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CONTINUING MEDICAL EDUCATION
ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Hematology Quiz – Case 71

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2025, 42(5):718–720

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A 15-year-old girl was admitted for the evaluation of a large abdominal mass. Imaging studies showed a large paraaortic mass, 10.0 cm by 8.0 cm by 3.0 cm that surrounded the aorta and infiltrated the left perirenal adipose tissue and left adrenal. Laboratory tests were notable for a haemoglobin level of 10.4 g/dL, a white-cell count of 2.7×10^9/L (neutrophil count: 1.2×10^9/L), a platelet count of 95×10^9/L, a lactate dehydrogenase (LDH) level of 914 U/L (normal range: 125–220), and serum ferritin 2,914 ng/mL (normal range: 4.63–204). A peripheral blood smear showed no circulating immature cells. A bone-marrow aspirate was obtained for evaluation of the pancytopenia, and showed replacement of the marrow by abnormal cells (figures 1–7; May-Grünwald-Giemsa stain). On flow cytometric analysis, the cells were CD3-, CD19-, CD33-, CD13-, CD56+, CD57+, CD81+, and CD117+.

Comment

The bone marrow is totally replaced (diffuse infiltration of the bone marrow) by monomorphic small- or medium-sized atypical round cells that have a high nuclear:cytoplasmic ratio, immature chromatin, and inconspicuous nucleoli resembling leukaemic blasts (primitive appearance). The most striking morphologic feature is the circular arrangement of the cells around an amorphous grey core (figures 6, 7). This structure is consistent with the Homer Wright rosette seen in neuroblastoma. Homer Wright rosettes are composed

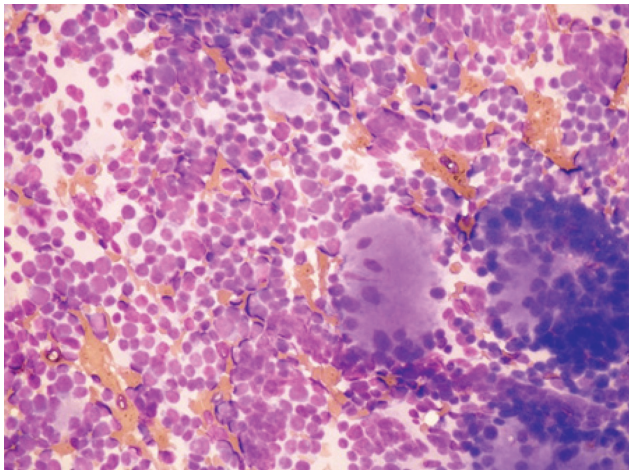

Figure 1.

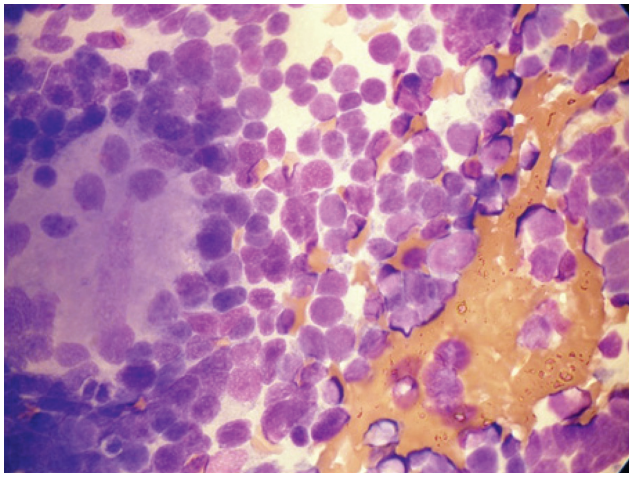

Figure 2.

