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Case Report

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The Role of Radiotherapy in Myeloid Sarcoma: A Case Report and Review of the Literature

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ABSTRACT

Myeloid sarcoma (MS) is a rare tumor characterized by the extramedullary proliferation of immature granulocytic cells. It can occur as a primary disease or as a manifestation of acute myeloid leukemia, with an incidence of three to five percent in patients diagnosed with the condition. This report presents a thirty-seven-year-old female patient with acute monoblastic leukemia (French-American-British classification, subtype M5) who achieved complete remission after induction therapy, and allogeneic bone marrow transplantation. Six-teen months after achieving remission, the patient experienced a relapse in the pituitary gland, which was initially misdiagnosed as a pituitary adenoma. Following gross total surgical resection, the diagnosis of MS was confirmed. Adjuvant craniospinal radiotherapy and systemic therapy with azacitidine and venetoclax led to sustained remission without further complications. This case highlights the diagnostic and therapeutic challenges of MS, particularly in its rare presentation involving the pituitary gland. Radiotherapy, in combination with systemic treatment, played a crucial role in the management of this relapse. The findings emphasize the importance of a multidisciplinary approach in managing rare cases of MS to optimize patient outcomes.

Keywords: Acute myeloid leukemia, myeloid sarcoma, craniospinal radiotherapy, pituitary relapse

Introduction

Myeloid sarcoma (MS) is a rare hematologic malignancy characterized by the uncontrolled proliferation of immature granulocytic cells in extramedullary tissues. It may occur *de novo*, represent an extramedullary manifestation of acute myeloid leukemia (AML), or develop following hematopoietic stem cell transplantation. In the literature, it is also referred to as granulocytic sarcoma, chloroma, or extramedullary myeloid tumor.

The reported incidence of isolated *de novo* MS is approximately 2 per 100,000 in adults and 0.7 per 1,000,000 in children. When presenting concurrently with AML, the frequency ranges between 2-9% in adults and 10.9-23.3% in pediatric patients. In the post-transplant remission setting, its incidence has been reported between 5% and 12%.

MS most commonly involves the skin, lymph nodes, soft tissues, and bones, while central nervous system (CNS) localization is exceedingly rare and typically does not cause parenchymal damage. Østgaard et al. [1] found that MS was located in the craniospinal system in only 0.4% of AML patients. Pituitary involvement represents only a very small fraction of CNS cases, with fewer than 10 cases of pituitary or sellar region MS described in the literature to date (10, 12, 20).

In this report, we present a rare case of pituitary relapse of MS occurring after allogeneic hematopoietic stem cell transplantation in a female patient with AML and discuss its diagnostic and therapeutic aspects in the context of current literature.

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Case Report

A 37-year-old female patient presented with gingival bleeding, fatigue, and skin rash. Hematological analysis revealed a hemoglobin level of 9 g/dL, leukocyte count of 300,000/µL, and platelet count of 34 k/µL. Due to gingival hypertrophy, AML-M5 was suspected, and subsequent bone marrow biopsy confirmed acute monoblastic leukemia with the feline mcdonough sarcoma-like tyrosine kinase 3/internal tandem duplication (FLT3/ITD) mutation. The patient underwent induction therapy consisting of continuous administration of cytarabine for seven days, in combination with idarubicin during the first three days, followed by midostaurin administered from days 8 to 21. Following confirmation of the diagnosis with bone marrow biopsy results, consolidation therapy was initiated with high-dose cytarabine, complemented by midostaurin administered during days 8 to 21 of each cycle. The patient achieved complete remission with minimal residual disease (MRD) negativity. After one month, the patient underwent myeloablative conditioning and received an allogeneic bone marrow transplant from her sibling. At 1, 3, 6, and 12 months post-transplantation, the patient exhibited 100% chimerism and maintained MRD-negative status. No graft-versus-host disease, cytomegalovirus infection, or other complications were observed. Six-teen months after treatment completion, the patient presented with somnolence, visual impairment, polydipsia, and fatigue. Hematological, hormonal, and biochemical analyses revealed normal results. However, magnetic resonance imaging (MRI) of the pituitary gland revealed a 19×17 mm diffuse contrast-enhancing lesion extending from the stalk level to the superior optic chiasm, initially diagnosed as a pituitary adenoma (Figure 1). In March 2023, gross total resection was performed, and the diagnosis was confirmed of MS (Figure 2). At this juncture, a bone marrow biopsy demonstrated remission, 100% chimerism, and a negative FLT3/ITD mutation. Postoperative cranial MRI

