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Anaesthetic Challenges Faced In A Paediatric Budd Chiari Syndrome With Congenital Neutropenia Posted For IVC And Portal Vein Dilatation.

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Abstract

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Introduction

Budd Chiari Syndrome (BCS)[1,3,4] is rare in children. It commonly results from vascular obstruction due to a web or a membrane in IVC. The reason for the occurrence of such a membrane is not clear, but may be congenital. Hyper viscosity is a central feature in the pathogenesis of BCS and may be responsible for complete occlusion or partial obstruction. In India, BCS due to chronic liver disease accounts for 7.4%.[1] Here, we are reporting a case of successful management of 11years old female, diagnosed with Budd Chiari Syndrome with congenital neutropenia planned for venography and IVC stenting.

11year developmentally normal female child with no other comorbidities presented with a history of multiple episodes of abdominal pain and distension for the past 4 years with 2 episodes of backache and abdominal pain in the present visit. She was diagnosed with congenital neutropenia (Kostmann syndrome) at 15 months of age by bone marrow biopsy, which showed maturation arrest in the myeloid lineage at the stage and eosinophilia, for which she was treated with repeated doses of GM-CSF. Later, at 11 years of age, Budd Chiari syndrome was diagnosed with clinical features of hepato-splenomegaly and dilated veins over the anterior chest wall, indicating characteristics of IVC obstruction. UGIE revealed grade 1 oesophageal varices. USG showed IVC obstruction with severe narrowing and major hepatic vein was partly visualized in the distal part with severe narrowing. Preoperative investigations are as follows: Hb 8.2 g/dl, total count- 4,810/mm^3 with neutrophils of 63 %. The airway examination did not reveal abnormalities. The patient was posted electively for diagnostic venography followed by balloon dilatation of IVC and portal vein. General anaesthesia was administered to the patient with ASA standard monitoring after securing wide-bore iv access. Perioperative course was uneventful.

Budd-Chiari syndrome (BCS)[1,2] is characterised by structural and functional abnormalities of the liver caused by obstruction to liver venous blood flow from small hepatic veins to the IVC at multiple sites. The aetiology of BCS is diverse, but many patients have been reported to have inferior vena cava (IVC) obstruction due to a web or a membrane. In our patient, myeloid arrest could be the cause of the development of this syndrome.

Treatment modalities vary depending on the extent of thrombosis and the amount of liver tissue functional to carry out its functions.[3] Management options in management are medical, surgical, or radiological[4]. Surgical treatment of these patients is difficult and is associated with high morbidity and mortality. The treatment of BCS with the interventional procedure has been reported in recent times. The balloon

dilatation of IVC is a safe and effective procedure and also provides a pathway for blood flow.

In Budd Chiari syndrome, obstruction of venous outflow results in portal hypertension and hepatocellular damage eventually leading to cirrhosis, being very rare in infants and young children, and the clinical presentation is varied among them. It may present as acute fulminant hepatic failure with pain, hepatomegaly, ascites, and rapid deterioration of liver function with encephalopathy or as acute non-fulminant BCS.

In preoperative management of BCS, the extent of liver damage and other organs involved should be assessed. Hypersplenism may result in thrombocytopenia and impaired production of clotting factors may lead to coagulopathy. Correction of coagulation abnormalities is indicated if there is active bleeding. Patients presenting for major surgery should have blood products arranged if preoperative coagulation abnormalities are noted. Pre-existing infections require appropriate antibiotic therapy.

Risk assessment could be carried out with PELD score, ASA physical status, and child Turcotte Pugh scoring[5]. These scoring systems would reveal the outcome and prognosis of the patient.

The goals of anaesthesia management of BCS[3] include preservation of hepatic blood flow and perfusion, maintenance of oxygen delivery, and adequate treatment of perioperative anticoagulant therapy. Regional anaesthesia is generally preferred to general anaesthesia due to preservation of hepatic blood flow. The choice of anaesthetic technique depends on the prognostication of the liver disease and the presence of coagulation may preclude regional anesthesia.

Dosing of drugs[5] is another important concern in liver disease, as all drugs are metabolized and detoxified in the liver. This should be taken into account prior to the administration of any drug. Inhalational agents such as enflurane and halothane should be avoided. Succinylcholine and vecuronium are avoided, whereas atracurium can be given safely as it undergoes Hoffman elimination. Fentanyl at moderate doses would not cause impairment in hepatic oxygen delivery and it can be administered in aliquots to prevent sympathetic response and pain relief.

Kostmann syndrome[6] is characterized by the arrest of neutrophil maturation in the promyelocyte stage. The resulting deep neutropenia leads to an increased susceptibility and severity of bacterial infections from early childhood These infections progress frequently to systemic illness and septicemia, hence the serious morbidity and mortality associated with this condition. BCS per se would lead to infections; here in our patient, it is associated with Kostmann syndrome, so pre and post-administration of antibiotics were mandatory.

In our case, all the aforementioned points had been taken into account and general anesthesia was administered to the child as regional anaesthesia was not feasible.

Anaesthetic management of a pediatric patient with the liver disease is a complex task.. The success rates of these cases lie in a proper preoperative workup, management of complications and infections, and appropriate postoperative care. Thromboprophylaxis and proper drug dosing need to be considered in these patients for a fruitful outcome. Additionally, a multidisciplinary approach is mandatory in the management of these cases.

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