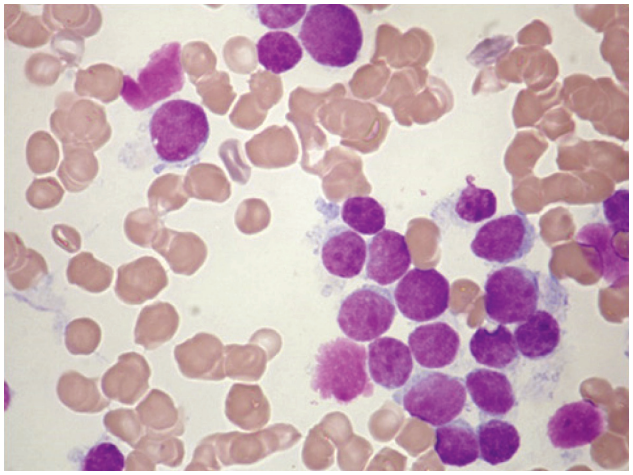

Figure 3

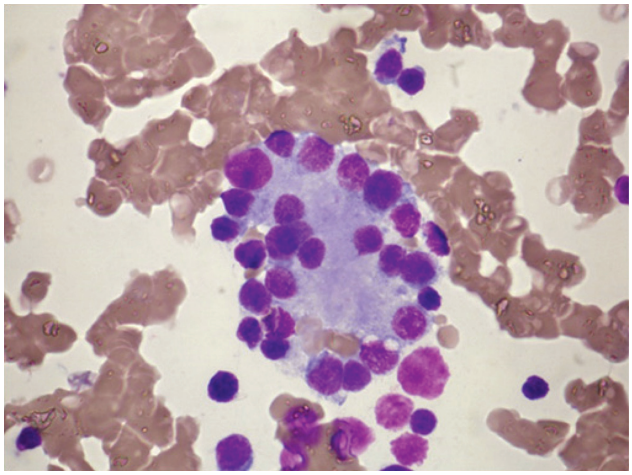

Figure 6

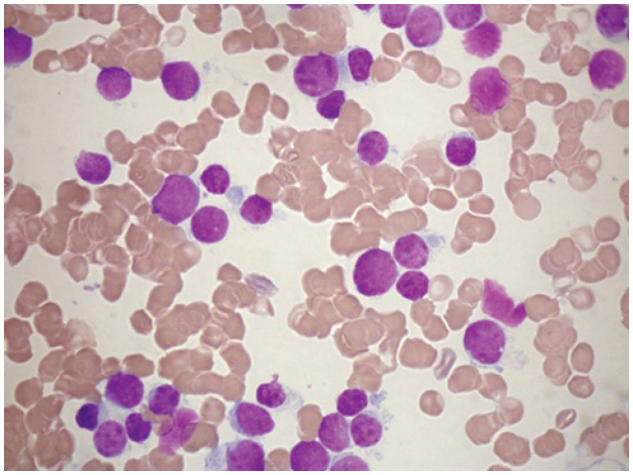

Figure 4

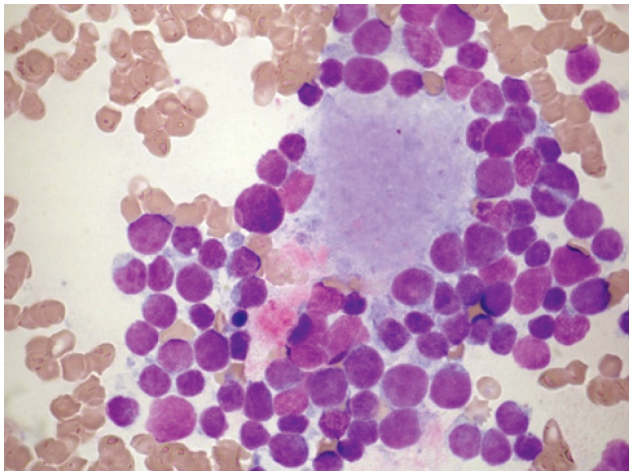

Figure 7

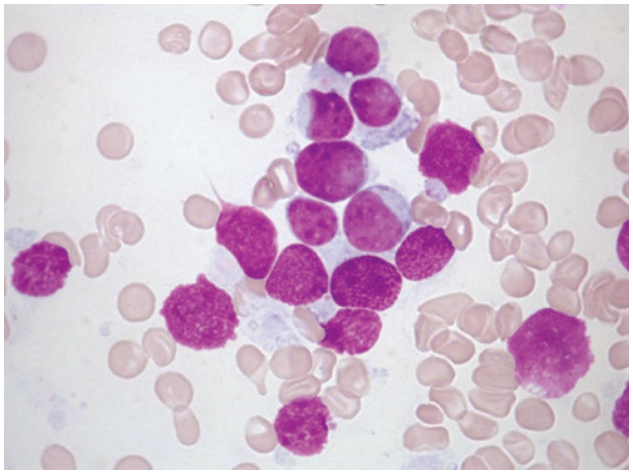

Figure 5

of tumor cells surrounding a central core of neurites/neurofibrillary material (neuropil). Another clue to metastatic neuroblastoma is the stippled nuclear chromatin of the blast cells which has been described as "salt and pepper chromatin".

Neuroblastoma is an embryonic tumour of the autonomic nervous system. The tumor arises in tissues of the sympathetic nervous system, typically in the adrenal medulla or paraspinal ganglia and, thus, can present as a mass in the neck, chest, abdomen, or pelvis. Neuroblastoma is one of the most common solid tumors of childhood. Median age at diagnosis is two years. There is a sharp decline in its incidence for patients between 14 and 18 years old or older than 18 years. Older children suffer from more aggressive tumours and have a worse prognosis than younger children.

Homer Wright rosettes were named after James Homer Wright (1869–1928) who wrote his classic report when he was director of the pathology laboratory of Massachusetts General Hospital. He

described a group of adrenal and sympathetic nervous system tumours, which became known as neuroblastomas, with characteristic ball-like arrangements of tumor cells that enclosed meshworks of fibers. James Homer Wright also developed Wright's stain. Homer Wright rosettes are seen in 25–30% of neuroblastomas.

Small round cell tumors of childhood, also called small round blue cell tumours of childhood, is a term used to describe a group of malignant neoplasms affecting children and adolescents. They include neuroblastoma, rhabdomyosarcoma (fig. 8), primitive neuroectodermal tumors (PNET), Ewing's sarcoma and Burkitt lymphoma.

The small round cell tumors of childhood share common morphologic features and sometimes immunophenotypic characteristics, making the differential diagnosis challenging. They should be considered in the differential diagnosis of young patients presenting with extensive bone marrow infiltration mimicking acute leukaemia and atypical/unusual blasts. Appropriate treatment demands accurate diagnosis.

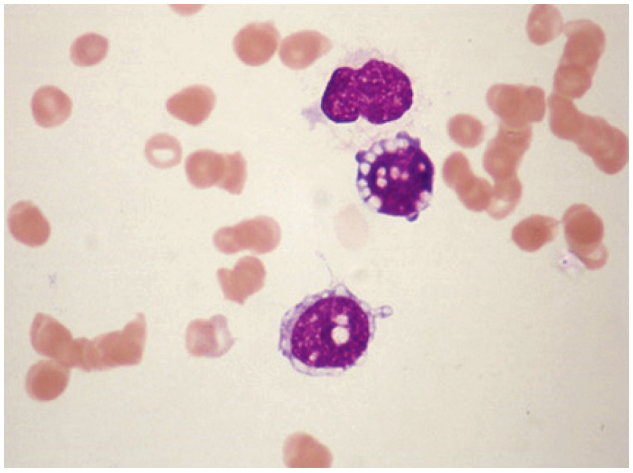

Figure 8

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