Pediatric Oncall Journal
Volume : 20, Issue 1:33
DOI: https://doi.org/10.7199/ped.oncall.2023.12

PEDIATRIC ONCALL
CHILD HEALTH CARE

TEACHING FILES (GRAND ROUNDS)

A CHILD WITH BUDD CHIARI SYNDROME AND RECURRENT THROMBOSIS

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ARTICLE HISTORY
Received 28 December 2022
Accepted 18 January 2023

KEYWORDS
Budd Chiari Syndrome,
Thrombosis, Coagulopathy.

Clinical Problem:

A 10-month-old girl presented with ascites. At 3 months of age, she had ascites and was diagnosed to have Budd Chiari syndrome (BCS) due to acute thrombosis of Middle and Right Hepatic veins (MHV and RHV). At the age of 3 months, coagulation studies showed decreased protein C activity, decreased Antithrombin III activity and increased serum homocysteine levels. Subsequently, she was started on injectable low molecular weight heparin, spironolactone, folic acid. She was also started on propranolol as ultrasound doppler of abdomen and portal system showed signs of portal hypertension. At 7 months of age she underwent venoplasty of Middle and Right Hepatic veins (MHV and RHV) and anticoagulation was continued with LMWH. Despite this, she had 2 hospital admissions for acute liver decompensation (ascites). She also developed gangrene of left middle finger at 8 months of age post venoplasty that auto-amputated inspite of being on LMWH. Despite being on treatment with diuretics, LMWH, propranolol, liver dysfunction continues. Current ultrasound of abdomen and doppler showed no demonstrable flow in hepatic veins, low velocity flow in inferior vena cava (IVC) along with liver cirrhosis and portal hypertension. INR was 2 with prothrombin time 24.2 sec (control 12 sec); partial thromboplastin time of 49.4 sec (control 28 sec). She underwent a repeat venoplasty of RHV and MHV. However, she continues to have persistent ascites post-venoplasty (diuretic resistant) and requires repeated ascitic tap. A repeat doppler after 1 week of venoplasty again showed that the MHV flow was blocked. She has had growth failure and her weight remained 6 kg in spite of all nutritional rehabilitation. Her albumin was 2.5 gm/dl and bilirubin was normal.

What is the cause of recurrent thrombosis in this child?

Discussion:

BCS is a rare disorder characterised by hepatic venous outflow obstruction which can be thrombotic (majorly) or non-thrombotic. It can occur anywhere along the venous course from the hepatic venules to the junction of the inferior vena cava (IVC) and the right atrium.[1] The most common causes of BCS in children are inherited hypercoagulable states such as Protein C and S deficiency.[2] In addition, decreased levels of in antithrombin III have also recently been linked to the cause of BCS which can be seen as increased values of PTT.[3] Another prominent risk factor for BCS is hyperhomocysteinemia.[4] Thus it is a proven fact that BCS is a thrombophilic state, but the coagulation function in BCS is unpredictable due to liver dysfunction-related coagulopathy.[5] Natural anticoagulant protein levels like protein C and protein S decrease progressively as the severity of liver disease increases. They reduce by 10% to 65% of normal, which is similar with the range of values observed in patients with inherited deficiencies.[6] To specifically identify the cause of coagulopathy in such patients, we can use the values of Tissue factor pathway inhibitor (TFPI) which is produced by endothelial cells. In patients with chronic liver disease, TFPI levels are normal or elevated. However, in conditions like protein S deficiency TPFI anticoagulant pathway is impaired, leading to low levels of TFPI.[6] In our patient TFPI could not be tested due to non-availability. Thus, in our patient, the most likely cause of repeated thrombosis is a hypercoagulable state.

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Compliance with ethical standards

Funding: None

Conflict of Interest: None

References:

1. Aydinli M, Bayraktar Y. Budd-Chiari syndrome: etiology, pathogenesis and diagnosis. World J Gastroenterol. 2007 May 21;13(19):2693-6.
2. Redkar R, Bangar A, Hathiramani V, Raj V, Swathi C. Pediatric Budd-Chiari Syndrome: A Case Series. Indian Pediatr. 2018 Oct 1;55(10):871-3.
3. Jagtap A, Shanbag P, Vaidya M. Budd-Chiari Syndrome due to Antithrombin III Deficiency. Indian J Pediatr.2004 Jan 1;70:1003-5.
4. Li XM, Wei YF, Hao HL, Hao YB, He LS, Li JD, et al. Hyperhomocysteinemia and the MTHFR C677T mutation in Budd-Chiari syndrome. Am J Hematol.2002 Sep 1;71(1):11-4.
5. Jain A, Dhore P, Meshram M, Bhatia S, Shukla A. Patients With Budd-Chiari Syndrome Have Variable Coagulation Status on Thromboelastography at Diagnosis. J Clin Exp Hepatol. 2018/10/25 ed. 2019;9(4):460-7.
6. Kujovich JL. Coagulopathy in liver disease: a balancing act. Hematology Am Soc Hematol Educ Program. 2015 Dec 5;2015(1):243-9.