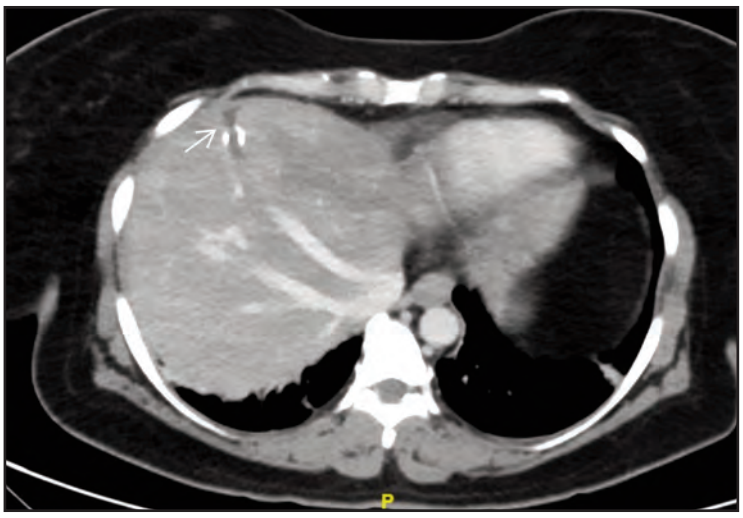

Fig. 3: Complete disconnection of the shunt (arrow) based on repeat CT assessment 2 months post-surgical ligation

DISCUSSION

Intrahepatic portosystemic venous shunt can either be congenital, acquired or iatrogenically formed for therapeutic purposes. At the present literature, it is still an extremely rare disease with an incidence of 1 in 30,000 to 1 in 50,000 for congenital, but still unknown for acquired.[3]

The pathophysiology of IPSVS centers on an abnormal vascular connection within the liver that diverts the blood from the portal vein to the systemic (hepatic) veins without passing through the intervening capillary (sinusoids) beds of the liver parenchyma. In acquired, theories have suggested the role of portal hypertension in cirrhotic liver that resulted with expansion of collateral circulation may cause the formation of a IPSVS which is usually located peripheral vessels of the liver, however in non-cirrhotic liver, it is still unknown, although there are theories in trauma-related which may have contributed from absorption of injured necrotic liver in combination with open inflow and drainage of adjacent vessels, creating a direct connection between the vessels, but are commonly reported as hepatic arterio-portal fistula.[4-6] For congenital IPSVS, it was thought to be persistence communication of cranial and caudal hepatic sinusoids formed by vitelline and umbilical veins.[7] Although this patient has chronic hepatitis B, she does not exhibit symptoms of portal hypertension and routine hepatobiliary system ultrasound, nor intraoperative examination of liver did not show any evidence of cirrhosis. The etiology of her condition is still not known, although suspecting it could possibly be congenital.

Most of the cases are asymptomatic which are diagnosed incidentally during routine imaging.[8] Symptoms of IPSVS are hepatic encephalopathy (50% of cases reported) which may sometimes be mistakenly diagnosed as having psychiatric disorder, hepatic dysfunction, cardiac dysfunction and renal dysfunction.[9] However, in children, hyperammonia was the common clinical manisfestation (85%), followed by jaundice (80%), cholestasis and or hypergalactosemia.[3]

Treatment is indicated in symptomatic patients.[5, 8-10] Our patient has developed symptoms relating to cardiac failure and hepatic encephalopathy and thus, a definitive treatment is indicated for her to prevent worsening of symptoms. Whereas for asymptomatic patient, studies have suggested to calculate the shunt ratio, which is calculated using the Doppler US, by dividing the blood flow volume at the shunt orifice by the total portal blood volume. The risk of symptomatic IPSVS increases proportionately with shunt ratio. If the shunt ratio exceeds 60%, treatment is indicated even if patient is asymptomatic.[7-9]

Majority IPSVS are reportedly treated via interventional radiological (IR) approach as this is the current widely adopted approach.[3-5,9] These shunts can be treated using embolisation principles. The choice of embolisation approach can either be embolic agents or embolic materials. Surgical approaches would be either surgical occlusion, ligation of the shunts, or hepatic resection.[5] Although, there have been reports of perioperative poor effectiveness, especially in cirrhotic liver patients. There was also observation study that have showed newly formed shunts at other locations.

Zhang et. al compared the efficacy and safety of surgical ligation versus endovascular embolisation for congenital extrahepatic portosystemic shunt (EPSVS).[11] Patients in both arms had significant clinical improvements with no recurrence after three years of follow-up. Serum ammonia returned to normal levels within six months. It had shown that both methods are equally effective, with intervention group showing shorter procedure time and less intraoperative blood loss, whereas in the surgical arm, ligation is feasible with safety and may avoid future recurrence.

A recent literature used the measurement of occluded portal pressure as guide to determine whether to perform staged or complete ligation of the shunt.[3] Using occluded portal pressure indicator of 26 cmH_2O, staged ligation is performed if the patient developed portal hypertension as evidence by portal pressure is > 26mmHg, and complete ligation is done if the portal pressure is < 26 cmH_2O. The importance of performing pre-operative occlusion portal pressure measurement is to evaluate the plasticity of the inthrahepatic portal vein. This helps to reduce the risk of post-operative portal hypertension, portal vein thrombosis and even mortality.

Our case reinforces that open surgical ligation can be performed safely with favourable outcomes, making it a valuable option in resource-limited centres.

CONCLUSION

IPSVS are incidentally diagnosed and are generally asymptomatic. Treatment is usually indicated for symptomatic patients and treatment is mostly via IR approach. However, a classical open surgical approach has shown beneficial symptomatic relief with no complication. This serves as a good alternative treatment to IR method when the expertise, cost & logistics are not feasible to perform.

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DECLARATIONS

This case report has obtained an informed consent from the patient herself for permission to publish. There are no conflicts of interest to be declared.

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