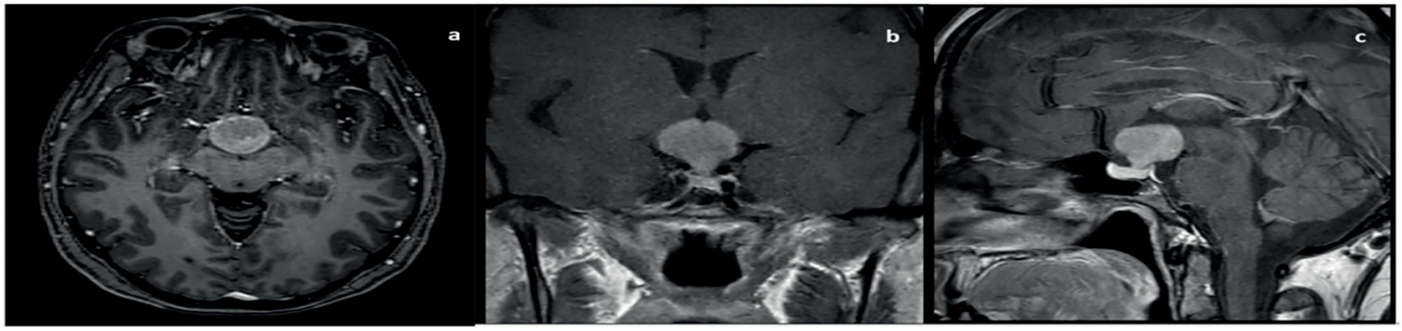

Figure 1. Preoperative contrast-enhanced brain magnetic resonance imaging (MRI) images, (a) axial section, (b) coronal section, (c) sagittal section. MRI of the pituitary gland revealed a 19×17 mm diffuse contrast-enhancing lesion extending from the stalk level to the superior optic chiasm

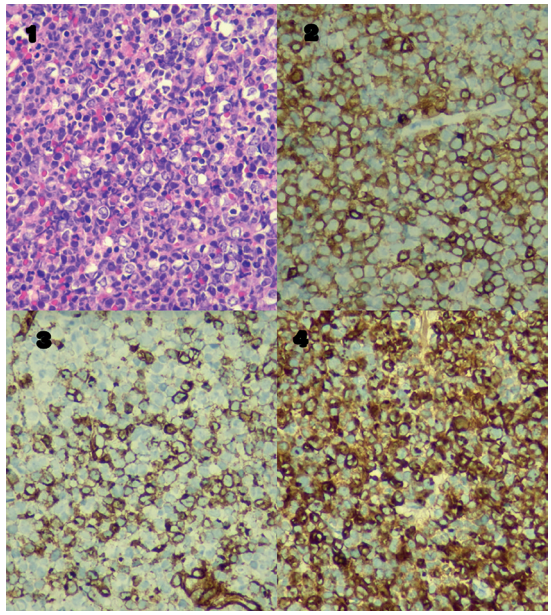

Figure 2. Pathology specimen images
1. The nucleus is irregularly elongated with occasional indentations, containing fine vesicular chromatin, a prominent nucleolus, and discohesive cells (hematoxylin and eosin, x400)
2. Positive reaction with cluster of differentiation 117 (CD117) (x400)
3. Positive reaction with CD34 (x400)
4. Positive reaction with myeloperoxidase (x400)

