Hyaline Globules In Ascitic Fluid In Primary Hepatic Undifferentiated Embryonal Sarcoma

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Citation

M Dharan, H Yodko. *Hyaline Globules In Ascitic Fluid In Primary Hepatic Undifferentiated Embryonal Sarcoma*. The Internet Journal of Pathology. 2004 Volume 4 Number 1.

Abstract

Undifferentiated Embryonal Sarcoma (UES) of the liver is a rare predominantly pediatric malignancy. Only one brief report of cytologic findings in ascitic fluid of a recurrent tumor is available. The characteristic eosinophilic hyaline globules (HG) with immunohistochemistry on fluid cytology, has not previously been documented.

A 20-year-old male presented with ascites. Cytology revealed a sparse number of pleomorphic malignant cells. Sections of the cell-block preparation revealed a loose myxomatous background and numerous atypical cells with marked pleomorphism, multinucleation and many intra- and extra- cytoplsmic eosinophilic HG that were periodic acid-Schiff (PAS) positive and diastase-resistant. They displayed a positive reaction to alpha-1-antitrypsin on immunoperoxidase staining. A needle biopsy of a liver mass detected by CT scan confirmed the diagnosis of UES.

Detection of sarcoma cells on fluid cytology is not often difficult. But an accurate diagnosis can be achieved only by detailed immunohistochemistry. Preparation of cell-blocks is of value in the work-up and management of patients presenting with malignant effusion.

INTRODUCTION

Undifferentiated embryonal sarcoma (UES) of the liver is a rare neoplasm that occurs mainly in pediatric patients, but occasionally has been reported in adults[1,2,3]. The tumor has a myxomatous stroma with a markedly pleomorphic cellular component and many multinucleated and bizarre cells. Numerous eosinophilic hyaline globules (HG) appearing both intra- and extracellularly are a characteristic feature of UES. The HG are PAS- positive, diastase-resistant and stain positively for alpha-1-antitrypsin by immunocytochemistry. These features can be readily recognized on FNA cytology which has an important role to play in the pre-operative diagnosis and management of these patients[4,5]. Allen et al has described the cytologic findings from peritoneal washings in a recurrent UES[6]. HG were not present in that report. A detailed description of cytomorphology of this rare entity in serous fluid cytology has not appeared in the literature.

CASE REPORT

In our case the patient, a 20-year-old man, presented with ascites. The cytologic smears were blood –stained and cellularity was scant. Isolated multinucleated, bizarre cells with clear abundant cytoplasm displayed indistinct borders. Cytoplasmic vacuoles were seen in large mononuclear cells. Nuclear chromatin was coarsely clumped and nucleoli were distinct. A few cells showed smudged nuclei. Of special interest were sparse multinucleate jelly-fish-like bizarre cells with transparent cytoplasmic tentacles (Fig 1 A,B).

Figure 1

Figure 1: Cytologic appearance of UES in ascitic fluid smear. 1A: A multinucleate tumor cell with abundant, clear cytoplasm(Papanicolaou x 600). 1B: Prominent nucleoli (arrows) are visible in this bizarre " jelly-fish"-like cell (Papanic

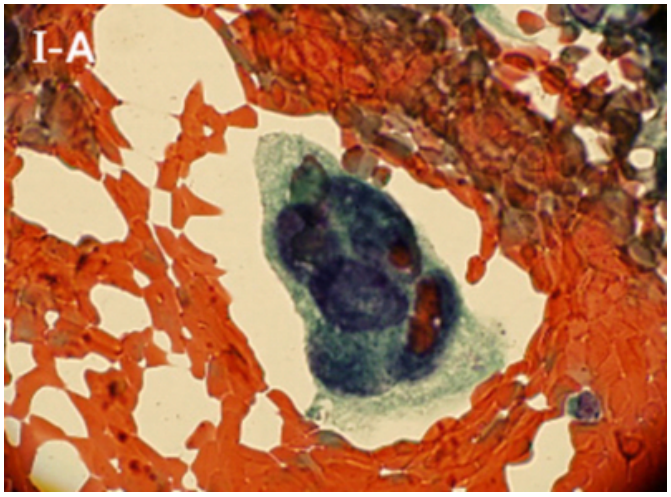
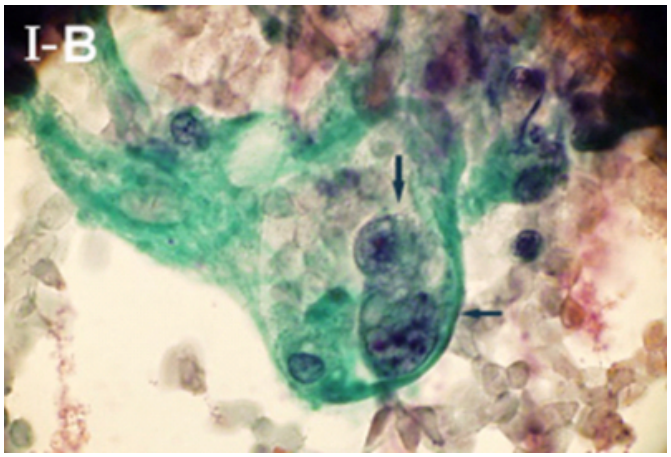


