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Small cells indifferentiated hepatoblasotma in childhood: a rare aggressive malignancy



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A new born of two months years old was admitted for abdominal mass which the echography confirmed the hepatic origin. The biological analysis showed an elevated Lactic Acid Dehydrogenase (LDH) serum level, but no elevated alpha fetoprotein neither catecholamins nor other marquers. The abdominal scan confirmed hepatic mass at the expense of two segments. The fine needle scan guided biopsy was done and revealed a neoplastic proliferation of cells, histologically slightly larger than lymphocytes and measure (7-8 µm) in size. They are round to oval with scant cytoplasm, relatively fine nuclear chromatin, inconspicuous nucleoli and only minimal mitotic activity. They grow in a diffuse pattern but usually form clusters either intimately inter-mixed with other epithelial cell

types or forming nests in an almost "organoid" pattern (A). Immunohistochemically, small cells showed positivity for pancytokeratin (B) as well as vimentin (D). They did not express alpha-fetoprotein (AFP) (C). Thus, the final diagnosis been retained: small cell undifferentiated hepatoblastomas (SCUD-HB) is associated with an aggressive biological course and poor survival. The negative prognostic impact is conferred even by small areas in an otherwise large tumor. Rarely, particularly in infants, the entire hepatoblastoma is composed of small undifferentiated cells; such tumors account for approximately 5% of all hepatoblastomas. SCUD-HB is associated with low or normal serum AFP levels. SCUD-HB retain expression of INI1 but a small subset present morphological rhabdoid features, including loss of nuclear expression of INI1. Transition between the two types has been described.



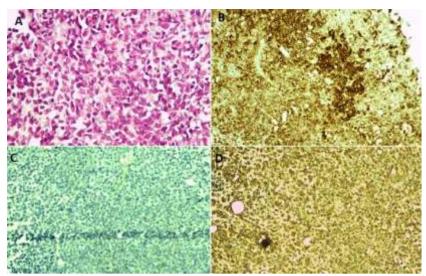


Figure 1: A) HEx20 neoplastic proliferation of small tumoral cells slightly larger than lymphocyte measuring (7-8 μ m) in size. They are round to oval with scant cytoplasm, relatively fine nuclear chromatin, inconspicuous nucleoli and only minimal mitotic activity; B) they grow in a diffuse pattern, immunohistochemicaly, the small cells stained positive for pancytokeratine; C) negative for alfa-foetoprotein; D) positive for vimentine