Morphology Update

Circulating lymphoma cells in intravascular large B-cell lymphoma

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A 92-year-old Caucasian man who was dialysis dependent due to autosomal dominant polycystic kidney disease presented with thrombocytopenia and weight loss. On examination, his spleen was palpable 6cm below the left costal margin but there was no palpable lymphadenopathy. His blood count showed a stable hemoglobin concentration of 101 g/l but a markedly reduced platelet count of $44 \times 10^9/l$ and a white cell count of $19.3 \times 10^9/l$. An automated differential count suggested that there was monocytosis and lymphocytosis but a blood film showed that the abnormal cells were all lymphoid, with the count being neutrophils $3.8 \times 10^9/l$, lymphoid cells $12 \times 10^9/l$, monocytes $0.6 \times 10^9/l$, and eosinophils $0.1 \times 10^9/l$. The abnormal cells were medium to large with a high nucleocytoplasmic ratio, a single large but indistinct nucleolus and moderately basophilic cytoplasm (top left). Flow cytometric immunophenotyping on peripheral blood showed the abnormal cells to be large B cells expressing CD19, CD20, CD79b and BCL2 with kappa light chain restriction. They were negative for CD5, CD10, CD23, CD43 and CD200. The proliferation fraction (Ki-67 positive) was 67%. A bone marrow aspirate was hemodilute, with some maturing hematopoietic cells but also lymphoid cells similar to those seen in the peripheral blood. Trephine biopsy sections were hypercellular for the patient's advanced age and showed atypical medium to large cells, with prominent nucleoli, within sinusoids and capillaries (top right, hematoxylin and eosin, $\times 40$ objective). Immunohistochemistry for CD20 (bottom left) confirmed that the abnormal cells were largely intravascular with smaller numbers of single cells and small aggregates in the interstitium. The neoplastic cells also expressed CD19 and CD79a while CD10, MUM1/IRF4 and EBER were negative. CD34 staining of endothelial cells confirmed their intravascular location (bottom right). The patient was given a trial of high dose corticosteroids but died shortly afterwards.

The prognosis of intravascular large B-cell lymphoma is usually adverse, at least in part due to the difficulty in making the diagnosis. In our patient the unusual presence of circulating cells led to examination of the bone marrow. Despite the speedy diagnosis, treatment in this elderly dialysis-dependent man was unsuccessful.

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