Case Report

An Unusual Case of Diffuse Small Blue Cell Tumor of Liver

Mudassira and Anwar Ul Haque

Department of Pathology, Pakistan Institute of Medical Sciences, Islamabad.

We report a case of diffuse small blue cell tumor of liver diagnosed on fine needle aspiration cytology (FNAC) in a 10 month old boy. He presented with chronic diarrhea and low-grade fever. On Physical and ultrasound examinations he had diffuse hepatomegally. However no discrete mass was present. The FNAC of the liver showed numerous atypical small blue cells, some of which were forming cords and trabeculae. A few macrophages with cytoplasmic vacuoles mimicking Neiman Pick cells were also seen. There was also evidence of extramedullary hematopoiesis. The Bone marrow examination revealed hyperplastic erythroid activity with few cells resembling Neiman Pick cells. Based on the cytological features of small blue cells a presumptive diagnosis of Hepatoblastoma was made.

Materials and Methods

The FNAC of enlarged liver was performed by using 23 gauge needle. The aspirate was obtained from three different sites and smears were prepared and fixed in 95% alcohol. The slides were stained by hematoxilin and eosin stain. Bone marrow aspiration was done from the right tibia; smears were made and stained with Giemsa, PAS and Sudan Black stains.

Report of The Case

A 10 month old boy presented with low grade fever, intermittent episodes of loose stools and progressive abdominal distention since birth. He had normal vitals except for temperature of 99 °F. Physical examination revealed hepatosplenomegally. Routine laboratory tests revealed WBC 15.0 x 10^9/ ul, H.B 8.6g/dl, HCT 30%, MCV 59fl, MCH 17 pg, MCHC 28 g/dl, Platelets 233 x 10^9 /ul. Peripheral blood film revealed microcytic hypochromatic anemia and leukoerythroblastic picture. Blood chemistry revealed alkaline phosphate 512U/L (range 85-365U/L), ALT 137 U/L (range 9-39U/L), Albumin 4.3 g/dl, and Triglycerides 435 mg/dl (range 59-200 mg/dl). Serum α1- fetoprotein was 7.2 ng/ml.

On ultrasound examination both liver and spleen were enlarged measuring 10 cm. FNAC of liver was performed. It revealed hypercellular smear composed of cords and sheets of atypical round blue cells. These cells had large hyper chromatic nuclei with high N/C ratio. Some cells had cleaved nuclei giving the evidence of extramedullary hematopoiesis. A few cells having foamy cytoplasm with vacuolated appearance were also seen. (Fig 2). A few normal hepatocytes were also present. No rosette formation was present.

Bone marrow aspirate showed hyperplastic of erythroid elements with mild megaloblastic change. Megakaryocytes were increased in number with left shift of maturation. A few cells resembling Neiman Pick cells having foamy cytoplasm containing clear vacuoles were seen. (Fig 3). These vacuoles were PAS and Sudan black positive. Phagocytic activity was evident by the presence of intracytoplasmic phagocytosed nuclei.

Discussion

Small, round, blue-cell tumors together comprise approximately 30% of childhood malignancies. These include neuroblastoma, non-Hodgkin lymphomas, Wilms tumors, retinoblastoma, rhabdomyosarcoma, osteosarcoma, Ewing sarcoma and hepatoblastoma. Hepatoblastoma is the most common primary hepatic malignancy in children. It is also the third most common intra-abdominal
malignancy in childhood, following neuroblastoma and Wilm's tumor.\(^3\)

FNAC is of great help in diagnosing and distinguishing small blue cell tumors of liver. This of course has important bearing on assessment for preoperative chemotherapy and radiotherapy. The small blue cells appear uniformly small in size; have rounded hyperchromatic nuclei and scant cytoplasm. The neoplastic cells have different cytomorphological patterns in different types of small blue cell neoplasia. The cells may be quite discohesive and monotonous in lymphomas. Rosettes and pseudorossettes are frequently seen in retinoblastoma and neuroblastoma. Strap cells favour rhabdoid differentiation. In the present case the cells were forming two cell thick trabeculae and about 6-7 cells long cords which suggested a diagnosis of hepatoblastoma.\(^4\)

The hepatoblastomas however in general form discrete masses and to the best of our knowledge no case of diffuse hepatoblastoma had been reported. At the time of diagnosis hepatoblastoma has an average diameter of 10-12 cm. The tumor most often is a unifocal, well-circumscribed mass, but it may be multinodular. When presenting as a solitary mass, the right lobe is more commonly affected.\(^5\) but in our case the presentation was of diffuse enlargement of liver. Diagnostic evaluation of hepatoblastoma may reveal normochromic normocytic anemia and thrombocytosis on peripheral blood film and complete blood counts.\(^6\) Evidence of extramedullary hematopoeisis can also seen which often accompanies hepatoblastoma.\(^7\) Liver enzymes are moderately elevated. Serum \(\alpha\)-fetoprotein levels are often elevated but low levels do not exclude the diagnosis of hepatoblastoma.\(^8\) A presumptive diagnosis of diffuse small blue cell tumor of liver favoring hepatoblastoma was made. The definite diagnosis required liver biopsy and CT scan but the patient could not be followed further.

Gaucher cells are known to be associated with round blue cell tumors. However such association is rarely found for Neiman pick cells. The cells with cytoplasmic clearing and vacuolations seen in liver aspirates as well as in bone marrow aspirates could be lipid laden macrophages, the lipid being derived from the cell membranes of phagocytosed cells. Such storage type cells can be seen in reactive conditions and hemorrhages.

The current case was of great interest as the small blue cells strongly suggested the possibility of hepatoblastoma as there was no evidence of other small blue cell tumors including neuroblastoma, Wilms tumor and leukemia and lymphoma. Neiman Pick type cells seen in liver and bone marrow could be reactive macrophages. Unfortunately the patient left against medical advise and several confirmatory tests including Splenic FNAC could not be carried out.
References


5. Mary Elizabeth McCarville, MD: Hepatoblastoma (online) Last Updated: March 8, 2005 Available from URL: http://www.emedicine.com/ped/PEDIATRICS.htm

