CASE REPORT

Portal vein aneurysm in thalassaemia

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Accepted 7 August 2017

SUMMARY

Arterial aneurysms are more common than visceral venous aneurysms. Portal vein aneurysms being the most common type of visceral venous aneurysms. Here, we present an 18-year-old young woman with thalassaemia major, who presented with headache, palpitation, shortness of breath and a recent increase in blood transfusion rate. On clinical examination, she had hepatosplenomegaly. Ultrasonography revealed hepatosplenomegaly with fusiform dilatation of extrahepatic portal vein, which was confirmed to be portal vein aneurysm on contrast enhanced CT. Though portal vein aneurysms were previously thought to be rare, recently they are increasingly diagnosed with the use of cross-sectional imaging. Recognition of this finding can help to avoid potential confusion with other periportal cystic masses of different aetiologies, especially on sonography.

BACKGROUND

Portal vein aneurysms are focal saccular or fusiform dilatation of splenoportal venous system and are defined as portal vein diameter greater than 2 cm.¹ It is the most common type of visceral venous aneurysm.² Though these are rare entities, portal vein aneurysms are increasingly discovered by ample use of modern cross-sectional imaging like CT and MRI. Portal vein aneurysms are either congenital or acquired. Patients may be asymptomatic or symptomatic, and it determines further management.³

CASE PRESENTATION

An 18-year-old young woman with thalassaemia major presented with severe pallor, headache, palpitation and shortness of breath. She was on monthly blood transfusion of one unit packed red blood cells since 10 years of age. She developed features of hypersplenism and experienced a recent increase in blood transfusion rate from once a month to thrice a month over last 6 months due to rapid fall in haemoglobin level. Clinical examination revealed severe pallor, mild pedal oedema and hepatosplenomegaly.

INVESTIGATIONS

Laboratory investigations showed microcytic hypochromic anaemia with haemoglobin of 4.9 gm%, leucopenia (total leucocyte count: $2.9 \times 10^9/L$) and thrombocytopenia ($61 \times 10^9/L$). Bilirubin levels and other liver function test parameters were within normal range.

Ultrasonography revealed hepatosplenomegaly and a tubular cystic lesion measuring 5×3 cm

extending from the head of pancreas to the porta hepatis which showed colour flow on Doppler (figure 1). Hepatic artery, common bile duct and intrahepatic biliary radicals were normal. Subsequent CT confirmed the portal lesion as extrahepatic portal vein aneurysm (figure 2).

DIFFERENTIAL DIAGNOSIS

Differentials of cystic lesions at porta hepatis are portal vein aneurysm, hepatic artery aneurysm and choledochal cyst. Portal vein aneurysm shows colour flow with undulatory monophasic flow. Hepatic artery aneurysms show colour flow with arterial waveform, but choledochal cysts do not show such colour flow and has communication with biliary channels.

TREATMENT

In view of rising transfusion rate and hypersplenism, she underwent splenectomy after triple vaccination.

OUTCOME AND FOLLOW-UP

At 1-year postsplenectomy, her haemoglobin was 6.9 gm/dL and her transfusion rate returned to the baseline of once a month. Total leucocyte and platelet counts were $5.4 \times 10^9/L$ and $110 \times 10^9/L$, respectively. Liver function test parameters were also normal.

Follow-up Doppler at the same time demonstrated regression of the portal vein diameter from 3 cm to 1.8 cm.

DISCUSSION

Arterial aneurysms are more common than visceral venous aneurysms; however, with extensive use of cross-sectional imaging, the detection rate of portal vein aneurysms has increased.⁴ The first case was reported in 1956 by Barzilai and Kleckner.⁵ Portal vein aneurysm represents about 3% of all venous aneurysms and has a reported prevalence of

Figure 1 Ultrasonography images near porta reveals fusiform dilatation of extrahepatic portal vein.

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To cite: Das S, Dey M, Kumar V, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2016-218551

Figure 2 Contrast enhanced CT scan of abdomen in (A) axial and (B) coronal planes reveal extrahepatic portal vein aneurysm and gross hepatosplenomegaly.

0.43%.^{6,7} There is no correlation between incidence of portal aneurysm and age or sex of the patient.⁷

Portal vein aneurysms may be congenital or acquired. However, the exact aetiology is still controversial. Congenital variety is believed to develop from persistent right vitelline vein which later on enlarges to form saccular aneurysm.⁸ Acquired aneurysms are commonly associated with cirrhosis and portal hypertension. In portal hypertension, there is intimal thickening and medial hypertrophy of the portal vein with gradual replacement by fibrous tissue resulting in weakening of the venous wall and rendering it susceptible to aneurysmal dilatation.⁹

According to Doust and Pearce, the maximum portal vein in normal population and those with cirrhosis never exceeds over 15 mm and 19 mm, respectively. Thus a diameter of greater than 20 mm is regarded as aneurysm. The intrahepatic portal branch aneurysms are considered if they measure greater than 7 mm in normal and 8.5 mm in those with cirrhosis.¹

The two most common locations for venous aneurysm are the main portal vein and at the splenoportal confluence.⁶ In this case, we encountered with aneurysmal dilatation of extrahepatic portal vein extending up to confluence of superior mesenteric vein and splenic vein.

The typical B-mode ultrasonography finding is a hypoechoic or anechoic structure at porta hepatis. Doppler shows monophasic colour flow in it, thus differentiating it from a liver cyst. It may mimic a tumour due to presence of echogenic mural thrombus. Ultrasound with Doppler study is an excellent tool for demonstrating and monitoring the size of the aneurysm. Contrast enhanced CT and MR angiography are helpful in case of equivocal ultrasound findings or to make out the exact anatomical detail for preoperative planning; however in most instances, ultrasound is sufficient to make final diagnosis.⁶

Most portal venous system aneurysms are asymptomatic. External compression and rupture are rare complications. Complete and non-occlusive thrombi in portal vein occur in 13.6% and 6%, respectively.¹⁰

The management protocol is still debatable. Asymptomatic aneurysms are usually treated conservatively, while surgical interventions are reserved for symptomatic cases or in cases of

Learning points

- ▶ Most cases of portal vein aneurysms are acquired in nature and the main aetiology is hyperdynamic circulation in the splenoportal axis.
- ▶ Most patients are asymptomatic leading to under-reporting of the condition.
- ▶ Imaging diagnosis of portal vein aneurysms can be made on the basis of ultrasonography and CT.

impending rupture or ruptured or thrombosed aneurysms.¹¹ Unlike splenic artery, pseudoaneurysms which are the most common arterial visceral aneurysms and need mandatory intervention to prevent catastrophic rupture, portal aneurysms do not need direct intervention but instead need treatment of primary underlying condition—portal hypertension in most cases.¹² In the present case, no surgical intervention was required for portal vein aneurysm per se and it regressed following splenectomy with correction of hypersplenism.

Contributors HL conceived the manuscript, SD prepared the manuscript, MD performed the literature review and collected the images, VS helped in manuscript preparation. The manuscript was reviewed by HL and all the authors approved the final manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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