view of the harmful effects of worm infection, we suggest that adequate measures be taken to prevent and treat these conditions.

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Wolman's Disease

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Wolman's disease is an extremely rare, fatal, autosomal recessive, lysosomal storage disease of cholesterol esters caused by deficiency of lysosomal lipase. About 50 cases have been reported in literature of which three cases are from India(l). We report here a new case because of its rarity and certain unusual features.

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Case Report

A two month old girl (Fig. *1)* first issue of a non-consanguinous marriage, belonging to the Maratha community, born of a full term normal delivery, was brought with progressively increasing abdominal distension noticed since birth. There was no history of any antenatal infection. The birth weight was 2.74 Kg. There was no history of fever, jaundice, convulsions, vomiting, diarrhea or bleeding from any site. The child had a social smile at six weeks of age.

On examination there was marked pallor and mild cervical and axillary lymphadenopathy. The weight was 3.2 Kg. The liver was firm, smooth and enlarged 7 cm below the costal margin. The spleen was enlarged firm and palpable 5 cms below the costal margin. Neurologic examination including fundoscopy was normal.

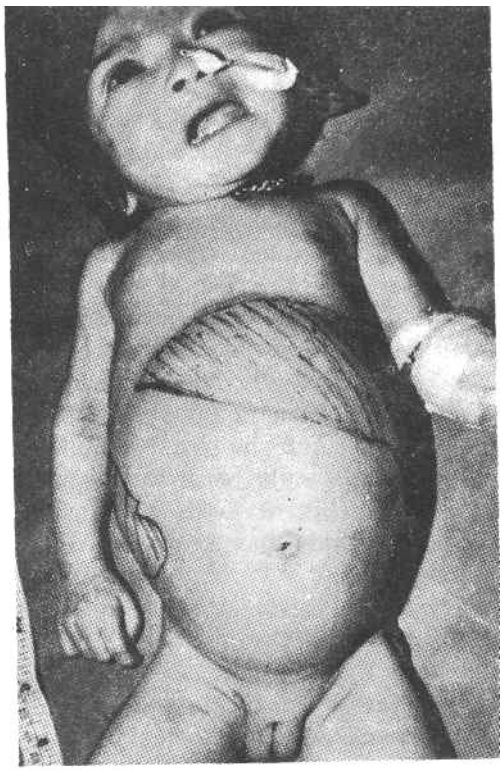

Fig. 1. Photograph of the patient showing markedly enlarged liver and spleen.

On investigation was the hemoglobin 6.7 g/dl, leucocyte count 22,600/cu mm and reticulocyte count 0.9% peripheral smear showed adequate platelets and vacuolated lymphocytes. The liver functions were deranged, with elevated liver enzymes (SGOT-270 IU/ml, SGPT-67 IU/ml, Serum bilirubin 2 mg/dl with a direct component of 1.9 mg/dl markedly prologed prothrombin time (more than 2 min) elevated LDH (1754 U/ml) and serum lipids (serum cholesterol 220 mg/dl serum triglycerides 472 mg/dl). Serum total protein level (8 g/dl) and serum albumin (3.5 g/dl) were normal. The 24 hour stool fat was normal (230 mg/24 hrs). X-ray abdomen *(Fig. 2)* showed bilaterally calcified adrenal glands, which was confirmed on ultrasound examination. Bone marrow aspiration showed numerous foam cells. The patient was diagnosed to have Wolman's disease on the basis of hepatosplenomegaly, calcified adrenals and foam cells in the bone marrow aspirate.

The child's condition deteriorated rapidly and she died of liver cell failure on the 14th day after admission.

