Acute Lymphoblastic Leukemia: An Unusual cause of Nephromegaly in Infancy

Sir,

We present here an infant who had acute lymphoblastic leukemia with massively enlarged kidneys due to leukemic infiltrates. A 7-month-old girl child presented with complaints of low grade fever and coryza for 6 days. The mother noticed angular deviation of mouth and tongue to right side for the past 4 days and decreased movements of right upper limb for same duration. On examination, the child had pallor and few petechie over the trunk. The vitals were HR 122/min, pulses well palpable, RR 48/min; blood pressure was 134/94 mm Hg. Her anthropometry was: weight 6.6 Kg (10th centile), length 65 cm (25th centile), and head circumference was 41.5cm (25th centile). Central nervous system examination revealed normal sensorium. There was left facial lower motor neuron palsy, right hypoglossal palsy and right sided hemiparesis (upper motor neuron type). There were no meningeal or cerebellar signs. She had a soft hepatomegaly (2.5 cm below costal margin) and splenomegaly (6 cm below costal margins). Bilateral kidneys were palpable and occupied most of the abdomen, their surface was smooth.

The mother had undergone ultrasonography during the antenatal period and no abnormality of fetal kidneys was reported. Initial investigations revealed hemoglobin 5.8g%, TLC 7500/µl, polymorphs 35, lymphocytes 65 and Platelet count of $26X10^9$/L. The peripheral smear had normocytic normochromic to mildly hypochromic red blood cells, platelets were diminished and 3% blasts were present. The biochemistry showed blood urea 45mg/dL, serum creatinine 0.8 mg/dL, serum ALT 24 IU/L, AST 25 IU/L. albumin 4.1 gm/dL, SAP 240 IU/L. The serum sodium was 133 mEq/L, K 4.0 mEq/L, calcium 8.2 mg/dL, and phosphates 5.4 mg/dL. A venous blood gas had a pH of 7.35, bicarbonate 21 mEq/L, base excess -5 mEq/L. A lumbar puncture examination done showed 80 cells, polymorphs 45 and lymphocytes 25 and presence of abnormal cells; CSF sugar was 87 mg/dL and protein 62 mg/dL. Glomerular filtration rate as calculated by the Schwartz method was 45 ml/min/1.73m^2.

An ultrasonogram of the abdomen revealed liver of normal size and echotexture. The spleen was enlarged 9.2 cm, right kidney (10.7X 5.0 cm); left kidney (10.2 X 5.5 cm), bilateral grossly enlarged kidneys with smooth outline extending from epigastrium to pelvic inlet compressing midline retroperitoneal vessels. Multiple dilated tubular structures were seen in cortex and medulla. A computed tomography of the abdomen showed enlarged smooth kidneys with no cystic dilatation with maintained corticomedullary differentiation. An MRI of the brain was suggestive of generalized brain atrophy. A bone marrow aspirate showed presence of 21% lymphobalsts (PAS & myeloperoxidase negative blasts) in the marrow. A diagnosis of acute lymphoblastic leukemia with renal and CNS infiltrations was made. A renal biopsy was not done in view of persistent thrombocytopenia.

The child was started on chemotherapy for standard risk: ALL (prednisolone, L-asparginase and vincristine), intrathecal methotrexate, hydrocortisone and cytarabine. After the first cycle of chemotherapy the liver, spleen had regressed and the kidneys were also barely palpable. A repeat ultrasound showed a significant reduction in the size of the kidneys. The child died after 2 wks of chemotherapy due to febrile neutropenia.

Clinically evident renal enlargement occurs in only 2-5% of leukemia patients. Renal failure consequent to such involvement occurs rarely.[1] Also signs and symptoms of central nervous system involvement are seldom observed at initial presentation.

Although renal infiltration is relatively frequent in acute lymphoblastic leukemia, nephromegaly is unusual.[2,3] Kidneys are the most frequent extramedullary site for leukemic cell infiltration. This infiltration is usually diffuse and bilateral with a predominant involvement of the renal cortex. Nodular and hypoechoeic lesions are seen with tumors like lymphoma. Renal infiltration is often associated with involvement of other extramedullary sites like central nervous system, testis and skin.

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