

carries a high mortality rate [4]. There is no precise data regarding the prevalence of ARDS during malaria infection; however, it is predicted nearly 20-30% of malaria patients admitted to ICU develop ARDS [5]. Proposed mechanism of development of ARDS is pulmonary vasculature dysfunction secondary to liberation of inflammatory mediators which increase vascular permeability, and parasitized RBCs' sequestration cause injury. Clinically, patients developing sudden onset tachypnea and dyspnea. Life threatening hypoxemia may develop within a few hours. Two of our patients developed ARDS, one died and other required mechanical ventilation.

Hemophagocytosis is associated with malignant, genetic, and autoimmune diseases. Viral infections as a cause are mainly limited to EBV infection. Malaria is a very rare cause and the mechanism of hemophagocytosis in malaria is unknown [3]. High levels of cytokines have been reported in malaria patients with hemophagocytosis which resolves soon after successful treatment of malaria [6-9]. Prolonged hemophagocytosis, has not been reported in patients with falciparum malaria. Once the cytokine cascade is triggered, hemophagocytosis may continue independent of the presence of the malarial parasite.

Thus, we had a family of three children, all with falciparum malaria with three unusual complications occurring in the same family.

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Acute Myeloid Leukemia Presenting as Obstructive Jaundice

BINITHA RAJESWARI, ANU NINAN, SINDHU NAIR PRASANNAKUMARI* AND KUSUMAKUMARY PARUKUTTYAMMA

*From the Division of Pediatric Oncology and *Division of Pathology, Regional Cancer Centre, Trivandrum, India.*

Correspondence to:

Binitha Rajeswari, Lecturer, Division of Pediatric Oncology, Regional Cancer Centre, Trivandrum, India.

rbinitha@yahoo.co.in

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Jaundice as a presenting feature of pediatric acute myeloid leukemia is rare. We report two cases of AML who presented with obstructive jaundice, one with a malignant stricture at the common bile duct and other with a granulocytic sarcoma obstructing the bile duct. The prognosis is poor in these patients.

Key words: *Acute myeloid leukemia, Granulocytic sarcoma, obstructive jaundice.*

O bstructive jaundice as the presenting feature of acute myeloid leukemia (AML) is rare in children. It may be due to a stricture of the biliary tree or a granulocytic sarcoma compressing the biliary tree. We report two such cases.

CASE REPORT

Case 1: A one year old female child presented to us with pancytopenia (hemoglobin 4.5g/dL, WBC count 2100/mm³, platelet count 13,000/mm³). A thorough evaluation

including bone marrow study did not reveal any definite evidence of malignancy. The blood counts normalized in a month. She presented two months later, with fever, followed by increasing jaundice, pale stools and abdominal distension. She was sick, with severe pallor, jaundice, generalized edema and massive ascites. Hepatosplenomegaly could not be assessed due to the massive ascites. Laboratory investigations revealed hemoglobin 4.6 g/dL, platelet count 33000/mm³, total count 5100/mm³, serum bilirubin 4.8 mg/dL (conjugated bilirubin 3.6 mg/dL), SGOT 76 IU/L, SGPT 30 IU/L, ALP 263 IU/L and prothrombin time 13 seconds. Serologies for HIV and HBsAg were negative. Anti HCV titers were not done. CT scan and ultrasound scan of the abdomen showed a soft tissue lesion 6×4cm wedged between the pancreas and liver. There was moderate bilobar intrahepatic biliary radicle dilatation and common bile duct dilatation upto pancreatic segments. There was bulky celiac axis, mesenteric and retroperitoneal lymphadenopathy with moderate ascites and bilateral minimal pleural effusion. Ascitic fluid cytology showed atypical cells suggestive of leukemia/lymphoma infiltration. Flow cytometry analysis done on the ascitic fluid revealed positivity for CD13, CD33, CD117 and CD7 markers, diagnostic of AML, possibly M5. Bone marrow study was deferred due to her poor general condition. The patient was started on subcutaneous cytosine arabinoside. In the following two weeks she improved with clearing of jaundice, reduction of abdominal distension and improvement of blood counts. A bone marrow study done showed 3% blasts with normal hemopoietic elements. Ultrasound scan of the abdomen showed disappearance of the mass and return of the biliary channels to normal size. Despite starting chemotherapy with intravenous cytosine arabinoside and daunorubicin, she developed sepsis and died.

