

Successful treatment of large intracranial granulocytic sarcomas

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ABSTRACT

Granulocytic sarcoma is highly sensitive to radiotherapy and chemotherapy, and surgery is usually performed to obtain a biopsy specimen for a definitive diagnosis. The effectiveness of surgical resection remains controversial. We herein describe a case of symptomatic granulocytic sarcoma that replaced intracranial hypertension. A 58-year-old woman suffered from a headache, motor aphasia, and right hemiplegia during treatment for acute myeloblastic leukemia. Computed tomography and magnetic resonance imaging of the brain revealed multiple subdural masses. The largest symptomatic mass was resected for decompression, and granulocytic sarcoma was diagnosed. The patient became symptom-free after the surgery, and whole brain radiotherapy was subsequently performed. Although chemoradiotherapy is generally used for treatment of granulocytic sarcoma, surgical resection for decompression is advisable in cases in which clinical symptoms are present and performance status is altered.

Introduction

Granulocytic sarcoma (GS) generally occurs as a complication of acute myeloblastic leukemia (AML). Among 633 children with myelogenous leukemia, GS was observed in 4.7% of those with AML and 1% of those with chronic myeloblastic leukemia. [1] GS involves subcutaneous tissue, the orbit, paranasal sinuses, lymph nodes, bone, and periosteum [1,2]. Involvement of the central nervous system (CNS) by GS is rare. If GS develops in the CNS, it tends to form extra medullary tumors, which invade the dura mater and subarachnoid space.

We encountered a patient with multiple symptomatic GS lesions under the dura mater who went into complete remission after surgery. We describe this case and discuss therapeutic strategies for symptomatic intracranial GS.

Case Report

The patient was a 58-year-old woman who underwent inpatient treatment for AML in the hematology department of our hospital. During her hospital stay, severe headache, motor aphasia, and right hemiplegia developed. Karnofsky performance status (KPS) decreased to 30%. Computed tomography revealed a large mass in the left convexity. Magnetic Resonance Imaging

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(MRI) revealed multiple, homogeneous, contrast-enhanced masses in both convexities, the cerebral falx, and the left cerebellopontine angle (Fig. 1A-D). The mass in the left convexity was 40 mm in the greatest thickness, and it severely compressed the brain. These findings strongly suggested GS. The mass in the left convexity was surgically resected to alleviate intracranial hypertension. Intraoperatively, the tumor was resilient and hard but did not adhere to the brain surface (Fig. 2A). The tumor was subtotally removed.

Upon pathologic examination of the surgical specimen, medullary-type proliferation of medium-sized and large atypical cells was noted, and the evidence of active cell division was observed (Fig. 2B). The diagnosis of GS was established. After the surgery, the patient's symptoms promptly remitted, and KPS was restored to 100%. The residual lesions were treated by whole brain radiotherapy (total 30 Gy), followed by high-dose chemotherapy with melphalan and methotrexate. Thereafter, peripheral blood stem cell transplantation was performed, and complete remission was achieved. The patient is now in complete hematologic remission and has no neurologic deficits. Follow-up MRI that was performed 18 months after surgery showed no evidence of the intracranial lesion (Fig. 3A, B).

Discussion

Involvement of the CNS by GS has been reported to occur in about 0.7% of patients with AML [3]. Intracranial GS arises in the dura mater and subarachnoid space and is sometimes found within the brain parenchyma [2,4,5]. Multiple GS lesions are sometimes noted in either intracranial or extracranial tissues [6,7]. In the present case, multiple lesions were detected in the dura mater. In cases of multiple lesions, contrast-enhanced MRI is useful to distinguish GS from subdural abscess or subdural hematoma [1,8].

The major treatment option for GS is chemotherapy combined with radiotherapy. Surgery is generally limited to obtaining a biopsy specimen for a definitive diagnosis [9,10]. However, surgical removal of the large mass in the brain was critical in our case. Pui et al [1]. Reported the treatment of 30 children with AML and GS. In 13 of the 30 patients, the tumors resolved completely within 3 months after treatment with chemotherapy alone. Six patients required additional radiotherapy. Among the remaining 11 patients, there were 2 recurrent tumors despite surgical resection and chemotherapy [1]. Responses of the 30 patients, the different to treatments varied.

