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Case report

Multifocal myeloid sarcoma in the central nervous system without leukemia

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1. Introduction

Myeloid sarcoma is a localized tumor composed of primitive myeloid cells at an extramedullary site, which has often been described in association with leukemia or myeloid proliferative disorders. It is previously known as granulocytic sarcoma, chloroma, or extra-medullary myeloid tumor, and the 2008 World Health Organization classification adopted the term "myeloid sarcomas" as a subgroup of "acute myeloid leukemias, not otherwise categorized" [1]. Myeloid sarcoma most commonly occurs in the soft tissues of the head and neck, bone, and skin, while rarely in the central nervous system (CNS), especially spinal cord [2]. Herein, we describe a patient with multifocal myeloid sarcoma in the CNS. In particular, no leukemic manifestations were detected in bone marrow and peripheral blood examinations.

2. Case report

In April 2012, a previously healthy 27-year-old man presented to us with a 6-month history of intermittent headache and a 3-month history of bilateral visual field defects. He was a radiology technician who had operated X-ray equipment for the previous 5 years. Physical examination revealed a homonymous right lower quadrant visual field defect. Brain MRI showed a parasagittal mass involving the occipital lobes bilaterally. The lesion was isointense on T1-weighted images with homogeneous contrast enhancement, and mildly hyperintense on T2-weighted images (Fig. 1). Baseline laboratory data were normal: leukocyte count 8910/μL with a normal differential, hemoglobin 132 g/L, and platelet count 204,000/μL. A presumptive diagnosis of parasagittal meningioma was made, and subsequently a corresponding en-bloc tumorectomy was designed, not including a frozen-section examination. Microscopically, gross total resection was performed of the intradural grayish mass with a well-defined cleavage plane. Immunohistological examination showed myeloid sarcoma (Fig. 2A) with positive staining for myeloperoxidase (MPO, Fig. 2B), CD34, CD45, CD68, CD99, and lysozyme. Postoperatively, his headaches resolved but the visual field defects remained. The postoperative MRI confirmed gross total resection. Subsequently, further cytogenetic examinations, bone marrow aspirates and adjuvant chemotherapy was recommended, nevertheless in vain due to the patient refusal.

Five months after discharge, without any related intervention in the interim, the patient was readmitted with a 2-month history of pain and numbness in his right thigh, sphincter dysfunction, and occasional pain in the right flank. Neurological examination revealed a loss of sensation below the L2 dermatome on the right, and grade 4/5 right lower extremity weakness. Spinal MRI showed a giant mass in the spinal canal at L2–L3 that extended through the right L2–L3 intervertebral foramen, and a smaller mass at

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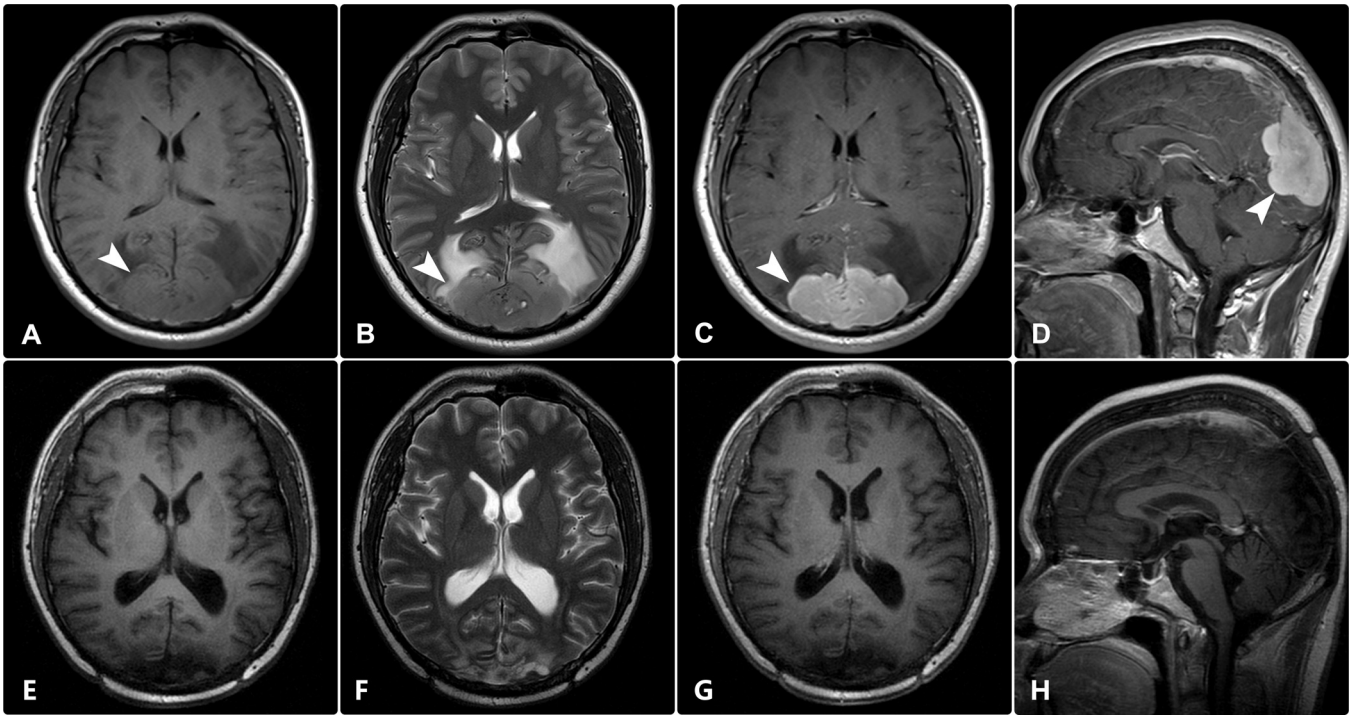

