

# 'Fatal journey of a cutaneous ulcer'

Ankur Jain,<sup>1</sup> Naresh Gupta<sup>2</sup>

<sup>1</sup>Department of Internal Medicine, Post Graduate Institute of Medical Education and Research College of Nursing, Chandigarh, Punjab, India

<sup>2</sup>Department of internal medicine, Maulana Azad Medical College, New Delhi, Delhi, India

## Correspondence to

Dr Ankur Jain,  
drankur589@yahoo.in

Accepted 20 July 2017

## 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  $17 \times 10^9$ /L, differential counts 85% neutrophils, 15% lymphocytes, platelets  $700 \times 10^9$ /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/m<sup>2</sup>, doxorubicin 50 mg/m<sup>2</sup>, vincristine 1.4 mg/m<sup>2</sup> and prednisone 60 mg/m<sup>2</sup> 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%).<sup>1</sup> 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/m<sup>2</sup>)) in high-risk DLBCL to PTCL as well.<sup>2</sup> Isolated CNS relapse in a case of primary cutaneous ALCL is extremely uncommon.

**Contributors** AJ wrote the draft and was involved in the patient's management. NG reviewed the manuscript.

**Competing interests** None declared.

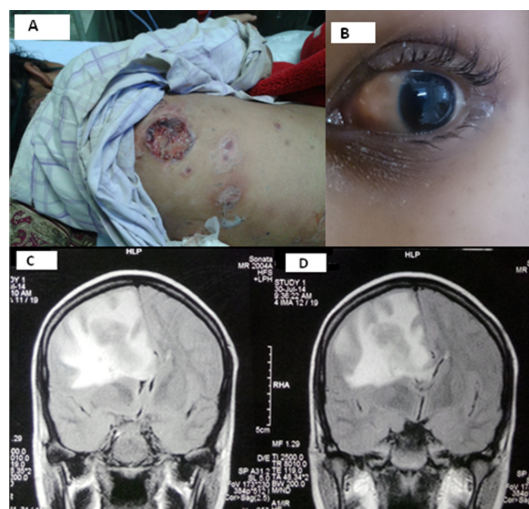
**Patient consent** Obtained.

**Provenance and peer review** Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2017. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

## REFERENCES

- 1 Rannan-Eliya YF, Pulford K, Johnson R, *et al*. Isolated cutaneous anaplastic large cell lymphoma progressing to severe systemic disease with myocardial involvement and central nervous system infiltration. *Pediatr Blood Cancer* 2008;50:879–81.
- 2 Gurion R, Mehta N, Migliacci JC, *et al*. Central nervous system involvement in T-cell lymphoma: a single center experience. *Acta Oncol* 2016;55:561–6.



**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).



CrossMark

**To cite:** Jain A, Gupta N. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-221250

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.  
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact [consortiasales@bmjgroup.com](mailto:consortiasales@bmjgroup.com)

Visit [casereports.bmj.com](http://casereports.bmj.com) for more articles like this and to become a Fellow