Figure 2



Sections of the cellblock revealed neoplasm with a myxomatous background with a sprinkling of inflammatory cells. Numerous bizarre atypical cells, many of them bi- or multi-nucleated, were conspicuous. In addition, large atypical mononuclear cells were seen. Numerous eosinophilic hyaline globules of varying sizes appeared intra- and extra-cellularly (Fig 2A). Eosinophilic material was also seen in membranous formations. These eosiniphilic deposits stained positive with PAS and were diastase resistant. Similar positive reaction was noted with immunoperoxidase staining for alpha-1- antitrypsin (AAT) (Fig 2C). The neoplastic cells also stained strongly positive for vimentin (Fig 2B) and negative for cytokeratin (CK), leukocyte common antigen (LCA), desmin, S100, alpha-fetoprotein (AFP) and carcino-embryonic antigen (CEA).

Figure 3

Figure 2: Cell block of ascitic fluid. 2A: Pleomorphic sarcoma cells in a myxomatous background. Inset shows an intracytoplasmic eosinophilic HG (Hematoxylin- eosin x 200). 2B: strong vimentin positivity of the tumor cells (Immunoperoxidase X 200). 2C: Intracytoplasmic AAT-positive HG (Immunoperoxidase X 600).

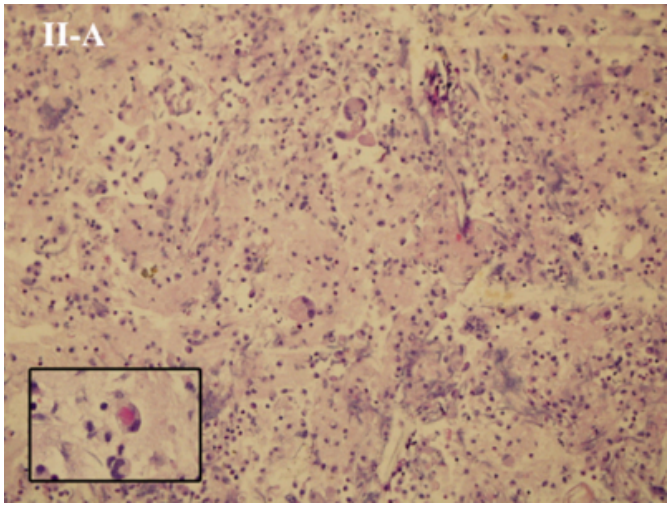


Figure 4

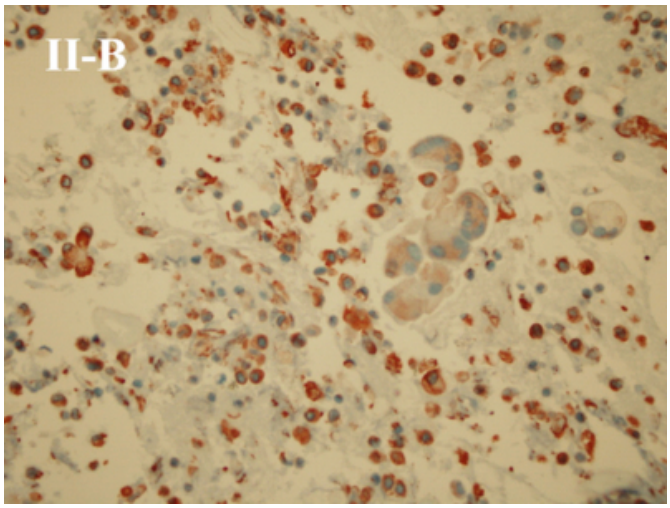
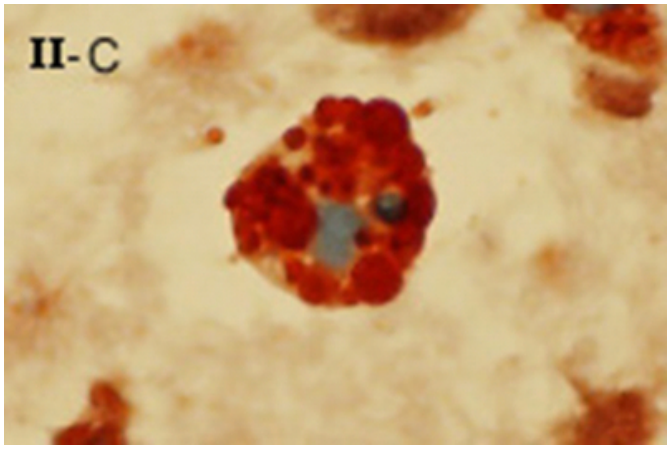


