Obstructive Jaundice due to Pancreatic Head Mass: A Rare and Unusual Presentation of Acute Myeloid Leukemia in Children

Ghasem Miri-Aliabad, Hamid Dahmardeh

1Department of Pediatrics, Children and Adolescent Health Research Center, Zahedan University of Medical Sciences, Zahedan, Iran
2Department of Radiology, Zahedan University of Medical Sciences, Zahedan, Iran

About 80% of hematologic malignancies in children are acute lymphoblastic leukemia (ALL), and 15–20% are acute myeloid leukemia (AML). AML occurs at any age and is classified into subgroups M0–M7. The clinical manifestations of AML are similar to those of ALL. However, gingival hypertrophy, disseminated intravascular coagulation, and chloroma are more common in some subtypes of AML. Granulocytic sarcoma (GS) or chloroma occurs more frequently in the neck, orbit, central nervous system (CNS), bone, and skin, and is rarely seen in the pancreas. Very few cases of GS of the pancreas have been reported in adults. Pancreatic involvement in pediatric leukemia is also very rare and has been reported in childhood ALL. Here, we report a 3-year-old girl with obstructive jaundice and a pancreatic mass as the initial manifestation of AML.

A 3-year-old girl was admitted to the emergency department with fever, abdominal pain, and jaundice. A heterogeneous mass measuring 23 × 27 × 49 mm was shown at the pancreatic head on abdominal ultrasound, along with dilatation of the common bile duct and intrahepatic bile ducts. The liver size was 116 mm and the liver parenchyma echo increased. The spleen size was normal. Contrast-enhanced abdominal computed tomography (CT) scan also confirmed the ultrasound findings as well as diffuse enlargement of the pancreatic body and tail (Figure 1A and B). Laboratory tests revealed obstructive jaundice (total bilirubin: 12.3 mg/dL, direct bilirubin: 10.4 mg/dL, aspartate aminotransferase: 145 IU/L, alanine aminotransferase: 235 IU/L, alkaline phosphatase: 3335 IU/L).

The complete blood count was as follows: White blood cell count: 20.7 × 10^3/mm^3 with 34% lymphocytes, hemoglobin: 9.8 g/dL, and platelet count: 68 × 10^3/mm^3. Serum amylase was 286 U/L (normal range, 30–100 U/L) and serum lipase was 330 U/L (normal range, 13–60 U/L). Erythrocyte sedimentation rate was 74 mm/h. Blood urea nitrogen, creatinine, blood glucose, sodium, potassium, calcium, phosphorus, albumin, lactate dehydrogenase, prothrombin time, partial thromboplastin time, and uric acid were all within normal range. The results of viral serological tests including hepatitis A, B, and C; HIV, cytomegalovirus, and Epstein–Barr virus were negative. Bone marrow aspiration (BMA) showed AML M2 with 60% blasts, and flow cytometric analysis was also confirmed. Cytogenetic analysis of bone marrow aspirate showed t(8; 21). Cerebrospinal fluid analysis indicated no evidence of CNS involvement. The patient underwent chemotherapy with the MRC 12 AML protocol. At the end of the induction chemotherapy, clinical and laboratory findings normalized. BMA also showed complete remission after induction chemotherapy. Pancreatic mass was completely resolved on CT scan at the end of the induction phase (Figure 1C and D). At last follow-up, the patient was asymptomatic 1 year after completion of treatment and was in remission.

In 20% to 40% of AML, extramedullary infiltration or GS is present at the time of diagnosis. Very few cases of GS have been reported in the pancreas, and mostly in adults. Obstructive jaundice is rare in pediatric malignancies and may occur in lymphoma, neuroblastoma, and rhabdomyosarcoma. However, pancreatic GS is an extremely rare presentation of pediatric
AML and only 2 cases have been reported in the literature so far (Table 1).

Rajeswari et al. reported 2 children with AML who presented with obstructive jaundice. In 1 case, GS was present between the liver and the pancreas, and in another patient, there was obstruction without evidence of a mass in the proximal part of the bile duct. Jaing et al. reported a 4-year-old child with AML M4 whose initial manifestation was obstructive jaundice and pancreatic head mass, similar to our case.

Pancreatoblastoma is one of the differential diagnoses for pediatric pancreatic masses, and it is the most common pancreatic malignancy in young children, despite its rarity. Other important differential diagnoses that should be considered include non-Hodgkin lymphomas, neuroblastoma, and rhabdomyosarcoma. In our case, abnormal findings in the peripheral blood smear, such as thrombocytopenia and blast, led to the decision to conduct BMA first rather than a pancreatic mass biopsy.

In conclusion, obstructive jaundice is a very rare manifestation of pediatric AML. Pancreatic involvement due to AML is very rare in children and may present as a mass in the head of the pancreas and obstructive jaundice. Therefore, in children with pancreatic involvement due to AML, suspicion for disease should be entertained.

Table 1. Reported Pediatric Cases of AML with Jaundice and Pancreatic Mass

| Author         | Age, Sex | Clinical Presentation | Imaging                                                                 | Definitive Diagnosis Method | Treatment                          | Outcome                          |
|----------------|----------|-----------------------|-------------------------------------------------------------------------|-----------------------------|-----------------------------------|----------------------------------|
| Rajeswari et al. | 1 year, F | Jaundice, abdominal distension, pallor | CT scan of the abdomen showed a soft tissue lesion 6x4 cm between the pancreas and liver | Ascitic fluid flow cytometry | Chemotherapy                      | Died due to sepsis               |
| Jaing et al.      | 4 years, M | Jaundice, abdominal pain | CT showed a mass in the head of the pancreas with biliary duct dilatation | BMA                         | Chemotherapy, bone marrow transplantation | Disease-free 15 months after diagnosis |
| Our Case         | 3 years, F | Jaundice, abdominal pain, fever | CT scan showed pancreatic head mass and diffuse enlargement of the pancreas | BMA                         | Chemotherapy                      | Disease-free 1 year after completion of treatment |
AML and obstructive jaundice, GS of the pancreas should be considered.

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