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Clinical Case Reports

Budd–Chiari Syndrome Secondary to Burkitt's Lymphoma in a 5-Year-Old Child

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Abstract

Summary: We report a rare case of Budd–Chiari syndrome secondary to Burkitt's lymphoma in a 5-year-old boy presenting with jaundice and abdominal pain. This case highlights the diagnostic challenges in paediatric patients presenting with hepatomegaly and cholestatic jaundice and emphasizes the importance of considering underlying malignancies in atypical cases of hepatic venous outflow obstruction.

Keywords: Burkitt lymphoma, Budd Chiari syndrome, venous thromboembolism/ etiology.

Introduction

Burkitt's lymphoma (BL) is a highly aggressive non-Hodgkin lymphoma (NHL) of B-cell origin, characterised by the translocation and deregulation of the MYC gene.[1,2] It accounts for 6–8% of childhood malignancies, with a median age of presentation of approximately 10 years. Paediatric NHLs are predominantly high-grade, and BL is known for its rapid progression and frequent extranodal involvement, particularly in the abdomen, bone marrow, and central nervous system.[1]

Budd–Chiari syndrome (BCS) is a rare disorder caused by obstruction of hepatic venous outflow, which may be primary (due to venous abnormalities) or secondary (due to external compression or invasion by tumours).[3] The Association of BL with BCS is extremely rare and can lead to diagnostic confusion, as both conditions may initially present with hepatomegaly and jaundice.

Case Presentation

A 5-year-old boy, resident of Bahria Town, Rawalpindi, presented to the emergency department with complaints of abdominal pain, anorexia, and yellow discoloration of the skin and sclera for 20 days. The pain was gradual in onset, intermittent, and generalised in nature. There was no history of itching, clay-coloured stools, fever, or bleeding tendencies.

Past and Family History

He had no history of transfusions, injections, or chronic illnesses. He was a product of normal vaginal delivery and had normal developmental milestones. His immunisation status was up to date. No family history of similar illnesses was noted.

Examination

The child appeared unwell and had deep icteric. Vitals were stable. The anthropometric measurements were between the 10th and 25th percentiles. There was no evidence of pallor, lymphadenopathy, or oedema.

Abdominal examination revealed hepatomegaly (4 cm below the costal margin, firm, 13 cm span) and splenomegaly (1 cm below the costal margin). Ascites was clinically present, with positive shifting dullness.

Investigations

CBC: Hb 10.3 g/dL, WBC 9500/µL, Platelets 531,000/µL

LFTs: Total bilirubin 9.9 mg/dL (direct 8.2 mg/dL), ALT 182 U/L, ALP 120.9 U/L

Serology: Hepatitis A, B, C, and E negative

Ultrasound: Mild hepatosplenomegaly with mild ascites

During hospitalisation, jaundice worsened, and the patient developed abdominal distension and tender hepatomegaly. The patient also presented with clay-coloured stools and low-grade fever.

Advanced Imaging

CT abdomen and pelvic CT revealed multifocal thrombosis of the inferior vena cava with hepatomegaly, nutmeg appearance of the liver, and flip-flop pattern suggestive of Budd–Chiari syndrome, along with acute pancreatitis and right renal vein thrombosis.

A triphasic CT scan revealed periportal hypodensities, hepatic and renal hypodense lesions, and mesenteric lymphoid masses, suggesting a lymphoproliferative disorder.

Ascitic Fluid Analysis:

The protein level was 49 g/L, LDH was 2296 U/L, and ADA was 34.3 U/L, with 80% lymphocytes and a few atypical cells.

Histopathology: Liver biopsy revealed sheets of monotonous intermediate-sized lymphoid cells with a "starry-sky" appearance, confirming Burkitt's lymphoma.

Final Diagnosis:

Budd–Chiari syndrome secondary to hepatic infiltration by Burkitt's lymphoma.

Discussion

This case demonstrates a rare presentation of BL, manifesting as Budd–Chiari syndrome. Burkitt's lymphoma can involve the liver either by direct infiltration or secondary thrombosis due to

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hypercoagulability and venous obstruction.[4] Paediatric lymphomas are known to induce a prothrombotic state, and studies have reported venous thromboembolism in up to 10–15% of cases within the first year of diagnosis.

In this patient, hepatic venous thrombosis and IVC obstruction were likely caused by lymphomatous invasion. The diagnosis was exigent due to overlapping symptoms of hepatic illness and the scarcity of such presentations in children. Imaging and biopsy are crucial investigations that lead to diagnosis.

The gold standard for the diagnosis of lymphoma is histopathology, which shows hyperchromatic monomorphic lymphoid cells with phagocytic histiocytes, the classic "starry sky" pattern.[5]

Management and Outcome

Burkitt lymphoma management includes chemotherapy or chemoimmunotherapy, depending on the disease stage. Prophylaxis for tumour lysis syndrome and febrile neutropenia and monitoring for thrombotic complications remain the main supportive treatments.

The child was then referred to the paediatric oncology unit for further management and initiation of chemotherapy after the diagnosis was confirmed.

Conclusion

This case highlights the importance of considering unusual diagnoses, such as malignancy, in paediatric patients presenting with unexplained hepatic venous obstruction. Imaging and histological evaluation early in the course of the disease are essential for timely diagnosis and management of rare diseases such as Budd–Chiari syndrome secondary to Burkitt's lymphoma.

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