MORPHOLOGY UPDATE

An aggressive NK-cell lymphoma with hemophagocytosis

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A previously fit 49-year-old man was admitted with a short history of drenching night sweats, fever and weight loss. On examination he looked unwell but no lymphadenopathy or organomegaly was detectable. His blood count showed Hb 133 g/L, WBC 4.3 × 109/L, and platelets 56 × 109/L. The blood film showed no specific features. He developed progressive liver failure, coagulopathy, hyperferritinemia, and hypertriglyceridemia. Serum lactate dehydrogenase was 4503 iu/L. Epstein–Barr virus (EBV) cell-free DNA was measured at 6.35 log 10 iu/ml. Computed tomography of neck, thorax, abdomen, and pelvis showed small bilateral pleural effusions with no evidence of lymphadenopathy, splenomegaly or hepatomegaly, although the liver had a diffusely abnormal texture. His clinical condition deteriorated rapidly, necessitating intubation, ventilation, and vasopressor support. A bone marrow aspirate showed markedly increased cellularity with prominent hemophagocytosis (upper images, ×100 objective). In addition there was a population of large, nucleolated lymphoid cells with finely granular, moderately basophilic cytoplasm (lower images, ×100). The latter were shown to have a CD45+, CD2+, CD56+ phenotype by flow cytometry; other surface T-cell markers and CD30 were not expressed. Cytogenetic analysis showed 92,XXYY,t(3;13)(q12;q14)[6]/46XY[4].

The diagnoses to be considered here are aggressive NK-cell leukemia and extranodal NK/T-cell leukemia/lymphoma. Some overlap between these diagnoses is recognized. We favor the diagnosis of aggressive NK-cell leukemia because of the bone marrow involvement and the associated hemophagocytosis. Both of these are rare in extranodal NK/T-cell leukemia/lymphoma, being seen in only one of 101 patients in one series.1 The patient was treated with high dose corticosteroids and cyclophosphamide but he developed multi-organ failure and died soon thereafter.

Aggressive NK-cell leukemia is a rare EBV-associated lymphoproliferative disorder, most prevalent in East Asian populations. Frequently, its presentation is complicated by multi-organ dysfunction, coagulopathy and hemophagocytosis. Numerical and structural cytogenetic abnormalities have been reported.  This neoplasm is frequently treatment refractory and the prognosis is dismal with a median survival of less than two months.

Conflict of interest

Nil

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REFERENCE

1. Su Y-J, Wang H, Chang H, et al. (2018) Extranodal NK/T‐cell lymphoma, nasal type: Clinical features, outcome, and prognostic factors in 101 cases. *Eur J Haematol*. 2018;101:379-388.

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