Secondary Budd-Chiari Syndrome due to Hepatic Tuberculosis in a Pediatric Patient Managed by Left Hepatic Vein Stenting

CASE

A 12 yr old boy presented to the pediatric OPD with complaints of gradual weight loss, decreased appetite for four months, and gradual distension of the abdomen for 1.5 months. The child was hemodynamically stable, cachectic, and icteric at presentation. He had pallor and gross ascites on examination. A detailed evaluation revealed low hemoglobin (9.1 g/dL), hyperbilirubinemia (2.8 mg/dL) with elevated liver enzymes (ALT–284 IU/L, AST–212 IU/L). Renal function parameters were within normal limits. Mantoux test was strongly reactive (17 mm of induration). Viral markers, serological screening for autoimmune hepatitis, and prothrombotic workup (Protein C, S, Factor V mutation, homocysteine levels) were negative. Ultrasonography showed gross ascites, splenomegaly, an atrophied right lobe of the liver with multiple echogenic calcific foci, and compensatory left lobe hypertrophy with heterogeneous echotexture. The right and caudate lobe had multiple hypoechoic lesions, causing occlusion of the left hepatic vein (LHV) near the ostium (Figure 1a). Doppler examination revealed monophasic flow within the LHV and multiple tiny intrahepatic veno-venous collaterals (Figure 1b, c). Additionally, contrast-enhanced CT demonstrated multiple abdominal wall collaterals and conglomerated and necrotic retroperitoneal lymphadenopathy (Figure 2). The ascitic fluid analysis confirmed its transudative nature with a slightly elevated ADA level; however, it was negative for acid-fast bacilli and malignant cells. With these findings, a provisional diagnosis of abdominal tuberculosis with hepatic involvement complicated by BCS was made.

Initially, ascites was managed with diuretic therapy and repeated paracentesis under IV albumin coverage. USG-guided plugged liver biopsy from the caudate lobe lesion revealed tubercular etiology. Then anti-tubercular therapy (ATT) was started considering the concurrent hepatopathy. Upon ascitic control and a decrease in hyperbilirubinemia to 1.6 mg/dL, the child was discharged on the 18th day of admission after repeated requests from his parents. The child was admitted again after two months due to worsening ascites. Repeat investigations showed a total bilirubin of 2.4 mg/dL, low serum albumin (2.2 mg/dL), and negative ascitic fluid analysis for tuberculosis. Repeat biopsy from the left lobe was supportive for hepatic venous outflow obstruction. Thus, LHV recanalization was planned after the multidisciplinary discussion to treat BCS.

After USG-guided percutaneous access of LHV using a micropuncture set (Neff percutaneous access set, Cook Medical), a 5F vascular sheath was placed. The venogram showed multiple intrahepatic veno-venous collaterals with non-opacification of IVC (Figure 3a). After multiple attempts, the stricture was negotiated with an 0.018″ guidewire (V-18, Boston Scientific). 0.018″ guidewire was snared from a previously obtained right jugular access (Figure 3b). A 4F catheter was advanced into the LHV from the jugular approach. Over an angled 0.035″-260 cm stiff guidewire, the tight stricture was sequentially dilated using 4 mm, 6 mm, 8 mm, and 10 mm balloon catheters (Figure 3c).

Significant stenosis persisted despite multiple attempts of angioplasty using a 10 mm balloon. Therefore, a 10 × 40 mm self-expanding metallic stent (Epic, Boston Scientific) was deployed across the stricture. The post-stenting venogram showed normal opacification of LHV with free contrast flow into the IVC (Figure 3d). In the end, the percutaneous transhepatic tract was embolized after removing the sheath using a 35-3-5 coil (Nester, Cook Medical). No periprocedural complication was observed.

Low molecular weight heparin (LMWH) was continued for five days after the procedure and was switched to oral Warfarin to maintain a target INR of 2–3. Low-dose diuretics and a salt-restricted diet were continued for two months. Follow-up USG showed patent LHV stent after one month (Figure 4), a decrease in intrahepatic veno-venous collaterals, and a marked reduction in ascites. Laboratory parameters showed normalization of bilirubin and improvement in serum albumin levels with only mildly elevated liver enzymes.