On post-morten examination, the liver was enlarged, firm and the cut surface was yellow and greasy. The adrenal glands were firm, symmetrically enlarged and contained flecks of gritty calcified tissue. The small intestine was

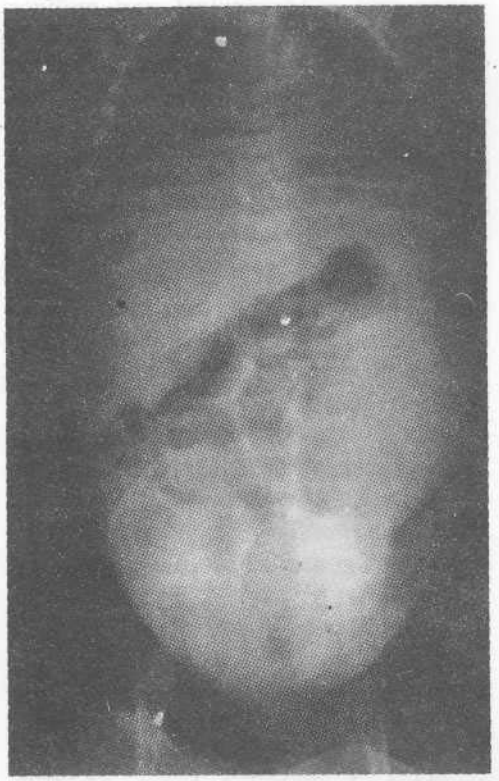

Fig. 2. X-ray abdomen showing bilaterally enlarged and calcified adrenal glands.

thickened and dilated with a dull, opaque yellow serosa. The spleen was grossly enlarged and firm with a reddish yellow cut surface. The kidneys and brain were grossly normal.

On microscopic examination, the hepatic parenchymal cells and Kupffer cells were enlarged and vacuolated. Large numbers of foamy histiocytes were found in the portal and periportal areas of the liver, in the adrenals, spleen and lamina propria of the small intestine. There were necrotic areas in the fasciculata and reticularis zones of the adrenals with prominent calcification.

Discussion

Wolman's disease occurs due to lysosomal acid lipase deficiency, leading to massive accumulation of cholesterol esters and triglycerides in most body tissues(2,3). The structural gene for the acid lipase enzyme is located on chromosome 10. The onset of symptoms is typically very early, within the first few weeks of life as in this case. Marked hepatosplenomegaly, mild lymphadenopathy, progresive inanition with anemia and deranged liver functions seen in this case were described in other cases reported in literature. But in this infant there was no evidence of fat malabsorption with forceful vomiting, diarrhea and steatorrhea which is striking in most reported cases(4). Plasma lipids though normal in the majority of cases were elevated in this case. Death occured by six months of age in most of the reported cases(5).

The definitive diagnosis of a patient and identification of carriers require the demonstration of decreased acid lipase levels in cultured skin fibroblasts or other tissues. However the demonstration of enlarged calcified adrenals on X-ray and ultrasound along with vacuolated lymphocytes on peripheral smear and foam cells in the bone marrow aspirate in an infant with hepatosplenomegaly and anemia is virtually pathognomonic of Wolman's disease(6). Autopsy and histopathology findings futher confirmed the diagnosis. Prenatal diagnosis is possible by assaying enzyme levels in cultured chorinonic villi or amniocytes(7).

The recommended management of these infants consists of strict cessation of breast feeding, avoidance of foods

containing fats and oils, vitamin supplementation and percutaneous application of minute amounts of sunflower oil to prevent essential fatty acid deficiency(8). Borie marrow transplantation, the use of lovastatin, and infusion of the missing enzyme are experimental therapies(9).

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Tuberous Sclerosis with a Brain Tumor

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Tuberous sclerosis or Bourneville disease is a neurocutaneous disorder characterized by the triad of seizures, mental retardation, and adenoma sebaceum. Intracranial lesions of tuberous sclerosis consist of calcified subependymal glial nodules and tubers occupying a cortical/subcortical location. Malignant transformation of the intracranial lesions of tuberous sclerosis occurs rarely(l). In a recent publication, none of the seven patients with this con-

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