Fig. 1. The cranial MR images show a parasagittal mass involving bilateral occipital lobes (arrowheads). The lesion is isointense on axial T1-weighted images (A), and mildly hyperintense on axial T2-weighted images (B). After Gd-DTPA administration, the axial (C) and sagittal (D) images show homogeneous contrast enhancement. The corresponding MR images in the last follow-up (E–H) show tumor-free patterns.

S2–S3 (Fig. 3). The masses were suspected as myeloid sarcoma because of multiple occurrence or CSF dissemination. Cerebrospinal fluid smear examination was performed, identifying several typical immature cells with distinctive Auer rods (Fig. 2C). Resection of the intra-spinal portion of the larger mass for local decompression and biopsy combined with six-cycle chemotherapy was scheduled. Intraoperatively, the larger mass was found epidural, and diffusely infiltrating into the right psoas muscle. Pathological examination

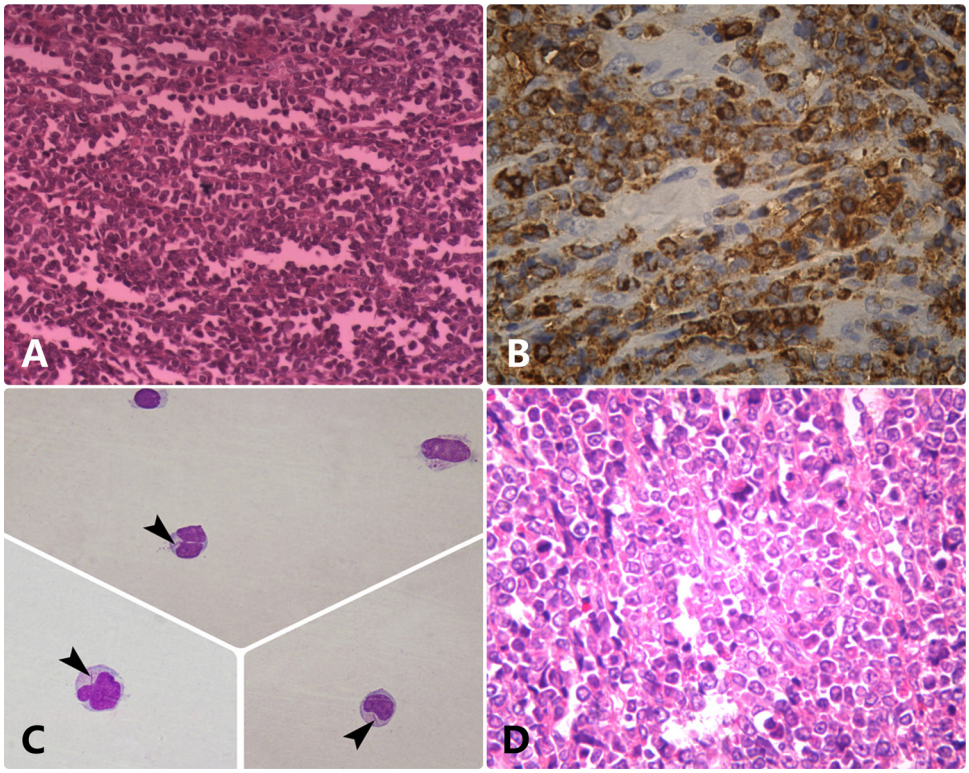

Fig. 2. H&E stain of the center of the resected brain tissue reveals myeloid sarcoma (A, ×200). Immunohistochemical stain shows positivity for myeloperoxidase (B, ×400). High-power views of cerebrospinal fluid smears show sporadic immature cells with Auer rods (C, ×1000). H&E stain of spinal tumor also reveals myeloid sarcoma (D, ×400).