'Fatal journey of a cutaneous ulcer'

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DESCRIPTION

A 14-year-old girl complained of multiple skin lesions for 1 month. Examination revealed multiple large well-defined cutaneous ulcers (6×6 cm) located over the trunk and a left-sided conjunctival nodule (figure 1A,B). Blood investigations revealed haemoglobin 78 g/L, white cell counts 17×10^9 /L, differential counts 85% neutrophils, 15% lymphocytes, platelets 700×10^9 /L and an elevated lactate dehydrogenase (LDH 450 U/L; normal range, 200–350 U/L). Biopsy from the ulcer edge showed anaplastic large cell lymphoma (ALCL, anaplastic lymphoma kinase (ALK) positive). PET-CT scan localised the disease to the skin. Treatment with six cycles of CHOP-21 (cyclophosphamide 750 mg/m², doxorubicin 50 mg/m², vincristine 1.4 mg/m² and prednisone 60 mg/m² times 5 days) chemotherapy led to complete resolution of the skin lesions as assessed by a repeat PET-CT scan. The patient presented to the emergency department with intense headache and generalised tonic-clonic seizures after being disease-free for 6 months. MRI brain with gadolinium contrast revealed a large parietal space-occupying lesion with surrounding oedema and midline shift (figure 1C,D). The patient succumbed to the raised intracranial pressure and tonsillar herniation despite steroids (dexamethasone 16 mg), anti-epileptics (sodium valproate 20 mg/kg) and decongestive measures (mannitol). Postmortem brain biopsy confirmed the diagnosis

Learning points

- ▶ Anaplastic large cell lymphoma (ALCL) must be kept in the differential diagnosis of 'cutaneous ulcer' in both children and adults.
- ▶ Regimens containing multiple chemotherapeutic drugs must be employed in the management of ALCL.
- ▶ CNS prophylaxis may be employed in cases with high-risk features predictive of CNS spread.
- ▶ CNS relapse in a case of ALCL is rare and imparts a grim outcome.

of ALCL, ALK positive. Primary cutaneous ALCL is rare in childhood. ALCL accounts for about 10%–15% of non-Hodgkin's lymphomas in children. Extranodal involvement is common at presentation (60%) with skin, bone and soft tissue being the most frequently involved sites. Central nervous system (CNS) involvement is unusual at diagnosis (<5%).¹ CNS relapse in peripheral T-cell lymphoma (PTCL) including ALCL is a rare event (2.4%). Features associated with a high risk of CNS relapse in diffuse large B-cell lymphoma (DLBCL) include elevated LDH (above institutional normal limit), more than one extranodal site of involvement and specific anatomical sites (testes, breast, paranasal sinus, epidural and bone marrow). Considering its rarity and lack of prospective studies, the British Committee for Standards in Haematology extends the recommendations of using CNS-directed prophylaxis (intrathecal methotrexate (12–15 mg) or systemic methotrexate (3–5 g/m²)) in high-risk DLBCL to PTCL as well.² Isolated CNS relapse in a case of primary cutaneous ALCL is extremely uncommon.

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Figure 1 (A) Large well-defined ulcer (6×6 cm) located over the back and a conjunctival nodule located over the nasal side of the left eye (B). MRI brain with gadolinium enhancement showing a large parietal space-occupying lesion with surrounding perilesional oedema and midline shift (C, D).

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