

## Fine needle aspiration cytology of undifferentiated embryonal sarcoma of the liver: a case report

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### Abstract

The cytological features of a rare case of undifferentiated (embryonal) sarcoma of the liver are presented. The cytology smears showed singly dispersed polygonal and spindle cells as well as loose clusters of cells held together in myxoid material. Neoplastic cells were generally large with round, oval or lobulated nuclei. The cytoplasm was variable in amount with ill-defined borders. Occasional multinucleated cells were also present. Hyaline globules were present on sections of the cell block. Immunohistochemical studies performed showed positivity for vimentin, cytokeratin and alpha-1-antitrypsin (AAT) in the tumour cells.

*Key words:* liver sarcoma, fine needle aspiration, undifferentiated embryonal sarcoma.

### INTRODUCTION

Undifferentiated (embryonal) sarcoma of the liver (UESL) is a rare malignant mesenchymal hepatic tumour with an incidence among liver tumours of 27.7%.<sup>1</sup> It occurs predominantly in children under the age of 15 years, with a peak incidence between 6 and 10 years. The prognosis is variable, although the mean survival is around 11 months.<sup>1</sup>

The term “undifferentiated (embryonal) sarcoma of the liver” was first used by Stocker and Ishak in 1978.<sup>2</sup> Although cases have been reported in literature<sup>3-5</sup> only a few described cytological findings in fine needle aspiration biopsy material.<sup>6-11</sup> Histologically, these tumours show features that mimic histiocytoid, rhabdoid, leiomyoblastic or fibroblastic tumours. Immunohistochemical studies show that UESL frequently expressed positivity for vimentin and occasionally, for cytokeratin, antichymotrypsin (ACT) and alpha-1-antitrypsin (AAT). In this report we describe the fine needle aspiration cytological characteristics of this rare tumour.

### CASE REPORT

A six-year-old Malay girl presented with a two week history of low grade fever and right hypochondrial pain. There was no history of jaundice, no loss of appetite and weight. Physical examination showed a thin and febrile girl with a temperature of 38.4°C. There was no

pallor, jaundice or any stigmata of chronic liver disease. The liver was enlarged up to seven cm below the costal margin and was firm and tender. There was no splenomegaly or ascites. Abdominal ultrasound showed a mass in the right lobe of the liver with multiple hypoechoic areas. A CT scan confirmed a liver mass in segment V and VI extending medially and crossing the midline. Ultrasound guided fine needle aspiration cytology was carried out. Direct smears taken were fixed in 95% alcohol and stained with Papanicolaou stain. A cell block was also prepared and sections cut and stained with Haematoxylin and Eosin (H&E). A cytological diagnosis of an undifferentiated embryonal sarcoma of the liver was made based on the cytological features and immunohistochemical findings. Four weekly courses of chemotherapy were given using the protocol consisting of vincristine, adriamycin, cyclophosphamide and actinomycin-D. The tumour showed partial response to chemotherapy as indicated by CT scan. An extended hepatectomy was performed leaving only the left lobe of the liver behind. At operation, the tumour appeared to be localised in the liver with no lymph node involvement. A repeat CT scan two weeks after completion of chemotherapy revealed no focal lesions in the left lobe. There were no abnormal masses elsewhere. The patient is well at the time of this writing.

**Cytological features**

The smears were generally sparsely cellular and comprised of large bizarre cells which were distributed singly and in loose groups held together by a myxoid stroma (Fig. 1). The single cells were of variable shapes. Some were spindly with plump, oval nuclei and prominent

nucleoli and others were polygonal, rounded, lobulated or multinucleated (Fig. 2). The cytoplasm was moderate to abundant in amount with poorly defined borders. Some pleomorphic bare nuclei were also present in the background which showed abundant myxoid material. No hepatic cells or **extramedullary** haematopoietic cells were observed. Examination of sections from the cell

