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IMAGE OF THE MONTH

Aggressive Langerhans cell histiocytosis transformation of T cell acute lymphoblastic leukemia detected on ^{18}F-FDG PET/CT

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A 12-year-old boy with clinical history of T cell acute lymphoblastic leukemia (T-ALL) 10 months ago presented with multiple palpable and enlarged left cervical lymph nodes, splenomegaly, and fatigue for 1 month. Bone marrow biopsy result of T-ALL shows obviously active proliferation of karyocyte, inhibited granulocytes, and hypoproliferation of red blood cell. The proportion of lymphocytes significantly increased with predominant small primitive cells and immature lymphocytes, which accounts for 79%. Immunohistochemical staining shows the tumor cells strongly expressed CD7; partially expressed CD34, TdT, and cCD3; slightly expressed mCD3 and CD2; and negatively expressed CD117, CD13, CD33, CD10, MPO, CD1α, CD99, and cCD79α. He accepted six cycles of routine chemotherapy and two cycles reinforcement treatment, and complete remission with incomplete blood count recovery (CRi) was achieved 3 months ago. Blood routine test shows moderate thrombocytopenia, hemoglobin was 105 g/L, WBC was 3.57×10^9/L, and platelets was 86×10^9/L.

The value of ^{18}F-FDG PET/CT in predicting prognosis and aggressive transformation in patients with acute leukemia had been illustrated [1]. On ^{18}F-FDG PET/CT imaging, multiple lesions in left multiple enlarged cervical lymph nodes (SUVmax = 6.9), spleen (SUVmax = 5.3), and left liver lobe (SUVmax = 3.6) with hypermetabolism on PET image (a) and PET/CT images (b–j) were found; leukemia relapse was highly suspected accordingly.

After biopsy of left enlarged cervical lymph node and lesion of left liver lobe, however, the histopathological results demonstrated that the lesions were Langerhans cell histiocytosis (LCH). HE-stained photomicrographs show a population of Langerhans' cell (HE × 100) (k), oval or lobulated nuclei, appeared with nuclear groove and indentation in partial cells (HE × 400) (l). There is a strong surface staining for CD1α (CD1α staining × 400) (m). The cells show strong expression of S-100 in a nuclear and cytoplasmic pattern (S-100 staining × 200) (n).

The center of spleen lesion also showed low density and hypometabolism, which suggests the lesion showed aggressive behavior. The subsequent aggressive LCH transformation was on average 18 months after maintenance chemotherapy for T-ALL, which indicated the resistance to chemotherapeutic agents; its prognosis was very poor [2–4].

It demonstrates that LCH should be considered in the differential diagnosis of T-ALL relapse detected on ^{18}F-FDG PET/CT imaging.

Jianjie Wang and Tianbin Song contributed equally to this work.

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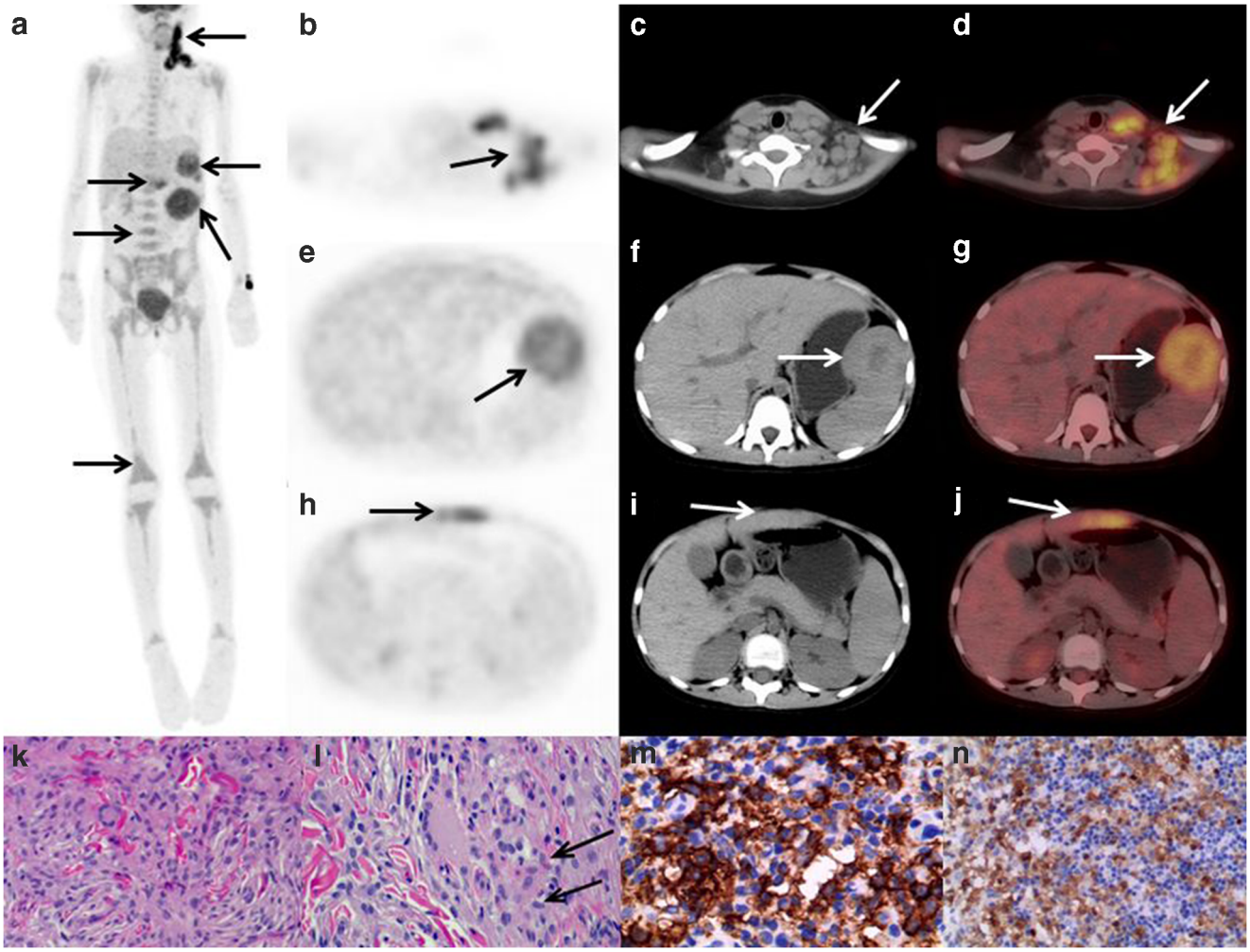

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with animals performed by any of the authors. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the principles of the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from the patient and his parents for the anonymous use of his clinical, imaging, and histologic data for publication.

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