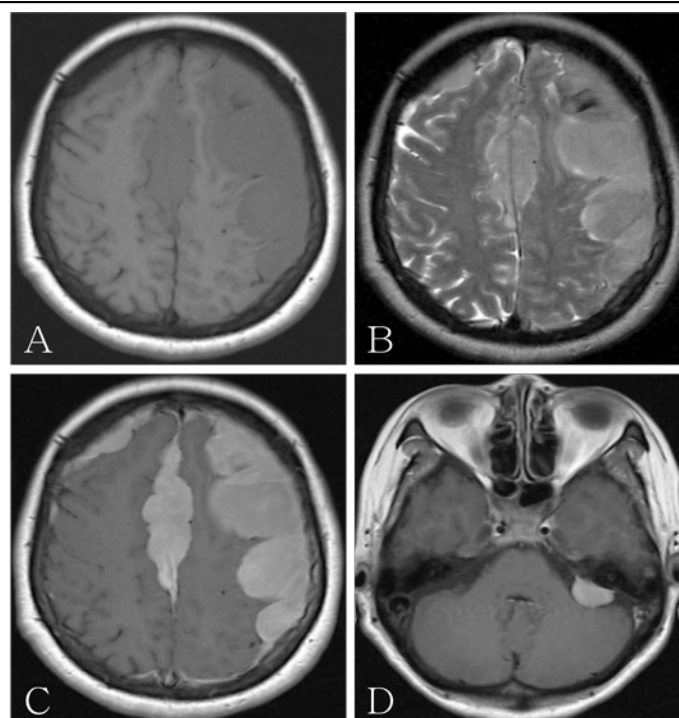


Figure 1: Preoperative magnetic resonance images. Multiple masses appear hypointense on the T1-weighted image (A), hyperintense on the T2-weighted image (B), and homogeneously enhanced on the contrast-enhanced T1-weighted images (C, D).

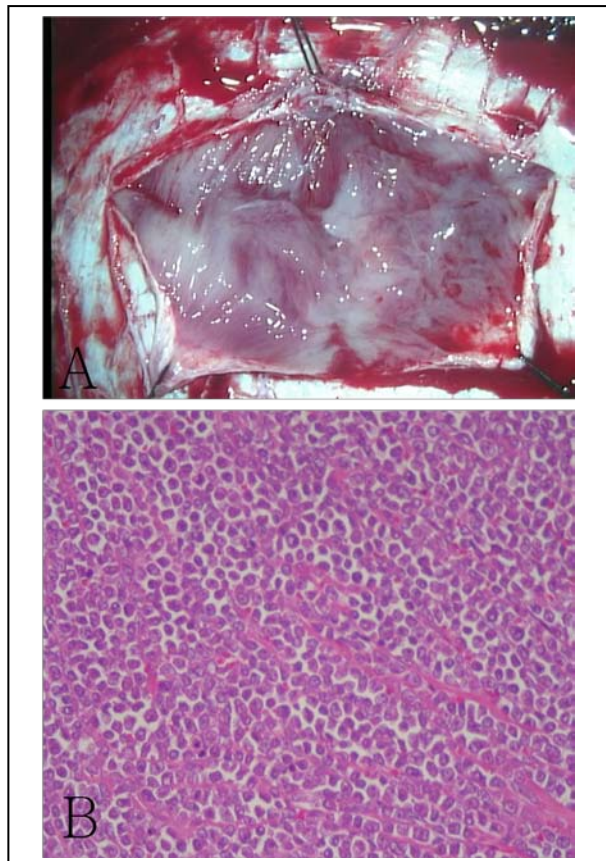


Figure 2A: Intraoperative photograph. The tumor had spread into the subdural space, and it did not adhere to the brain surface.
Figure 2B: Histologic (hematoxylin-eosin, $\times 400$) appearance of the surgical specimen. The tumor contained myeloid cells at different stages of maturation. Note the variation in size and lobulated nuclei.

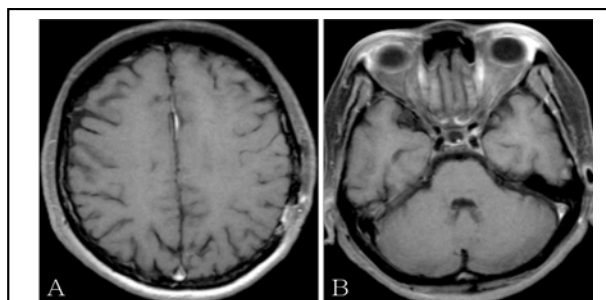


Figure 3: Post-treatment magnetic resonance image. No tumors are seen on contrast-enhanced T1-weighted images after 18 months (A, B).

It remains undetermined what size intracranial GS can be treated by radiotherapy and chemotherapy. Furthermore, reduced KPS may delay the introduction of systemic chemotherapy, which might be life-threatening in patients with systemic disease such as AML. Velasco et al [11]. reported that surgical resection should be considered when there is a large intracranial GS with significant peritumoral edema. Thus, we aggressively

performed surgical resection to improve the performance status of our patient.

Although the prognosis for AML with intracranial GS is generally poor, complete remission was achieved in our case. We consider aggressive surgical resection as an ideal option to improve performance status of AML patients with symptomatic intracranial symptomatic GS. This will allow subsequent treatment to be introduced without hindrance.

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