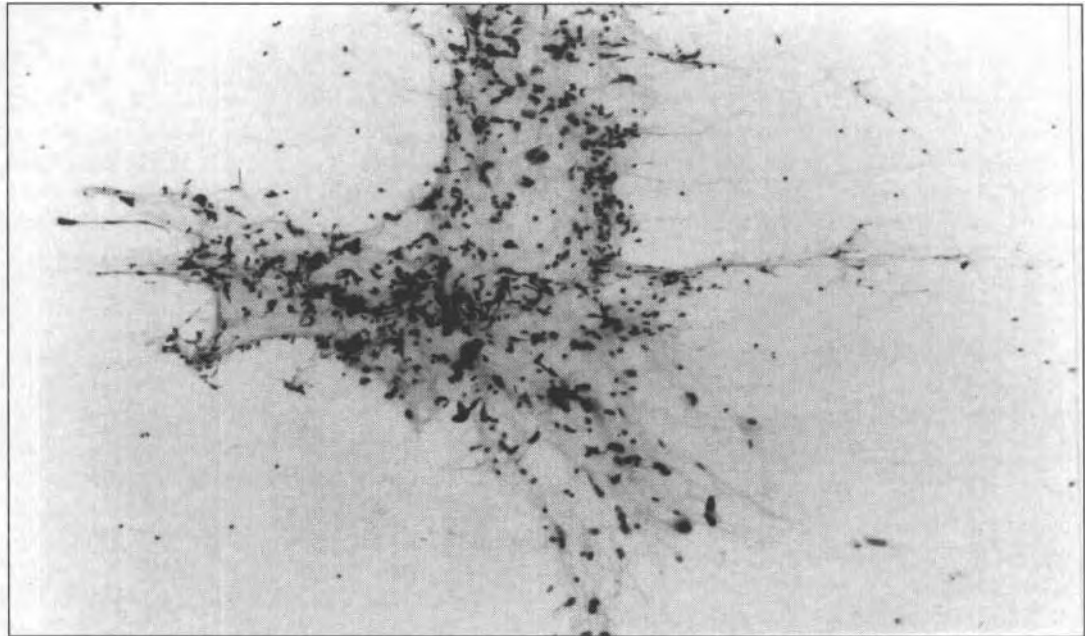


FIG. 1: Smear shows malignant cells loosely held together in myxoid stroma. Papanicolaou stain x 100.

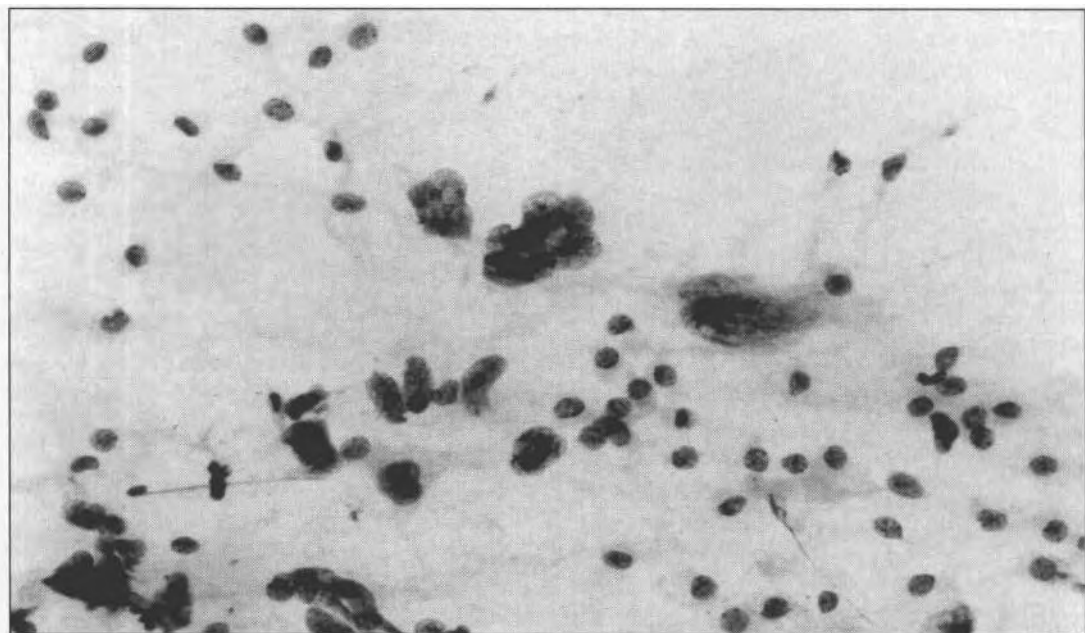


FIG. 2: Pleomorphic malignant cells stripped of cytoplasm displaying coarse nuclear chromatin, one of which is multinucleated. Papanicolaou stain x 400.

block showed an undifferentiated tumour composed of primitive looking cells in loose myxomatous stroma. Also noted was prominent vacuolisation (Fig. 3) of the stromal tissue and a few interspersed spindly and stellate neoplastic cells. Small rounded and distinct hyaline globules were seen (Fig. 4), some outside the

tumour cells and some within the cells. Immunohistochemical studies showed the tumour cells to be immunopositive to alpha-1-antitrypsin (AAT), vimentin and cytokeratin. A cytological diagnosis of undifferentiated embryonal sarcoma of the liver was given.

