Case report

B cell acute lymphoblastic leukaemia presenting with isolated soft tissue tumours: the non-myeloid sarcoma

Mohammad Tousifullah, Aditya Kumar Gupta , Jagdish Prasad Meena, Rachna Seth

Department of Pediatrics,
All India Institute of Medical
Sciences, New Delhi, India

Correspondence to
Dr Aditya Kumar Gupta;
adivick@gmail.com

Accepted 21 September 2019

SUMMARY

Soft tissue leukaemic masses are a well-described clinical feature of myeloid haematological neoplasms. In paediatric acute lymphoblastic leukaemia (ALL), such soft tissue leukaemic sarcomas have not been reported as a presenting feature. Here we report a 3-year-old boy with ALL who presented to us with isolated soft tissue swellings for a duration of 9 months. The significance of these 'non-myeloid' sarcomas in paediatric ALL is uncertain.

BACKGROUND

Acute lymphoblastic leukaemia (ALL) is the most common cancer in children. ALL usually presents as progressive pallor, petechiae or bleeding and frequent infections. Lymphadenopathy, hepatomegaly and splenomegaly are seen in 60% of children. Involvement of the central nervous system is seen in 2%–5% of patients and testicular disease in 1% of patients. Acute myeloid leukaemia (AML) is more likely to present with extramedullary deposits, called myeloid sarcomas or granulocytic sarcomas. Extramedullary deposits in other organs or the skin can be found in lymphoblastic lymphomas, but these are less common in ALL.¹

CASE PRESENTATION

A 3-year-old boy presented to our hospital with multiple soft tissue swellings on the left forearm, forehead and scalp for the past 9 months. He had presented to our dermatology outpatient department, after which he had been referred to a paediatric oncologist. Six months ago the child had received 2 weeks of oral corticosteroids with the suspicion of systemic panniculitis, after which the swellings had transiently diminished in size. His blood counts had been normal at that time point. The swellings had been progressively increasing in size over the past 6 months. On examination the child had a soft 3×4 cm swelling on the dorsal aspect of the left forearm. There was another swelling of 3×3 cm on the forehead and on the left parietal region of the scalp ([figure 1](#)). There was no pallor, rash, lymphadenopathy or organomegaly.

INVESTIGATIONS

On investigation, the child had a haemoglobin of 110 g/L, total leucocyte count of 5.4×10^9 /L and

platelet count of 170×10^9 /L. The peripheral blood smear was normal.

X-rays of the skull and the left forearm were normal. The ultrasound of the swellings revealed multiple, hypoechoic, well-defined densities in the subcutaneous plane. With the suspicion of a soft tissue malignancy, a positron emission tomography scan was done and revealed highly fluorodeoxyglucose-avid subcutaneous soft tissue masses on the forehead, scalp and left forearm ([figure 2](#)).

A biopsy was done from the scalp mass, which on histopathological examination showed diffuse infiltration by small-to-intermediate-sized atypical lymphoid cells, extending into the dermis and having a blastoid morphology ([figure 2](#)). These atypical lymphoids were immunopositive for CD79a, CD20, CD10, BCL2 and CD34, and negative for terminal deoxynucleotidyl transferase (TdT), myeloperoxidase (MPO) and CD3.

A bone marrow aspirate was then done and showed numerous MPO-negative lymphoid blasts ([figure 2](#)). Flow cytometry of the bone marrow revealed 15% blasts that were positive for CD45, CD34, CD79a, CD19, human leucocyte antigen-DR (HLA-DR and CD38). The blasts were negative for MPO, CD3, CD15, CD33 and CD20. The molecular analysis showed the presence of the MLL gene rearrangement.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis considered was AML, as it is the leukaemia that is more likely to present with soft tissue leukaemic deposits. The flow cytometry in our case, however, confirmed that the diagnosis was ALL.

TREATMENT

A diagnosis of partially treated ALL was made and treatment was started. After the induction chemotherapy consisting of prednisolone, vincristine, L-asparaginase and daunorubicin, the masses resolved, the bone marrow was also in morphological remission, and the minimal residual disease was negative.

OUTCOME AND FOLLOW-UP

The child suffered from a very early relapse after the second phase of chemotherapy and the family declined further treatment.

