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ONLINE INTERESTING MEDICAL IMAGE

Rare Presentation of Aggressive T/Natural Killer Lymphoproliferative Disorder in a Leukaemic Phase in an Omani Patient

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عرض نادر لمريض عماني مصاب بلمفوما الخلايا اللمفاويه الطبعيه القاتله

اروى الريامیه، خليل الفارسي، ساهيمه المعمريه، احمد الفرقاني، مرتضى الخابوري

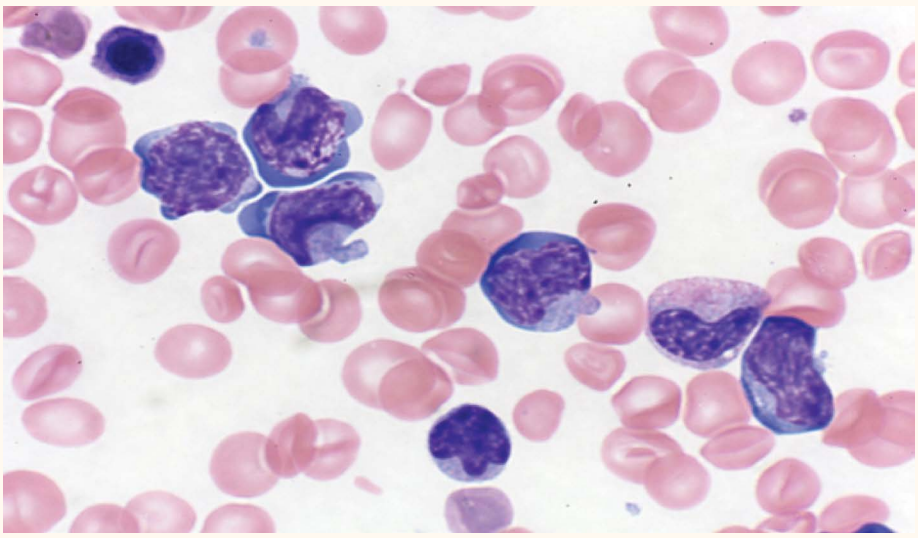

Figure 1: Blood film showing large lymphoid cells with prominent nucleoli and deep basophilic cytoplasm.

Natural killer (NK) cells are cytotoxic cells that target tumour, bacteria- and virus-infected cells.[1] They produce cytokines such as interferon-γ and tumour necrosis factor-α. Morphologically, NK cells appear as large granular lymphocytes with abundant pale cytoplasm with azurophilic granules. They are negative for surface CD3; positive for CD16, CD56 and CD57, and variably express some T cell antigens like CD2, CD7 and CD8.[2] NK and T/NK lymphomas are very rare, and are mainly described in areas of East Asia, where they account for 7–10% of all lymphomas.[3] They are commonly associated with the presence of Epstein-Barr virus in the neoplastic cells. To the best of the authors' knowledge, no cases from the Gulf region have been reported in the medical literature. Herein, is a description of a presentation of this rare disease in an Omani patient.

A 46-year-old Omani female with schizophrenia presented to the Department of Haematology at the Sultan Qaboos University Hospital in Muscat, Oman, in January 2013. She was referred for evaluation of bilateral adrenal masses, abdominal lymphadenopathy, hepatosplenomegaly, transaminitis and elevated lactate dehydrogenase levels at 540 u/L (normal range: 135–225 u/L). She had a two-week history of vomiting, abdominal pain and jaundice. A complete blood count revealed a total white cell count of 17.0 x 10^9/L (normal range: 2.4–9.5 x 10^9/L), lymphocytosis of 11.4 x 10^9/L (1.2–3.8 x 10^9/L), haemoglobin of 6.6 g/dL (11.0–14.5 g/dL) and a platelet count of 48 x 10^9/L (150–450 x 10^9/L). The patient's blood film showed medium to large lymphoid cells with prominent nucleoli and deep basophilic cytoplasm with occasional vacuolation and cytoplasmic granulation [Figure 1].

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Peripheral blood flow cytometry showed that T/NK cells made up 25–30% of the patient's blood count. The T/NK cells were uniformly positive for CD56, CD16, CD2, CD7, CD8 (dim) and α/β. However, they were negative for surface CD3, CD4, CD5 and γ/δ. Interestingly, the cells were also CD45-negative. The diagnosis of T/NK lymphoproliferative disorder in a leukaemic phase was made and a bone marrow aspiration and biopsy procedure were planned. However, the patient deteriorated rapidly with increasing oxygen requirements. She eventually went into cardiac arrest and died.

Comment

This case illustrates the leukaemic phase of a disseminated T/NK lymphoproliferative disorder. The presentation and the rapid deterioration of the patient were consistent with aggressive disease. CD45-negative NK lymphomas are very rare, with only three cases described in the medical literature from East Asia of extranodal NK/T cell lymphoma-nasal type, with nasal cavity involvement.[3] The current case represents the first described case of CD45-negative T/NK lymphoproliferative disorder in the leukaemic phase.

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