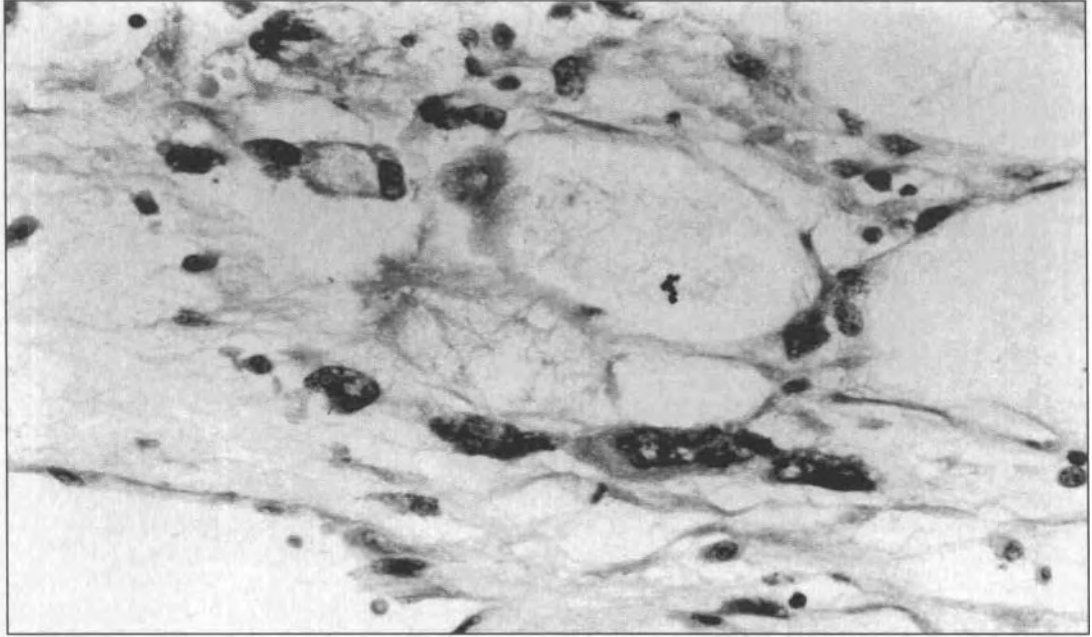


FIG. 3: Section from cell block showing malignant cells interspersed in loose vacuolated stroma. H&E x 400.

