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CORRESPONDENCE

Multiple Granulocytic Sarcomas: A Rare Presentation of Acute Myeloid Leukemia

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Dear editor,

A 16-year-old boy, presented with gradually progressive, painless swelling over the back for 1-month duration followed by similar swellings over head and chest. He also noticed bulging of the right eyeball with double vision in last 1 week.

Clinical examination revealed a thin built and cachexic young boy. He had multiple swellings over scalp (6 × 4 cm), chest wall (8 × 5 cm) and sacrum (5 × 2 cm, 3 × 2 cm) that were firm to hard, non-tender and adherent to the underlying structure (Fig. 1a, c, d). He also had proptosis of the right eye (Fig. 1b). Ocular movements and pupillary reflex were preserved. Rest of the clinical examination was unremarkable.

His complete blood count showed, Hemoglobin 138 g/L, WBC 3.6 × 10^9/L, and Platelets 260 × 10^9/L with a normal peripheral blood smear. A Whole-body PET CT showed intense FDG avid lesions corresponding to the clinically apparent sites along with a retro-orbital mass (Fig. 2). The histopathology report of the biopsied tissue from the sacral swelling was suggestive of non-Hodgkin lymphoma. Bone marrow aspiration-biopsy showed 26% MPO negative blast, which was positive for CD13, CD33, CD34, CD38, CD117 and HLA DR and Cytogenetic studies revealed t (8:21) (AML/ETO or RUNX1/RUNX1T1). Diagnosis of Acute myeloid leukemia with recurrent cytogenetic abnormality t (8:21) (AML/ETO or RUNX1/RUNX1T1) was made. Because of the deplorable general condition, patient and the family agreed for hypomethylating agents as a bridge to the standard induction therapy. The patient succumbed to the illness after the first cycle of Decitabine due to sepsis and multi-organ dysfunction.

Granulocytic sarcoma (GS), also known as myeloid sarcoma or chloromas have been described since 1811 [1]. Along with leukemia cutis, it is a well-defined extramedullary manifestation of acute myeloid leukemia. It constitutes a collection of immature cells of granulocytic origin and it is seen in 2.5–9.1% of AML cases [2]. Granulocytic sarcoma can precede, concur or follow the AML. It can present as a solitary lesion or multifocal lesion [3]. It can be virtually located in any part of the body and when present confer a poor clinical outcome [4]. Common differential diagnosis includes non-Hodgkin's lymphoma, dendritic cell neoplasm, Ewing sarcoma, and melanoma [5]. Diagnosis depends on morphology, immunohistochemistry and flow cytometry. PET-CT is useful in detecting anatomically hidden GS, planning radiotherapy and assessing the response to therapy [4, 6]. The outcome of chemotherapy alone in patients presenting with granulocytic sarcoma is often disappointing. The standard treatment constitutes induction chemotherapy (7 + 3) followed by hematopoietic stem cell transplantation [2]. A favorable risk is attributed to patients with t (8:21), and a good prognosis is expected from chemotherapy alone, but the outcome is significantly poor in the presence of GS even in the presence of t (8;14) and thus necessitates HSCT in complete remission after induction chemotherapy [4, 7].

The index case represents clinically apparent multiple granulocytic sarcoma as presenting feature of AML, which is extremely rare and may confuse the clinician with other solid malignancy. In the absence of clinical suspicion, even

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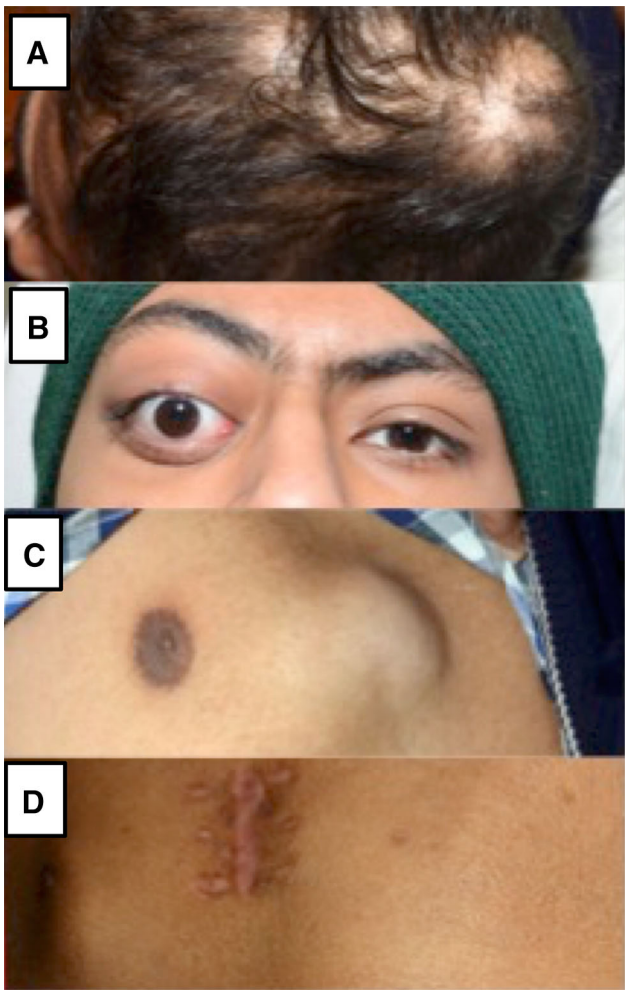

