



OPEN ACCESS

Isolated CNS leukaemic relapse in acute myeloid leukaemia

Amr Essa ¹, Maryam Gbadamosi-Akindele,¹ Alan Lichtin²

¹Internal Medicine, Creighton University School of Medicine, Omaha, Nebraska, USA

²Hematology and Oncology, Cleveland Clinic Foundation, Cleveland, Ohio, USA

Correspondence to

Dr Maryam Gbadamosi-Akindele;
maryamgbadamosi@creighton.edu

Accepted 10 December 2019

DESCRIPTION

A 61-year-old woman with acute myeloid leukaemia (AML) without maturation (M1) with trisomy 13 who achieved complete remission after her initial diagnosis with cytarabine and idarubicin. Two years later, she relapsed and underwent successful haematopoietic stem cell transplantation afterwards, and she entered complete remission. Sixteen months later, she presented to our emergency department with a 2-week history of worsening fatigue, progressive expressive aphasia and then she lapsed coma. On the workup, there was no identifiable reversible metabolic or infectious cause of her presentation. Brain MRI revealed multifocal patchy abnormalities in the white matter ([figure 1A,B](#)). Her lumbar puncture (LP) revealed blasts on the cytocentrifuge preparations of cerebrospinal fluid (CSF) ([figure 1C](#)). Flow cytometry on the CSF confirmed the presence of a myeloblast population with 14% blasts with expression of CD13, CD33, CD34, CD45 and Human

Leukocyte Antigen – DR isotype (HLADR). And fluorescence in situ hybridization test was negative for Promyelocytic Leukemia/Retinoic Acid Receptor alpha (PML/RARa) translocation. There was no evidence of blast cells on the peripheral blood smear. And, bone marrow aspirate and biopsy revealed no evidence of blasts, indicating isolated central nervous system (CNS) relapse as the patient had no signs of other extramedullary involvement manifestation. Despite treatment with intrathecal methotrexate, she continued to deteriorate and died.

CNS complication in patients with leukaemia can be related to several mechanisms; direct involvement, vascular complications and infectious complications related to the disease itself or the treatment. CNS involvement in adults with AML is less common when compared with acute lymphoblastic leukaemia and is rare in general, and routine evaluation for CNS involvement is not recommended in absence of CNS symptoms.¹ Symptoms of CNS involvement depends generally on the involved anatomy and pathology. LP should be the initial workup for suspected CNS involvement and identification of leukaemic blasts on CSF should confirm the diagnosis.² CNS imaging studies should be performed based on the manifestations and to exclude other suspected possible pathology (eg, stroke, inflammatory or vascular lesions). Treatment include intrathecal chemotherapy and radiation therapy. Particular clinical features associated with a higher risk for CNS involvement in AML have been reported in the literature.^{1,3}

We thought the MRI finding, in this case, was related to myeloid sarcoma in the setting of isolated CNS leukaemic relapse. Myeloid sarcoma is a term for a tumour mass of myeloblast or immature myeloid cells occurring in an extramedullary site or the bone.⁴

This case demonstrates a good example of CNS imaging in CNS leukaemic relapse that can be an

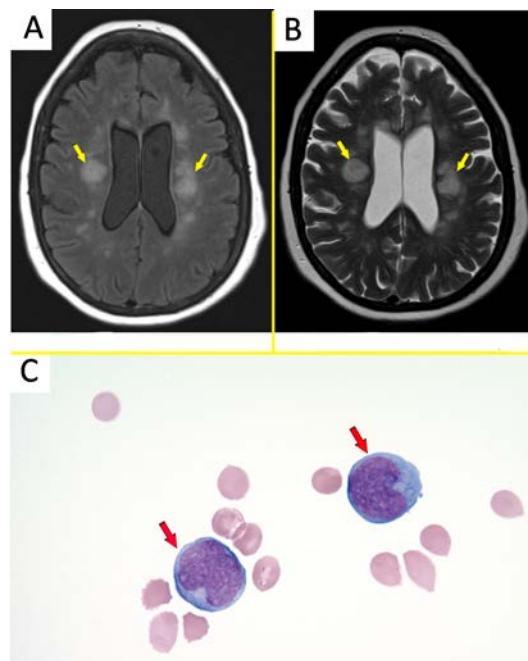


Figure 1 (A,B) Brain MRI T2 weighted sequence showing periventricular flair and hyperintensity (yellow arrows) with multifocal patchy abnormalities in the white matter characterised as restricted diffusion. (C) Cerebrospinal fluid showing large cells with increased nuclear size, increased nuclear/cytoplasmic ratio, fine chromatin and prominent nucleoli, consistent with blasts (red arrows).



© BMJ Publishing Group Limited 2019. Re-use permitted under CC BY-NC. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Essa A, Gbadamosi-Akindele M, Lichtin A. *BMJ Case Rep* 2019;**12**:e233499. doi:10.1136/bcr-2019-233499

Learning points

- ▶ Central nervous system (CNS) involvement in acute myeloid leukaemia is rare compared with acute lymphoblastic leukaemia.
- ▶ Lumbar puncture with cerebrospinal fluid analysis for blasts should be the initial workup once CNS involvement is suspected.
- ▶ CNS imaging should be directed towards a specific clinical presentation, and it can aid in the diagnosis.

instructive case for other clinicians. Also, it highlights the importance of imaging and pathology along with other clinical data in yielding the final diagnosis.

Contributors AE: obtained the images; prepared the manuscript; prepared the submission paperwork. AL: helped in obtaining the images; reviewed the manuscript. MG-A: reviewed the manuscript; did the submission process.

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 Not required.

Provenance and peer review Not commissioned; externally peer reviewed.

Open access This is an open access article distributed in accordance with the Creative Commons Attribution Non Commercial (CC BY-NC 4.0) license, which permits others to distribute, remix, adapt, build upon this work non-commercially,

and license their derivative works on different terms, provided the original work is properly cited and the use is non-commercial. See: <http://creativecommons.org/licenses/by-nc/4.0/>.

ORCID iD

Amr Essa <http://orcid.org/0000-0003-1310-643X>

REFERENCES

- 1 Alakel N, Stölzel F, Mohr B, *et al*. Symptomatic central nervous system involvement in adult patients with acute myeloid leukemia. *Cancer Manag Res* 2017;9:97–102.
- 2 Arber DA, Borowitz MJ, Cessna M, *et al*. Initial diagnostic workup of acute leukemia: guideline from the College of American pathologists and the American Society of hematology. *Arch Pathol Lab Med* 2017;141:1342–93.
- 3 Shihadeh F, Reed V, Faderl S, *et al*. Cytogenetic profile of patients with acute myeloid leukemia and central nervous system disease. *Cancer* 2012;118:112–7.
- 4 Brunning RD, Bennett J, Matutes E. Acute myeloid leukemia not otherwise categorised. In: Jaffe ES, Harris NL, Stein H, *et al*, eds. *Who classification of tumours; pathology and genetics of tumours of haematopoietic and lymphoid tissues*. Lyon: IARC Press, 2001: 104–5.

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