Case 2: A previously normal 10 month old female child, presented with history of progressively increasing jaundice, clay colored stools and high colored urine of 2 months duration, followed by swellings over both parotid regions and multiple ecchymotic patches one month later. The child was sick and malnourished with deep jaundice, pallor, ecchymoses on the face and generalized lymphadenopathy. There was massive hepatosplenomegaly with liver palpable 10 cm below the right costal margin, reaching up to right iliac fossa and spleen palpable 8 cm below the left costal margin, crossing the midline beyond the umbilicus. Laboratory investigations showed hemoglobin 11.7 g/dL, platelet count 23000/mm³, WBC count 62000/mm³ (Neutrophils 24%, lymphocytes 42%, myelocytes 10%, abnormal cells 24%), serum bilirubin 38mg/dL (conjugated bilirubin

30.6 mg/dL), SGOT 108 IU/L, SGPT 50 IU/L, gamma glutamyl transferase 499 U/L and LDH 1132 U. Serology for HIV and HBsAg were negative. Anti HCV titers were not done. Peripheral blood smear examination showed 33% peroxidase positive myeloid blast cells and a diagnosis of AML was made. The review of the parotid gland biopsy slides also showed infiltration by peroxidase positive myeloid blasts. Bone marrow studies could not be done due to the poor general condition of the patient. Ultrasonography and CT scan of the abdomen showed intrahepatic biliary radicle dilatation with distension of the gall bladder and dilatation of the proximal common bile duct. No mass was visualized. Magnetic resonance cholangiopancreatogram confirmed the above findings and revealed an obstruction at the level of proximal common bile duct possibly due to a malignant stricture. There was also distension of the gall bladder with dilatation of the right and left hepatic ducts and cystic duct. (**Fig. 1**) Chemotherapy could not be instituted because of the poor general condition of the patient and she succumbed to her illness.

DISCUSSION

Obstructive jaundice as a presenting feature of pediatric malignancy is rare. Lymphoma and neuroblastoma may present with biliary obstruction. Rhabdomyosarcoma of the biliary tract may also occur. Jaundice as a presenting symptom in AML is rare. It can occur due to drug induced hepatocellular damage, post transfusion viral hepatitis, infiltration of the liver by the leukemic process or obstruction of the biliary tract. Obstruction may be due to granulocytic sarcomas compressing the biliary tree or due to stricture of the biliary tree. There are very few case reports of AML presenting as obstructive jaundice, especially in children. Jaing, *et al.* [3] have reported a 4 year old boy with extrahepatic obstruction of the biliary



FIG. 1 MRCP showing obstruction at common bile duct with distension of gall bladder and dilatation of hepatic ducts.

tract in AML. In their patient, on CT scan of the abdomen, there was a mass lesion at the pancreatic head associated with biliary dilatation. This patient responded well to chemotherapy, followed by bone marrow transplantation and was disease free 15 months after diagnosis.

The granulocytic sarcoma of biliary tree may be detected radiologically as a stricture or thickening of the biliary tree [1,2,4,7,8] or as a mass causing extrinsic obstruction of the biliary tree [3,5,6]. The mass obstructing the biliary tree in AML is usually a granulocytic sarcoma. This may occur concurrently with leukemia or may precede the occurrence of leukemia by weeks to months [5-8]. In our first patient, imaging studies showed a mass lesion wedged between the pancreas and liver, producing compression of the biliary channels and so we considered the jaundice to be obstructive even though the alkaline phosphatase levels were normal. The mass was a granulocytic sarcoma causing extrinsic compression of the biliary tree. In our second patient, AML presented as a stricture of the biliary tree producing obstructive jaundice. In this scenario, the major differential diagnosis to be considered is a secondary sclerosing cholangitis which in children could be due to langerhans cell histiocytosis, immunodeficiency, sickle cell anemia or autoimmune diseases. In our patient, since the peripheral smear was diagnostic of AML, the obstruction was probably due to a malignant stricture.

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