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# Hepatic Hemangioendothelioma and Neonatal Hepatitis

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Infantile hemangioendothelioma is an uncommon vascular tumor of the liver(1). Many authors have described a triad of hepatomegaly, congestive heart failure (CHF) and cutaneous hemangioma(2-5). But occurrence of jaundice has been very rarely encountered with hemangiomata of the liver(4). During neonatal period when pepatic hemangioendothelioma is associated with jaundice, it poses a great problem in diagnosis. So far 21 cases of hemangioendothelioma with jaundice have been reported(4). We describe a 2-month-old infant who had hemangioendothelioma and neonatal hepatitis. Clinically his presentation simulated neonatal hepatitis. The rare association of hepatic hemangioendothelioma with neonatal hepatitis without cutaneous hemangioma and CHF prompted us to document this case.

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## Case Report

A two-month-old male infant was born to a fourth gravida mother, at term. On 5th day of life, the mother became aware of jaundice and since then it was waxing and waning. Stools were intermittently clay coloured while the urine was high coloured. Mother did not have the history of jaundice or rash during pregnancy. On examination, the infant was deeply jaundiced. He weighed 3.8 kg with a length of 56 cm and head circumstances of 35.3 cm. He had pedal edema. There were no clinical features to suggest intrauterine infection. There were no skin hemangiomas. The liver was palpable 4 cm below right costal margin, firm in consistency with smooth surface and no bruit was heard. Spleen was palpable 2 cm below left costal margin. Rest of systemic examination was normal.

On investigation, hemogram showed a hemoglobin of 12 g/dl with normal total and differential leucocyte and platelet counts. Liver function tests revealed a serum bilirubin of 15 mg/dl with conjugated bilirubin of 10 mg/dl; SGOT/SGPT was 50/63 IU; alkaline phosphates was 18 KAU, prothrombin time index was 48%; PTTK-57 seconds (control 40 seconds) and total proteins 4.6 g/dl (albumin 2 g/dl). *X*-ray chest and abdomen were normal. Blood culture received after the death was sterile. Investigations for intrauterine infections like toxoplasma, syphilis, rubella and cytomegalovirus were negative. Alpha-1-antitrypsin levels were normal. HBsAg was negative. Ultrasonography and CT scan of liver could not be performed because of short stay.

The child had downhill course and succumbed to sepsis within 48 hours of hospitalization. Necropsy revealed enlarged bile stained liver with 6 × 6 cm size hemangioma on the surface of the liver. On cutting slices, another hemangioma (5 × 5 cm) at porta hepatis was found. This was around the area of formation of common hepatic duct. Intrahepatic ducts were not dilated. The rest of the viscera including brain were unremarkable. There were no hemangiomata elsewhere. Histopathology of liver and hemangiomas showed changes of neonatal hepatitis (giant cell hepatitis) with cholestasis and hemangioendothelioma, respectively (*Figs. 1 a & b*).

## Discussion

Infantile hemangioendothelioma is a rare benign tumor of the liver(1,2). There are about 166 cases reported in English literature(1,3,5). One infant had been reported earlier from India(6). The exact incidence and etiology of the tumor is unknown. Hemangioendotheliomas can be single or multicentric. These may be associated with hemangiomas of the skin and other organs of the body(1).

Mclean *et al.*(2) described the triad of hepatomegaly (96%), high output CHF (60%) and cutaneous hemangioma (87%). Twenty one cases of hepatic hemangioendothelioma with hyperbilirubinemia have been reported so far and they constitute 12% of documented cases(4,5). The present case had jaundice and hepatomegaly. There were no cutaneous hemangiomas and CHF or gastrointestinal symptoms. His presentation simulated neonatal hepatitis. Elevation of serum bilirubin documented so far ranged from 8 to 15.9 mg/dl. Majority of them had conjugated hyperbilirubinemia (3.9-7.2 mg/dl)(4). The present patient had serum bilirubin of 15 mg/dl with conjugated fraction of 10 mg/dl. The mechanism of hyperbilirubinemia is believed to be due to