© BMJ Publishing Group Limited 2019. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Tousifullah M, Gupta AK, Meena JP, et al. *BMJ Case Rep* 2019;**12**:e230982. doi:10.1136/bcr-2019-230982

Figure 1 Leukaemic sarcoma on the forehead in a child with acute lymphoblastic leukaemia (left) and an image of the child after the first phase of chemotherapy (right).

DISCUSSION

Soft tissue masses are more common in AML and they are called myeloid sarcomas or granulocytic sarcomas. They arise from the collection of the leukaemic cells at an extramedullary site, and the most common sites involved are the bone, orbits, skin and lymph nodes.² ALL presenting as a soft tissue mass is very rare. Orbital involvement in ALL, similar to the orbital involvement by a myeloid sarcoma, had been reported in the literature.³ At relapse ALL is known to involve unusual sites. Jain *et al*⁴ in 2014

Figure 2 Clockwise from top left: skin biopsy under 10× magnification; skin biopsy under 40× magnification showing infiltration up to the dermis by atypical lymphoid cells; bone marrow aspirate showing the presence of numerous lymphoid blasts; and PET scan showing the FDG-avid scalp lesion (the mild uptake in the arm on the right is the site of the FDG injection). FDG, fluorodeoxyglucose; PET, positron emission tomography.

Learning points

- ▶ Acute lymphoblastic leukaemia (ALL) can also present as soft tissue masses.
- ▶ In ALL these soft tissue masses can be the initial presentation.
- ▶ Whenever possible, steroids should be avoided in such situations until a biopsy has been obtained; otherwise, a sinister diagnosis could be masked.

described nine patients with unusual sites of relapse, of which the predominant site was ocular, and in one case the patient had presented with a soft tissue mass on the shoulder. The pathogenesis of these leukaemic sarcomas is unclear. For myeloid sarcomas in AML, it has been suggested that different chemokine/chemokine receptor interactions underlie the homing and retention of AML blasts in the skin. Stefanidakis and colleagues⁵ suggested that myeloid sarcomas represent an ability of the leukaemic blasts to invade the surrounding tissues, and showed that specific interaction between the matrix metalloproteinase-9 and the leucocyte surface beta (2) integrin is required for the migration of AML-derived cells. As the presence of these leukaemic sarcomas in ALL is not common, their significance is uncertain. Orbital involvement is known to confer a poor prognosis in ALL.³

This case highlights that leukaemic sarcomas could be non-myeloid in origin, and rarely ALL can present with such soft tissue masses.

Contributors MT and AKG were involved in planning, conduct and design, and acquisition of data. AKG, RS and JPM were involved in the analysis and interpretation of the findings. All authors approved of the final version of the manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

ORCID iD

Aditya Kumar Gupta <http://orcid.org/0000-0002-3730-8944>

REFERENCES

- 1 Clarke RT, Van den Bruel A, Bankhead C, *et al*. Clinical presentation of childhood leukaemia: a systematic review and meta-analysis. *Arch Dis Child* 2016;101:894–901.
- 2 Gupta AK, Radhakrishnan N, Sachdeva A. Disseminated myeloid sarcomas-a rare presentation of acute myeloid leukemia. *Pediatr Hematol Oncol* 2014;31:140–2.
- 3 Ramamoorthy J, Jain R, Trehan A, *et al*. Orbital mass in a child with acute lymphoblastic leukemia: a case report and review of the literature. *J Pediatr Hematol Oncol* 2016;38:646–8.
- 4 Jain R, Trehan A, Singh R, *et al*. Unusual sites of relapse in pre-B acute lymphoblastic leukemia. *J Pediatr Hematol Oncol* 2014;36:e506–8.
- 5 Stefanidakis M, Karjalainen K, Jaalouk DE, *et al*. Role of leukemia cell invadosome in extramedullary infiltration. *Blood* 2009;114:3008–17.

Copyright 2019 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <https://www.bmj.com/company/products-services/rights-and-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

Customer Service

If you have any further queries about your subscription, please contact our customer services team on +44 (0) 207111 1105 or via email at support@bmj.com.

Visit casereports.bmj.com for more articles like this and to become a Fellow