revealed an approximately 3 cm hemorrhage in the suprasellar region and postoperative changes in the right temporal area (Figure 3). One month after surgery, the patient received adjuvant craniospinal radiotherapy with 18 Gy in 10 fractions and an additional dose of 23.4 Gy in 13 fractions to the tumor bed (Figure 4). The patient continued to receive systemic treatment. At the third month after radiotherapy, the patient experienced restricted eye movements with limited upward gaze in the right eye, and limited outward gaze in the left eye. However, MRIs showed changes consistent with the post-surgical effects. The patient received 11 cycles of azacitidine and venetoclax for maintenance therapy, and remained in remission with negative bone marrow, and extramedullary involvement and 100% chimerism. The patient was monitored without complications and received supportive treatment for the pituitary insufficiency. The patient's data used in this study have been fully anonymized, and no identifiable information is included. Therefore, consent to publish is not required.

Discussion

MS is a rare tumor characterized by extramedullary localization of immature granulocytic cells. It is also known as a myeloblastoma, chloroma, or granulocytic sarcoma. The term chloroma is derived from the green staining of cells due to myeloperoxidase content [2]. This condition was first described in the orbit by Allan [3]. Regarding its pathogenesis, it is hypothesized that there is an aberrant expression of homing signals of leukemic blasts in the extramedullary region compared to the bone marrow [4]. Avni and Koren-Michowitz [5] reported that AML blast cells express chemokine receptors that are absent in normal bone marrow and peripheral blood blasts.

MS occurs most commonly with AML, followed by chronic myeloid leukemia. It can present *de novo* or as an initial symptom in patients who have not yet been diagnosed with AML. In addition, it may appear as an early relapse manifestation after bone marrow transplantation in patients with known AML. The incidence of AML in patients ranges from 3% to 5%. It can also develop as a transformation of myeloproliferative diseases or myelodysplastic syndromes [4]. Our patient presented with an MS relapse while in remission from AML after allogeneic bone marrow transplantation. MS can manifest at any age without a sex predilection [6]. A meta-analysis encompassing CNS MS cases reported a median age of 35 years, with a higher incidence observed in males [7]. MS occurs more frequently in cases of French-American-British classification M4/M5 types, cytogenetic abnormalities such as t, (8; 21), inversion 16, infant leukemias, chromosomal 11q abnormalities, cellular immunodeficiency, and following allogeneic stem cell transplantation [2]. Although the patient in this study did not exhibit any of these chromosomal abnormalities, the FLT-3/ITD mutation, which is associated with a poor prognosis, was present. Furthermore, allogeneic bone marrow transplantation, which increases the frequency of MS, constitutes a risk factor for patients [8]. A review conducted by Paydas et al. [8] determined that the interval between previous hematological diseases and MS diagnosis ranged from 5 to 60 months [9]. A meta-analysis by Lee et al. [6] reported a mean duration of 25.5 months between AML remission and MS diagnosis. This meta-analysis did not demonstrate the impact of the duration between AML and MS on patient mortality. The patient in this study experienced an MS relapse 17 months after AML remission. The most common sites for MS are the subperiosteal regions of the skull, paranasal sinuses, sternum, ribs, vertebrae, pelvis, lymph nodes, and the skin. CNS involvement is infrequent and typically manifests as leptomeningeal or extra-axial cranial bone-based masses. Parenchymal involvement rarely occurs subsequent to meningeal involvement [10]. Due to its rarity, MS presents diagnostic challenges, with an overall misdiagnosis rate of up to 40%. Biopsy remains the preferred diagnostic method [11].