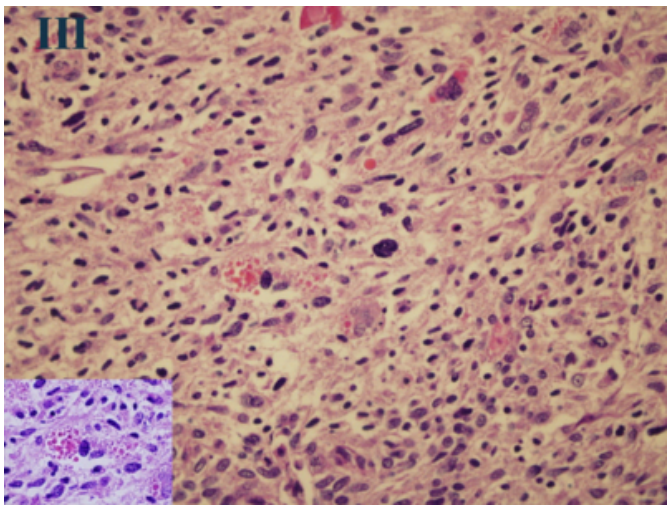
Figure 5



Based on the above findings a diagnosis of UES was made. A CT scan of the abdomen showed an enlarged liver with a mass in the right lobe. A needle biopsy confirmed the diagnosis (Fig 3).

Figure 6

Figure 3: Histologic section of biopsy from UES of the liver. Note the marked pleomorphism of tumor cells (Hematoxylin-eosin X 200). Inset shows numerous intracytoplasmic HG (X 600).



DISCUSSION

A few publications have described the cytological features of UES of the liver on FNA[4,5], but only one partial presentation of cytomorphology of the cells in serous fluid has been reported[6]. A diverse pattern of differentiation, including partial myogenic phenotype has been noted in different studies. The myogenic features seem to be more prominent in adult cases[3]. Atypical pleomorphic cells, though sparse, were consistent with a malignant mesenchymal tumor. Sarcomas account for only 3-6% of malignant effusions[7]. Usually the diagnosis is facilitated by knowledge of the primary neoplasm. However, the cells often exhibit a variety of morphologic features that can differ from those of the primary neoplasm which may preclude the correct cytologic diagnosis[7]. Generally non-lymphomatous sarcomas share certain features in cytologic smears such as sparse, single cells, multinucleation, indistinct cell borders, pleomorphism and proteinaceous background[7]. In the appropriate clinical setting cytologic sub-classification of sarcomas may be attempted. However, an accurate diagnosis usually requires immunocytochemistry. In laboratories where Thin Prep technique is not available, cell-block preparation will be of great value for this purpose. In a recent publication Selvaggi[8] demonstrated the added benefit of cell-block preparations in the cytologic analysis of peritoneal washings. Certain histologic aspects of the lesion may be revealed in cell-block preparations. More importantly, specific diagnostic features can be easily recognized and verified by special staining methods, as was possible in this case.

The differential diagnosis of sarcomas in fluids has been discussed[7]: consideration should be given to exclude anaplastic carcinoma, malignant melanoma, mesothelioma and large cell lymphoma; a panel of CK, LCA, vimentin and S100 will be helpful in this process. Further identification of specific neoplasms will require markers such as HMB-45, desmin and smooth muscle actin[7,9].

Eosinophilic material in the form of HG and membranous deposits was the most striking finding in this case. In a female patient, similar structures with positive staining for AFP can be observed in certain ovarian germ cell tumors[10].

From a clinical viewpoint, cytologic diagnosis of UES is important because preoperative chemotherapy approach may prove to be useful in this highly malignant neoplasm[11].

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