DISCUSSION

Hepatic tuberculosis is a rare cause of hepatic vein obstruction, causing secondary Budd-Chiari syndrome (BCS). To date, only two cases of secondary BCS due to hepatic tuberculosis have been reported.[1,2] Ongoing inflammation initiates a fibrotic response which causes hepatic fibrosis and a vascular occlusion in hepatic

Abbreviations: ATT: anti-tubercular therapy; BCS: Budd-Chiari syndrome; LHV: left hepatic vein; LMWH: Low molecular weight heparin

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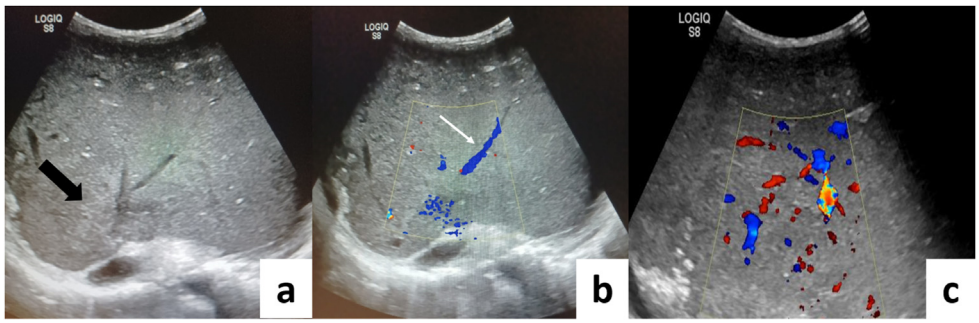

Figure 1 (a) USG shows an enlarged left lobe with heterogeneous parenchymal echotexture, an ill-defined iso to hyperechoic mass (black arrow) involving LHV; (b, c) Colour Doppler shows monophasic flow in LHV, s/o LHV stenosis with multiple tiny intrahepatic veno-venous collaterals.

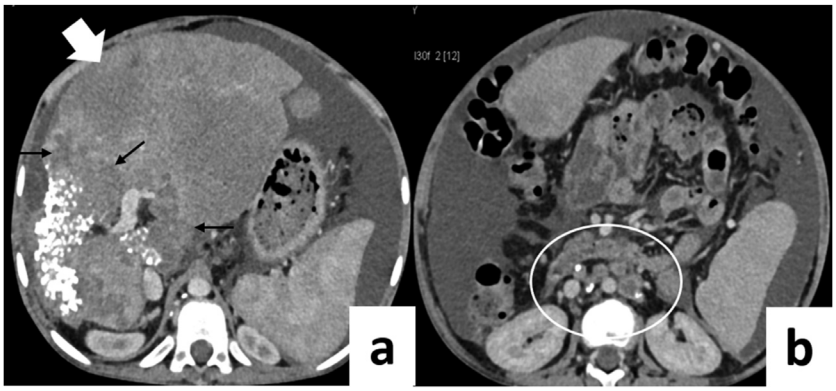

Figure 2 (a) Axial CECT image shows atrophied right lobe, multiple discrete and conglomerated hypoechoic lesions in the right and caudate lobe (black arrows) with enlarged and congested left lobe (white arrow); (b) Associated findings include multiple enlarged and necrotic retroperitoneal lymphadenopathy with few of them showing calcifications (white circle b).

tuberculosis.[1] Our case demonstrated an enlarged and congested left lobe with multiple intrahepatic veno-venous collaterals due to occlusion of the left hepatic vein by a tubercular lesion. The right lobe was atrophied in this case. Thus, occlusion of the only remaining HV (i.e., LHV) sufficed to cause hepatic decompensation.

Upon diagnosis of BCS, anticoagulation is started unless contraindicated. The presence of liver failure necessitates liver transplantation. The underlying primary disorder, if any, should be addressed to prevent the recurrence of BCS. Endovascular intervention is the recommended therapy, and it has been shown to have prolonged 5-year survival of up to 75% in BCS.[3] A stepwise approach is followed in managing BCS, starting from anticoagulation, angioplasty ± stenting, creation of portosystemic shunt, and finally, liver transplantation. Anatomical recanalization of HV and/or IVC shows improved liver synthetic functions compared with direct IVC to portal vein shunt (DIPS)[4]; hence, HV recanalization was considered first in the present case. Acute and anterosuperior angulation made endovascular recanalization of tight LHV stricture not feasible. Thus, the percutaneous approach was preferred in this case. The long segment and fibrotic nature of LHV stricture made the procedure technically challenging; however, we could negotiate it after multiple attempts. This case shows successful recanalization of HV using a combined percutaneous and transjugular approach with good functional outcomes.

