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A Rare Cause of Paraplegia: Myeloid Sarcoma

Nadir Bir Parapleji Nedeni: Miyeloid Sarkom

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To the Editor,

Myeloid sarcoma (MS), also known as granulocytic sarcoma or chloroma, is a rare extramedullary tumor consisting of myeloblasts or immature myeloid cells that disrupt the normal architecture of the involved tissue and typically occurs concurrently with acute myeloid leukemia (AML) [1,2]. It can also occur in association with accelerated-phase chronic myeloid leukemia, myelodysplastic syndrome, as an extramedullary relapse of AML, including in the post bone marrow transplant setting, and, occasionally may present as the first manifestation, even before bone marrow involvement [3,4]. Bone, periosteum, skin, orbit, lymph nodes, gastrointestinal tract and central nervous system are the most commonly involved sites in patients presenting with MS; however, skin and orbital localizations are the most often reported sites in children [4]. Here we present a four year old male patient who was referred to the pediatric hematology oncology clinic due to a thoracolumbar mass, and subsequently diagnosed with MS.

A four year old boy was referred to the pediatric hematology oncology clinic with the complaint of hemiparesis and a subsequent thoracolumbar mass detected by magnetic resonance imaging (MRI) (Figure 1A). On physical examination, bilateral lower extremity paralysis was noted and deep tendon reflexes were absent. Complete blood count, blood biochemical analysis were normal, and no blasts were detected on peripheral blood film. Bone marrow aspiration showed 30% blasts compatible with AML. The pathology of the mass revealed MS. After administration of radiotherapy, given at a dose of 18 Gy in 10 daily fractions in 2 weeks, and dexamethasone therapy, the patient achieved neurological improvement. He was treated with AML-BFM 2012 protocol and achieved both remission and mass reduction following AML induction chemotherapy. The patient is still in remission without any residual tumor on following MRI (Figure 1B).

Myeloid sarcoma may ocur at any site of the body, and therefore clinic manifestations of MS exhibit diversity depending on its specific location and size, which leads to significant diagnostic challenges, in particular in patients without initial bone marrow involvement. Incorrect diagnosis of malignant lymphoproliferative disorders, Ewing's sarcoma, thymoma, melanoma, round blue cell tumours, or poorly differentiated carcinoma has been reported at a rate of 25-47% in patients subsequently diagnosed with MS. In this regard, any atypical cellular infiltrate, should raise the suspicion for MS to make correct diagnosis timely, and proper management [2,4,5]. Diagnostic tools for the correct diagnosis of MS are also important in this context, and should include MRI and/or computed tomography scan for evaluation of size and location of the tumour and distinguishing tumour from other lesions, morphological and flow cytometric analysis of bone marrow and peripheral blood, or biopsy of the tumour and immunohistochemical staining in patients without bone marrow involvement [4]. Treatment of MS includes AML-based protocols and, as in our case, surgery and/or radiotherapy may be indicated for symptomatic lesions or tumors causing local organ dysfunction [5]. Considering the most common presentation sites in children with MS, which are skin and orbital localizations, the current patient is presented to highlight a rarely encountered presenting feature of MS.

Keywords: Myeloid sarcoma, children, paraplegia

Anahtar Sözcükler: Miyeloid sarkom, çocuk, parapleji

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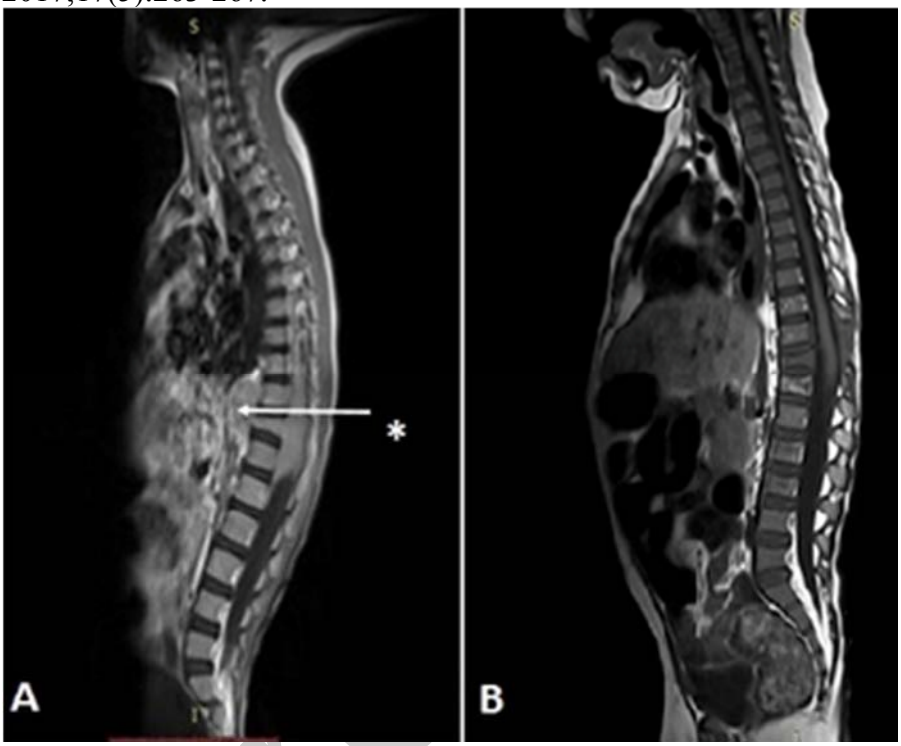

Figure 1A: Sagittal T1-weighted MRI image showing an epidural, hypointense, craniocaudal 4.5 cm diameter mass compressing spinal cord at the level of D 10-12; Figure 1B: The image of the mass 1 month before completion of acute myeloid leukemia maintenance therapy