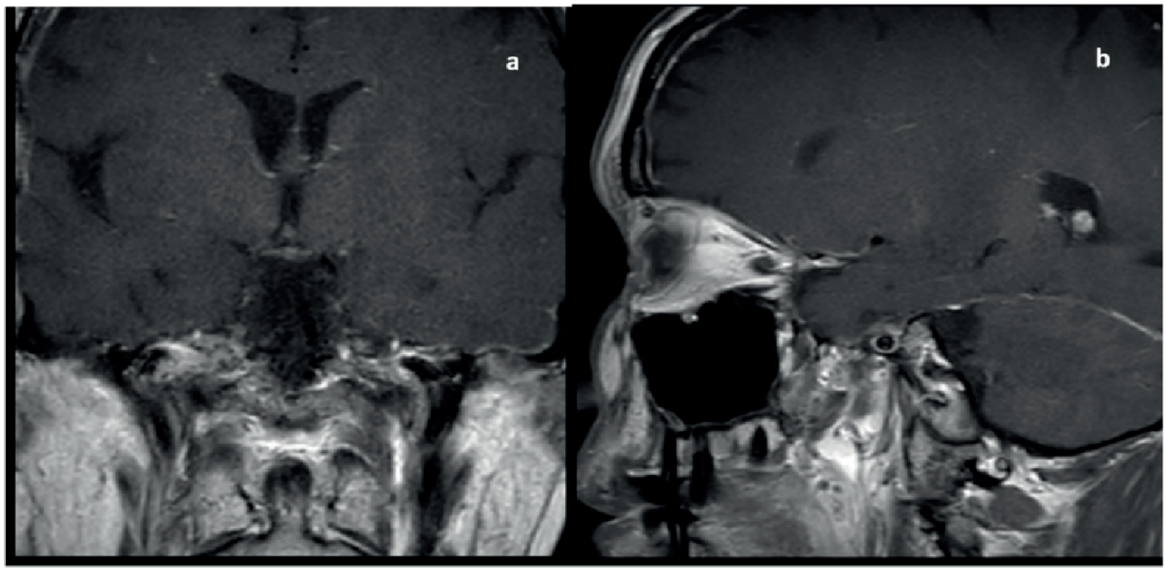

Figure 3. Postoperative cranial magnetic resonance imaging images, (a) coronal section, (b) sagittal section. An approximately 3 cm hemorrhage in the suprasellar region and postoperative changes in the right temporal area

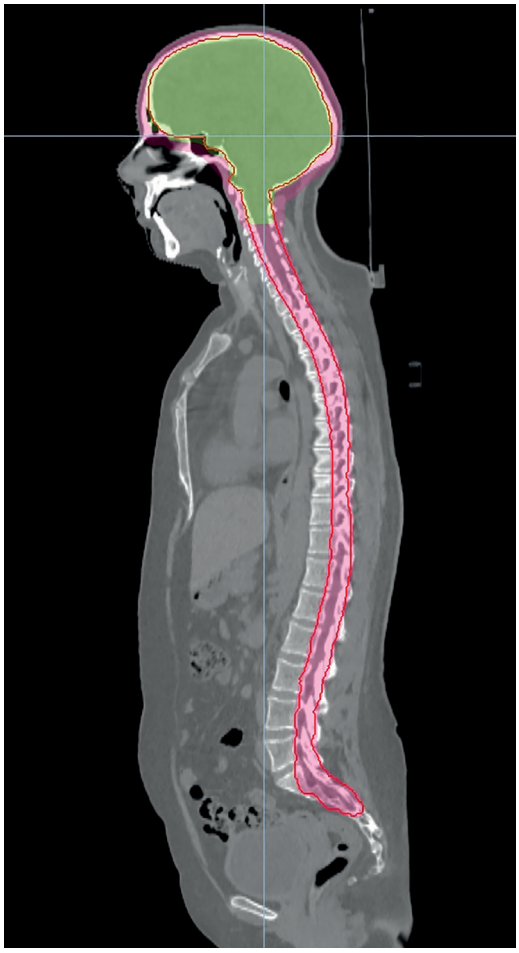

Figure 4. Treatment field of a patient undergoing craniospinal radiotherapy with helical intensity-modulated radiation therapy technique on a radixact device

