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Case Reports

Congenital Myeloid Leukaemia

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SUMMARY

Rare things are rare but they do occur. We present here a case of congenital myeloid leukaemia who presented with massive hepatosplenomegaly without other manifestations at the time of diagnosis.

INTRODUCTION

Many newborns present with congenital abdominal masses, that should be investigated to determine the underlying pathology. Common intra-abdominal masses are hepatic, splenic and renal. Here we report a case who presented with progressive abdominal distension since birth.

CASE REPORT

A two month old male child presented with progressive abdominal distension since birth. This baby was born by spontaneous vaginal delivery (SVD) following full term unremarkable pregnancy. According to the mother he was a weak child at birth.

There was no history of jaundice, fits, fever, vomiting, diarrhea or constipation. Baby was breast fed, had social smile and horizontal visual gaze. He was the 3rd issue from a consanguinous marriage with one healthy 3 years old sister and one intrauterine death (IUD) of a male child 2 years back.

Examination revealed a 4.5 kg healthy looking baby with normal anthropometric data without any dysmorphism. He was moderately anaemic and the anterior fontanella was open. The abdomen was protuberant with everted umbilicus, liver was palpable 4 cm below the costal margin in mid clavicular line, firm in consistency with a smooth surface. Spleen was 8-10 cm firm with a palpable notch heading towards the left iliac fossa. No free fluid was detected in the peritoneal cavity.

A differential diagnosis of congenital intrauterine infections, storage disease and congenital leukaemia was considered, and appropriate tests ordered.

On laboratory investigations, blood glucose was 77 mg/dl, bilirubin 1.4 mg/dl, ALK PO4 U/L with normal BUN, creatinine and serum electrolytes. Ophthalmological examination was negative for cherry red spot or any other intraocular abnormality. Ultrasound examination confirmed hepatomegaly and marked enlargement of spleen. No obvious lymph node enlargement was detected.

A peripheral blood smear revealed (along with Hb of 8.5 g/dl), TLC $63x10^9/l$, with poly 31, lymphos 31, mono 08, eosinophils 3, basophils 1, bands 7, myelocyte 7, metamyelocyte 8, promyelocyte 1, mononuclear cells 3, few nucleated red cells seen with marked degree of shift to the left and platelets 72,000/cm.

Bone marrow aspirate from tibial tuberosity showed hypercellular smear and fragments. Erythropoiesis was active and normoblastic. Myelopoiesis was hyperplastic with mature forms increased i.e. myeloblasts, myelocytes and metamyelocytes, megakaryocytes were increased in number, lymphos were prominent of mature type. No extramedullary cells were seen. A diagnosis of chronic (congenital) myeloid leukaemia was confirmed (Fig. 1).

DISCUSSION

Congenital leukaemia is a condition occurring very rarely. In a review in 1993, 175 cases were reported. Congenital leukaemia accounts for only 3% of childhood leukaemia. These are diagnosed within the first month of life at a rate 4.7 per million live births. Among the congenital

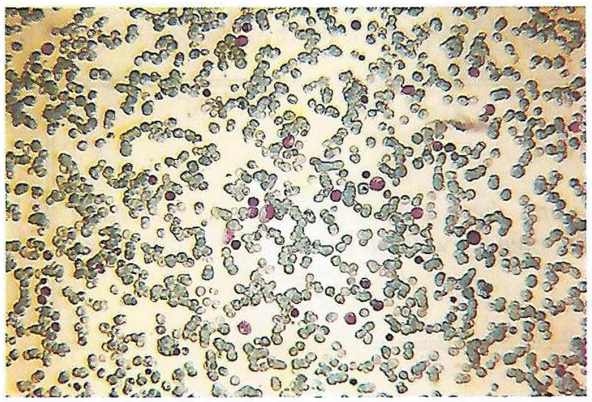

Fig. 1: Bone marrow aspirate from tibial tuberosity. Myeolopoiesis with mature forms increased myeloblasts, myelocytes and metamyelocytes.

leukemias, chronic myeloid leukaemia occurs with least frequency. Myeloid leukaemia appears to be predominant in this group (1) generally cases present with marked leukocytosis, petechiae, ecchymoses, and extramedullary involvement with massive hepatosplenomegaly, cutaneous nodules and CNS leukaemia (2). In our patient however, predominant features were hepatosplenomegaly without lymph and skin involvement.

Neuroblastoma and leukaemoid reactions secondary to erythroblastosis fetalis and severe congenital bacterial infection or viral infections may mimic congenital leukaemia but these can be ruled out by appropriate laboratory studies. More difficult to differentiate is transient myeloproliferative disorder which occurs primarily in neonates with trisomy 21 or chromosome 20 mosaicism. Most transient myeloproliferative disorders undergo spontaneous remission within a few weeks[3].

Our patient was two month old and had no features of trisomy 21. Congenital leukaemia has a poor prognosis. Although the short latency period suggests genetic predisposition. Studies suggest that intrauterine exposure to carcinogens is responsible for at least some cases of leukaemia in very young children[4].

The family refused treatment with chemotherapy but baby still survived till the age of 3 months with massive hepatosplenomegaly. There was no involvement of skin or lymph nodes.

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