



An Unusual Cause of Thigh Swelling: Extramedullary Myeloid Tumor

Uylukta Şişliğin Nadir Rastlanan Bir Sebebi: Ekstramedullar Myeloid Tümör

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To the Editor,

Extramedullary myeloid tumor (EMMT) is a rare neoplasm of immature myeloid cells that arises at an extramedullary site [1]. The most common sites of EMMT are the bone, lymph nodes, skin, and soft tissue [2]. EMMT rarely infiltrates the lower extremities.

A 47-year-old male patient was admitted to the orthopedics clinic because of swelling and pain in the right thigh for 1 month. His past medical history was unremarkable. Physical examination indicated a 10 cm-long solid soft tissue lesion in the anterior and lateral parts of the right thigh. Complete blood count results were as follows: white blood cell count of $15.1 \times 10^9/L$, hemoglobin level of 11.9 g/dL, and platelet count of $94 \times 10^9/L$. Magnetic resonance imaging (MRI) of the right thigh demonstrated a heterogeneous mass extending towards the distal part in the anterolateral section of the right femoral neck, completely involving the vastus lateralis and intermedius muscles (Figure 1). The patient underwent Tru-Cut biopsy of the right thigh. Pathology of the Tru-Cut biopsy showed large blastic cells infiltrating the soft tissue with hyperchromatic nuclei stained positively with CD117, CD34, and myeloperoxidase (Figure 2). After the Tru-Cut biopsy, blood count results were: white blood cells, $21.5 \times 10^9/L$, hemoglobin, 11.5 g/dL, and platelets, $74 \times 10^9/L$. Bone marrow aspiration showed 60% blasts, which were intensely positive for myeloperoxidase. Flow cytometry

performed on the bone marrow revealed a blast population that expressed CD34, CD117, CD33, CD15, CD13, CD19, and HLA-DR. As a result of cytogenetic testing, a new complex karyotype related to chromosomes 8, 10, and 21 and trisomy 8 were detected. The patient was started on an acute myeloid leukemia (AML) induction chemotherapy regimen consisting of idarubicin (12 mg/m², daily for 3 days) and cytosine arabinoside (ara-C; 200 mg/m² continuous infusion for 7 days). After 4 weeks, the control bone marrow aspiration was completely normal. The lesion had also disappeared completely in the control MRI of the thigh. The patient was administered a high-maintenance dose of ara-C at 3 g/m² for 6 days for consolidation. Treatment is ongoing.

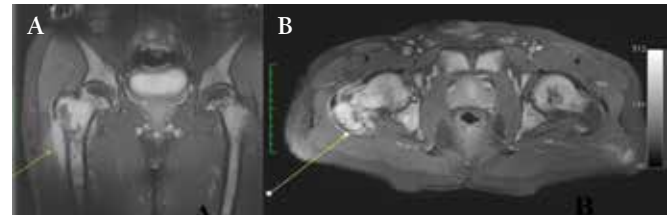


Figure 1. A) Metaphysodiaphyseal paracortical heterogeneous enhancement of the proximal femur in the post-contrast T1-weighted coronal image. B) Paracortical minimal signal increase is seen in the proximal femoral metaphysodiaphyseal part in coronal T2-weighted fat-suppressed image.

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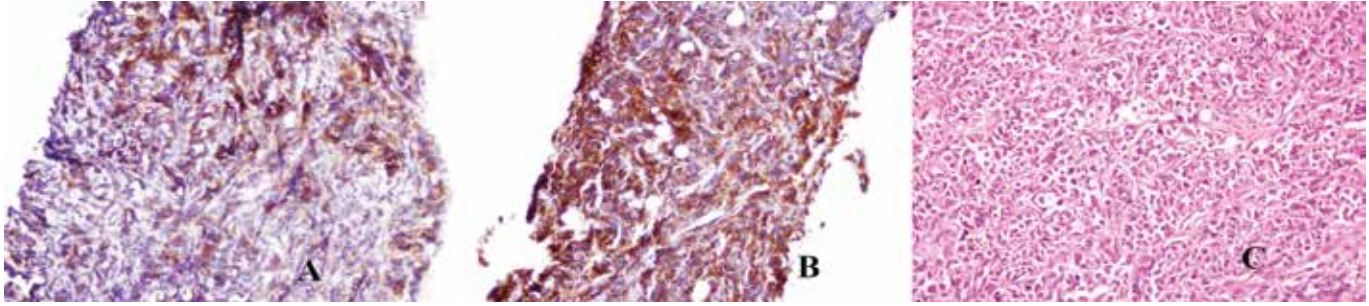


Figure 2. A) Selected cytoplasm, large hyperchromatic nuclei, and tumor infiltration of cells showing marked pleomorphism (H&E, 40x). B) Diffuse strong membranous/cytoplasmic staining for CD34 in tumor cells and associated vascular structures (H&E, 40x). C) Membranous/cytoplasmic staining strong in some places and moderate in some places in tumor cells (CD117, 40x).

EMMTs are composed of myeloid blasts. They can easily be confused with lymphomas or soft tissue sarcomas [3,4]. EMMTs may accompany AML in 35% of patients at presentation, 38% of patients following diagnosis of AML, and 27% of patients without diagnosis of AML [5]. Cytogenetic abnormalities like translocation (8;22) or inversion 16 and 11q23 were reported in EMMT [6]. An optimal treatment approach does not exist due to the lack of randomized studies. Intensive chemotherapy regimens containing idarubicin plus ara-C are usually administered in the treatment of EMMT. According to the risk factors (age; molecular and cytogenetic study results), allogeneic stem cell transplantation may also be considered. In the case of residual infiltration as shown by imaging, radiotherapy should be considered [7].

Consequently, EMMT might be taken into consideration in the differential diagnosis of venous thromboembolism and soft tissue malignancies in the case of swollen thighs.

Conflict of Interest Statement

The authors of this paper have no conflicts of interest, including specific financial interests, relationships, and/or affiliations relevant to the subject matter or materials included.

Key Words: Acute myeloblastic leukemia, Granulocytes, acute leukemia, Hemophagocytic lymphohistiocytosis

Anahtar Sözcükler: Akut miyeloblastik lösemi, Granülositler, Akut lösemi, Hemofagositik lenfohistiositöz

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