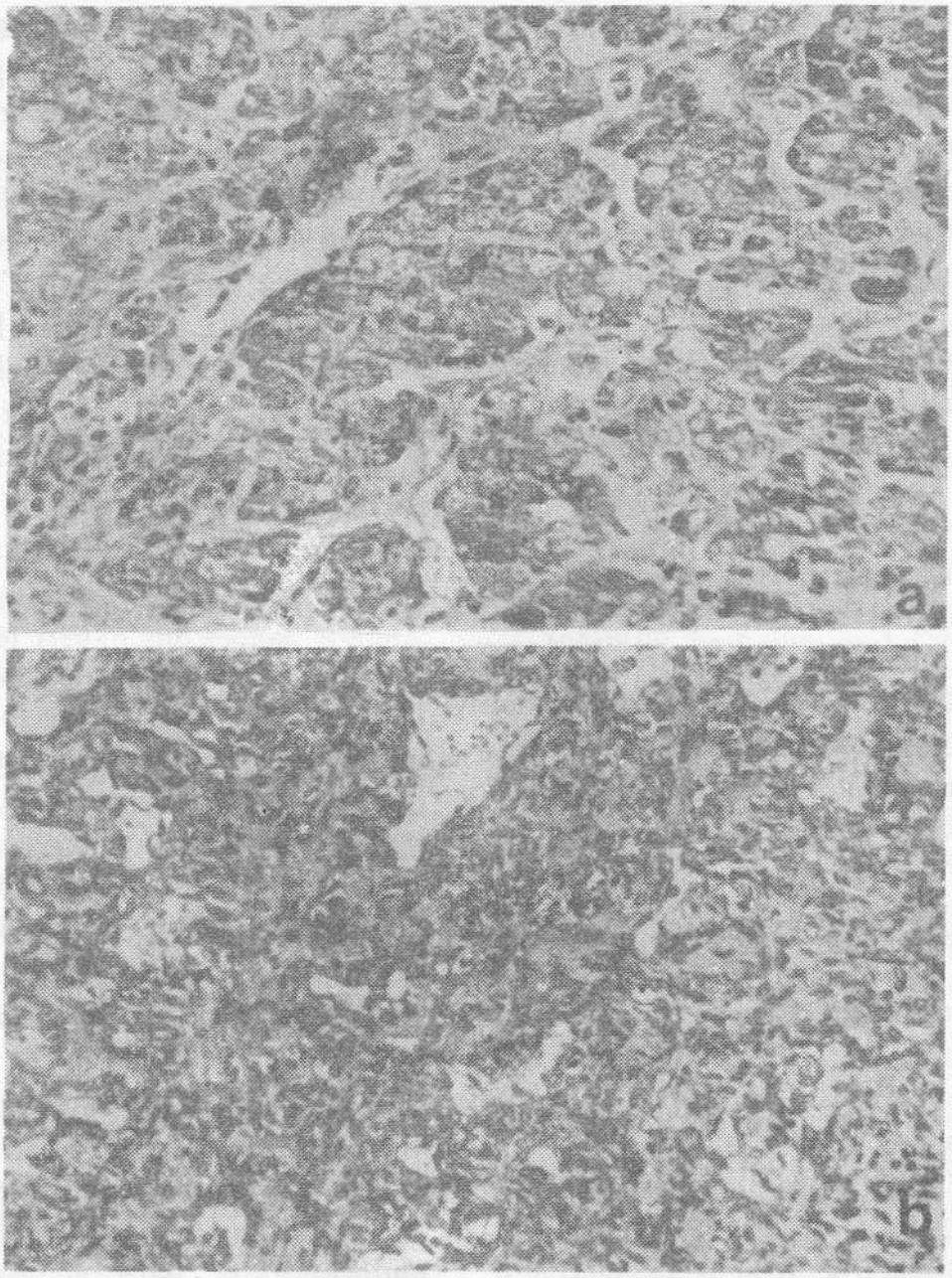

*Fig. 1. (a) Giant cell hepatitis: The parenchyma consists of multinucleated giant cells and shows infiltration by mononuclear cells (H & E × 140). (b) Hemangioendothelioma: Multiple anastomoting vascular channels lined by plump endothelial cells (H & E × 140).*

mechanical interference of bile flow from intrahepatic ducts due to compression by the tumor(4). Hemangioendothelioma around the porta hepatis would have been responsible for the compression of intrahepatic ducts and common hepatic duct in the present patient. In addition he had evidence of neonatal hepatitis (giant cell hepatitis) with cholestasis. The hepatic parenchyma also showed necrosis (cellular damage) which could be responsible for hyperbilirubinemia. Hypoproteinemia was documented and was responsible for pedal edema.

Tumors have not been stressed as differential diagnosis of hepatomegaly with hyperbilirubinemia. In case of obstructive cholangiopathy, hepatic hemangioendothelioma should be considered as one of the causes though rare. In presence of cutane-

ous hemangioma, CHF and hepatic bruit with diagnosis of hemangioendothelioma with jaundice these were present in 44.4, 55.5 and 16.6% cases, respectively(4). It is further stressed that absence of these clinical features does not rule out the presence of hepatic hemangioendothelioma. The infant reported from India had an enlarged firm nodular liver with single cutaneous hemangioma(6). These infants should be investigated further. Non-invasive techniques like ultrasonography and CT scan of the liver can delineate the tumor(4,6). Selective celiac arteriorgraphy can confirm the diagnosis of hemangioendothelioma(4). Close liver biopsy is contraindicated as the tumor is vascular. Open liver biopsy confirms the diagnosis(6). Ultrasound guided fine needle aspiration cytology can be attempted.

A majority (82.3%) of cases of hepatic hemangioendothelioma associated with hyperbilirubinemia had a fatal outcome(4). The present patient also succumbed. Our earlier experience shows that if diagnosed in life, it can be treated effectively with steriods(6). Associated CHF can be treated with concomitant decongestive therapy(2). Radiation of the tumor and surgical ligation of hepatic artery can facilitate the involution of the tumor(5,6). The prognosis can be improved upon by early diagnosis and by institution of appropriate medical treatment with steroids(6). It is suggested that as a differential diagnosis of neonatal hepatitis, possibility of hepatic hemangioendothelioma should also be thought of.

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# Spondylo-Epiphyseal Dysplasia Tarda with Progressive Arthropathy


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Spondylo-epiphyseal dysplasia tarda with progressive arthropathy is a rare inherited dysplasia with clinical resemblance


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