MS infrequently presents as intracranial MS (IMS) of the skull [12]. The most prevalent locations for IMS are the temporal lobes, cerebellum, and falx or parasagittal regions, accounting for approximately 30.9% of IMS cases [12]. In the present case, the pituitary mass was initially diagnosed as pituitary adenoma. Differential diagnosis for pituitary masses includes adenoma, sarcoidosis, optic nerve glioma, aneurysm, craniopharyngioma, Rathke cleft cyst, and teratoma [13]. Based on our current knowledge and literature review, this case is considered one of the exceedingly rare reported instances of isolated pituitary MS. Clinical symptoms of MS vary according to anatomical location. Symptoms arise from the tumor's mass effect or organ dysfunction, due to infiltration. In instances of pituitary involvement, symptoms can include visual disturbances, diabetes insipidus, and panhypopituitarism [14]. In the present case, the patient exhibited symptoms of decreased vision, somnolence, and polydipsia due to the mass effect of the pituitary tumor. The treatment of MS remains a subject of debate, with a lack of prospective randomized controlled trials [15]. Chemotherapy regimens employed for AML are applicable to both isolated MS and MS occurring concurrently with AML. Patients with isolated multiple sclerosis MS who undergo local treatment exhibit a higher rate of progression compared to those receiving systemic treatment [16,17]. Imrie et al. [17] observed that chemotherapy in isolated MS patients was associated with improved overall survival rates. While the efficacy of combining radiotherapy with chemotherapy is not fully elucidated, radiotherapy is frequently administered in conjunction with chemotherapy. Radiotherapy has been demonstrated to enhance progression-free survival in MS patients, although its impact on overall survival remains uncertain [18]. Lan et al. [15] reported no significant difference in overall survival between groups receiving chemotherapy with radiotherapy and those receiving chemotherapy without radiotherapy in MS patients. The treatment of IMS primarily comprises radiotherapy and chemotherapy. A meta-analysis conducted by Lee et al. [6] demonstrated that the addition of chemotherapy or radiotherapy to IMS treatment significantly reduces mortality. No correlation was identified between surgical resection and its extent and mortality. Although surgical resection is essential for histological diagnosis, its role remains uncertain. It is observed that surgical resection may enhance quality of life, in patients experiencing neurological symptoms or mass effect [7]. In the case reported, the patient initially received systemic treatment for AML and subsequently presented with symptoms related to the pituitary mass while in remission. Post-surgical radiotherapy was administered with the intention of preventing local recurrence, and subsequent chemotherapy resulted in remission.

Conclusion

In conclusion, MS is a neoplasm that may arise from the myeloid lineage in any part of the body, while sellar involvement is exceedingly rare. Nonetheless, it should be considered in the differential diagnosis of pituitary and parasellar masses, and it is important to note that no consensus currently exists regarding its optimal management. Radiotherapy provides rapid local control and symptomatic relief when administered in combination with systemic therapy, may further improve clinical outcomes. Reporting such cases is of great importance, as it contributes to the accumulation of collective experience and guides the development of treatment strategies in these rare clinical scenarios.

Ethics

Informed Consent: The patient's data used in this study have been fully anonymized, and no identifiable information is included. Therefore, consent to publish is not required.

Footnotes

Authorship Contributions

Surgical and Medical Practices: S.M., Z.G., A.U., Concept: S.M., Z.G., F.A., Design: S.M., Z.G., Ç.Ş.K.Y., F.A., Data Collection or Processing: S.M., A.U., Analysis or Interpretation: S.M., Ç.Ş.K.Y., Literature Search: S.M., Writing: S.M., Z.G., F.A.

Conflict of Interest: No conflict of interest was declared by the authors.

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