Fig. 1 Clinical photograph showing swellings over scalp (**a**), chest wall (**c**) and sacrum (**d**) and proptosis of the right eye (**b**)

the morphological appearances on histopathology may mimic a non-Hodgkin lymphoma and unnecessary delay in diagnosis of one of the most aggressive malignancies that require early diagnosis and prompt therapy.

Compliance with Ethical Standards

Conflict of interest There is no conflict of interest between the authors.

Ethical Approval 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 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Human and Animals Rights No animals were involved in the study.

Informed Consent Informed signed written consent was taken from the patient involved.

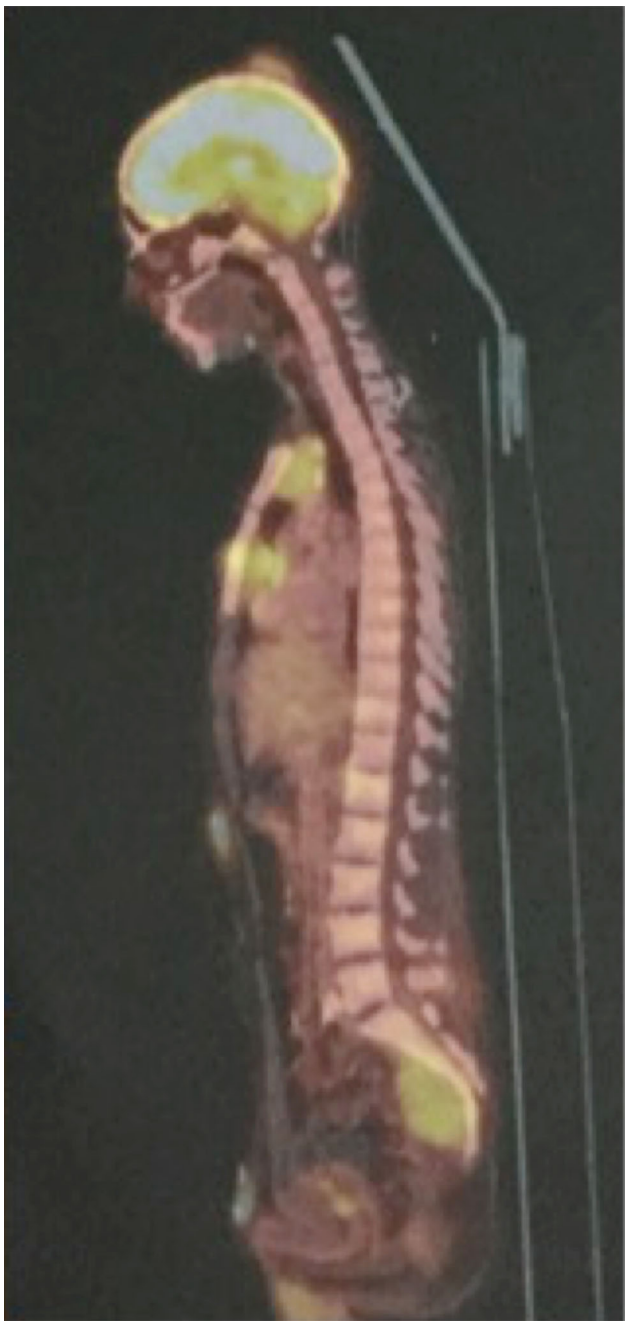

Fig. 2 PET-CT showing intense FDG avid lesions corresponding to the clinically apparent sites of scalp, eye, chest wall, and sacrum

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