Secondary BCS due to hepatic tuberculosis is clinically challenging to manage. Besides anti-tubercular therapy and other supportive measures, endovascular intervention plays a pivotal role in dealing with the BCS component.

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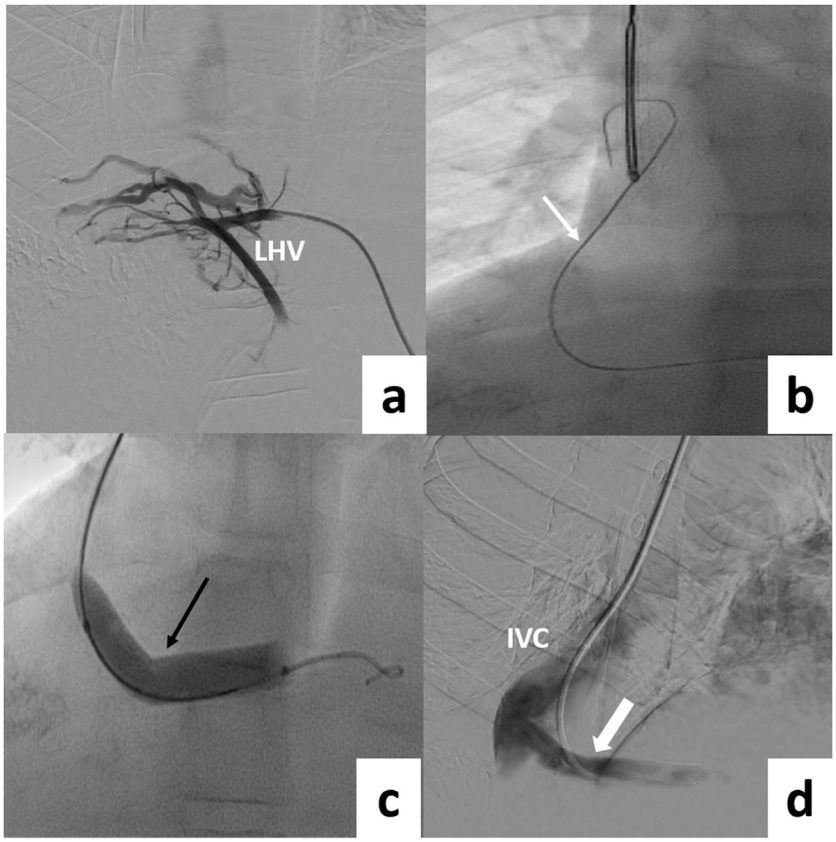

Figure 3 (a) Left hepatic venogram following percutaneous transhepatic access shows long segment occlusion of LHV with multiple collaterals around the LHV and non-opacification of IVC; (b) Stricture was negotiated, and a 0.018″ guidewire (white arrow) was snared through transjugular route to obtain a through and through access. (c) The stricture was serially dilated using a 4–10 mm balloon; (d) Venogram after stent deployment (black arrow) shows free flow into the IVC (white arrow).

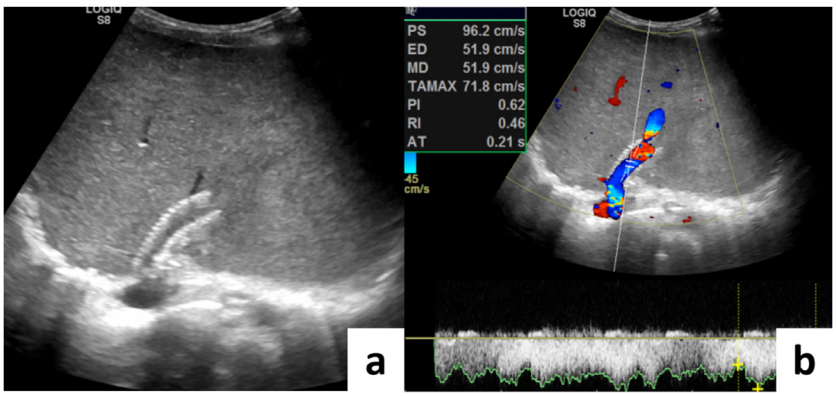

Figure 4 (a, b) Follow-up USG at one month shows the stent *in situ* with the flow within the stent.

INFORMED CONSENT

Informed consent was obtained from the patient's parents for publication of this case report and accompanying images.

CONFLICTS OF INTEREST

The authors have none to declare.

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