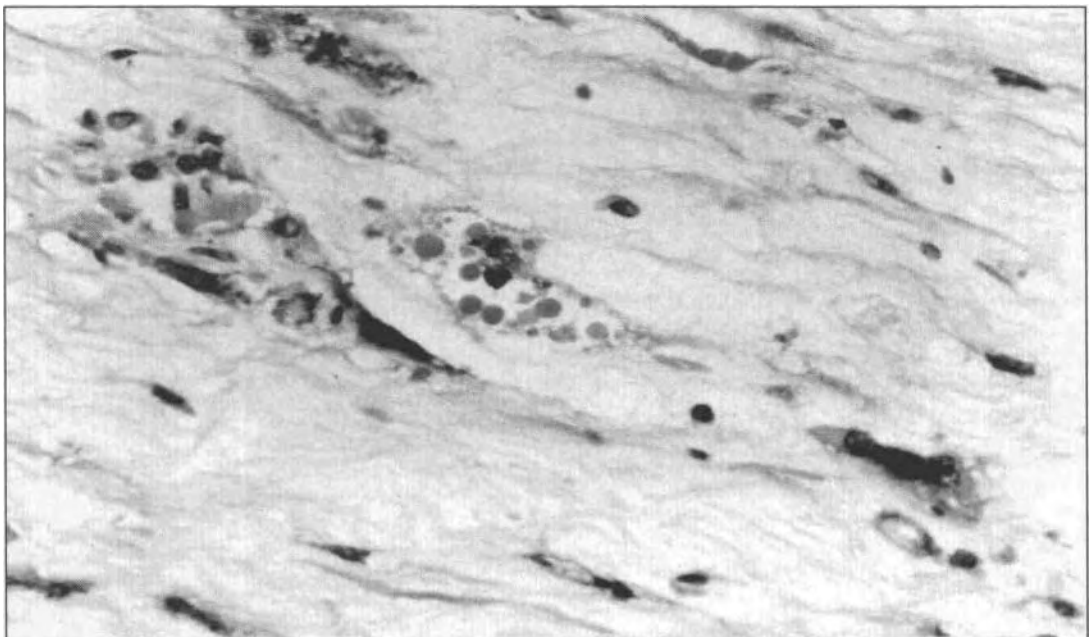


FIG. 4: Section from cell block showing small rounded hyaline globules. H&E x 400.

### Pathological features

Macroscopically the tumour appeared lobulated, measuring 12x10.5x6cm in diameter. A pseudocapsule separated the tumour from the adjacent normal liver. The cut surface showed solid and cystic areas, areas of haemorrhage and necrosis.

Histological sections showed large areas of necrosis with residual tumour tissue present only at the periphery. The tumour cells were generally spindle or stellate, with some multinucleated cells and were loosely arranged in an abundant myxoid background. Most distinct however, was the presence of numerous sharply defined, eosinophilic hyaline globules of various sizes which were periodic acid-Schiff positive and resistant to diastase digestion. Small groups of residual normal hepatocytes were seen in between the tumour cells and areas of haemorrhage. The surgical margins were free of tumour. Immunohistochemical stains demonstrated alpha-1-antitrypsin (AAT) in some cells and vimentin and cytokeratin in all tumour cells.

### DISCUSSION

UESL is a rare neoplasm that occurs almost exclusively in children.<sup>12</sup> No sexual predilection, racial predominance or geographic localisation has been identified.<sup>2</sup> The chief presenting complaint in the majority of patients is an abdominal mass with or without right upper quadrant pain.<sup>2</sup> Uncertainty as to the histogenesis of this entity has resulted in many synonyms for it, including malignant mesenchymoma, fibromyxosarcoma and lipofibrosarcoma.<sup>2</sup> UESL is now considered to be a distinct clinicopathological entity. The histological features of UESL have been well described; some reports describing cytomorphological characteristics are also available.<sup>6-11</sup>

The differential diagnosis of UESL include other primary paediatric hepatic neoplasms such as hepatoblastoma, hepatocellular carcinoma and sarcomatoid lesions of the liver in childhood such as embryonal rhabdomyosarcoma (intrahepatic), angiosarcoma, fibrosarcoma and malignant fibrous histiocytoma. In hepatoblastoma, the tumour cells are usually smaller, are arranged in cohesive clusters and acinar pattern<sup>13</sup> and are usually immunopositive to alpha-fetoprotein. Hepatoblastomas lack the myxoid stroma and marked pleomorphism which characterise UESL. Hyaline globules are also absent in hepatoblastomas. Childhood hepato-

cellular carcinomas are unlikely to be a source of confusion since these tumours are distinctly epithelial.<sup>14</sup>

Differential diagnostic problems may arise occasionally with UESL and rhabdomyosarcoma (embryonal), angiosarcoma, fibrosarcoma and malignant fibrous histiocytoma. This lies in the fact that some areas of the tumour may present histiocytoid, rhabdoid, fibroblastic or leiomyoblastic differentiation giving these tumours areas of great cellular heterogeneity histologically.' The phenotypic diversity of UESL has also been documented in immunohistochemical and ultrastructural studies.<sup>3,4</sup> Positivity for vimentin indicates a myogenous differentiation of these tumours.<sup>15</sup> In UESL, co-expression of vimentin and cytokeratin is observed. Occasionally ACT and AAT are present but AFP is rarely expressed.

The prognosis of these tumours is usually considered to be very poor. Median survival is in the range of 12 months. Resection before or after chemotherapy improves the prognosis,<sup>16</sup> hence the importance of recognising the cytological features of this tumour so that early diagnosis and treatment can be facilitated. The most distinctive cytological features of UESL are spindle, stellate, pleomorphic and multinucleated giant cells, hyaline globules and a myxoid matrix. The cytopathological features together with immunohistochemical findings in this case were sufficient to permit a confident cytological diagnosis.

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