Meningeal leukemia in acute lymphoblastic leukemia revealed by an intracranial mass

Leucémie méningée au cours d’une leucémie lymphoblastique aiguë révélée par une masse intracrânienne

Case report

We report here on the case of a 13-year-old girl, with a history of two relapses of precursor B-cell acute lymphoblastic leukemia (B-ALL) post-stem-cell transplantation, who presented with paresthesia and uncontrolled movements in the right leg. Cerebrospinal fluid cytology was negative for leukemic cells. Magnetic resonance imaging (MRI) of the brain revealed a large parafalcine mass, with a dural tail, and leptomeningeal enhancement of the adjacent left frontal sulci. The mass showed isosignal intensity to grey matter on both T1- and T2-weighted images, diffusion restriction and homogeneous enhancement after contrast administration (Figs. 1–3). The mass was found underlying the dura during surgical biopsy. Histological examination revealed diffuse proliferation of lymphoblasts. Immunohistochemical staining demonstrated positivity of the tumor cells to terminal deoxynucleotidyl transferase (TdT) and cluster of differentiation (CD) 79a, while myeloperoxidase, CD3 and CD20 were negative (Fig. 4).

References


Bo Yin, Li Liu, Xing-rong Chen, Ke Li, Dao-ying Geng*
Department of Radiology, Huashan Hospital, Fudan University, No.12, Wulumuqi, Road Middle, Shanghai 200040, China
Correspondences

Figure 2  Coronal (A) and sagittal (B) contrast-enhanced T1-weighted MR images show the extra-axial mass with leptomeningeal enhancement of the adjacent frontal sulci (A, B) and a dural tail (A, arrows).

Figure 3  Axial diffusion-weighted image (A) and the apparent diffusion coefficient map (B) show restricted diffusion in the mass.

Figure 4  Hematoxylin–eosin staining of a frozen section from a brain biopsy reveals a dense blastoid infiltrate with high mitotic activity (arrow). The cells express CD79a (B) and nuclear terminal deoxynucleotidyl transferase (C), thereby confirming their B-cell origin.
These findings are consistent with precursor B-ALL. Bone-marrow aspiration revealed concomitant leukemic bone-marrow relapse. On imaging, the tumor disappeared gradually with irradiation, and administration of systemic and intrathecal chemotherapy. Unfortunately, the patient died a few months later from gram-negative sepsis.

ALL is the most common type of cancer in childhood. Metastatic involvement of the meninges in children occurs primarily in ALL and in primary brain tumors. In patients with leukemia, it is often called 'leukemic meningitis' or 'meningeal leukemia'. In ALL, it is diagnosed either by detection of lymphoblasts in cerebrospinal fluid or by neuroimaging. Intracranial leukemic masses rarely occur in myeloid and lymphoid leukemias. However, in ALL, intracranial leukemic masses have been described in the literature using descriptive terms such as "tumor masses" [1] and "tumefactive presentations" [2], and also as "myeloid" (or "granulocytic") "sarcomas" or "chloromas" [3,4]. The latter terms are inaccurate and should be reserved only for leukemic masses consisting of myeloid cells expressing the enzyme myeloperoxidase.

So far, in the English literature, there have been four biopsy-proven cases of an intracranial leukemic mass either as the initial presentation [2] or as a relapse [1,3,4] of B-ALL. In the Japanese literature, however, a further four patients with ALL and an intracranial leukemic mass were reported in a retrospective study of 65 children with leukemia [5]. All tumors appeared to be homogeneously enhancing meningeal-based mass [1–5] that was either pachymeningeal (dural or falcine) or leptomeningeal. Peritumoral edema, presence of a dural tail and subperioveal tumor extension were also seen in a patient with a dural-based mass [2]. Yet another patient presented with a leptomeningeal mass with peritumoral edema [1].

Our present patient presented with a leptomeningeal mass with a dural tail that was not caused by dural invasion, as the tumor was found underlying the dura. One neuro-oncological report [4] mentions an intra-axial (intracerebral) mass; however, MRI findings were consistent with an extra-axial mass bulging into the temporal lobe and causing peritumoral edema. Surgery and pathology demonstrated attachment to the dura with no invasion of the brain parenchyma, indicating that this was another case of a dural-based mass.

In addition to these proven cases, one other patient with ALL and multiple intracerebral (intra-axial) nodules has been described in the literature [6]. The nodules were not biopsied, but were assumed to be metastatic brain tumors of ALL. However, as the imaging features of these nodules are nonspecific, there is room for doubt over the leukemic origin of the nodules in this patient.

In conclusion, all of the reported and biopsy-proven intracranial leukemic masses in ALL were extra-axial tumors. They may be pachymeningeal or leptomeningeal, and they can all be designated as cases of meningeal leukemia.

Disclosure of interest

The authors have not supplied their declaration of conflict of interest.

References


L.J.L. De Cocker a,∗
T. Tousseyn b
F. Van Calenbergh c
A. Uyttebroeck d
P. Demaereel e

a Department of Radiology, University Hospitals Leuven, Herestraat 49, 3000 Leuven, Belgium
b Department of Pathology, University Hospitals Leuven, Leuven, Belgium
c Department of Neurosurgery, University Hospitals Leuven, Leuven, Belgium
d Division of Pediatric Hematology and Oncology, Department of Pediatrics, University Hospitals Leuven, Leuven, Belgium

∗ Corresponding author. Tel.: +32032 16 332211.
E-mail addresses: laurens.decocker@uzleuven.be, laurens.de_cocker@hotmail.com (L.J.L. De Cocker)

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Spontaneous Cervical Epidural Hematoma Mimicking Stroke

Hématome épidural cervical spontané évoquant un accident vasculaire cérébral

A 62-year-old man presented to the emergency department after awakening with neck pain. En route to the hospital, he experienced progressive weakness and a sensation of heaviness in his right arm and leg. The patient reported a past medical history of hypertension and hyperlipidemia and a history of dancing at a party the previous night. The patient experienced “neck spasms” treated with aspirin the morning of presentation to the emergency department. Physical examination revealed 3/5 strength on