Turkish Archives of Pediatrics

SCIENTIFIC LETTER
DOI: 10.5152/TurkArchPediatr.2023.22216

Myeloid Sarcoma Presenting as Brain Tumor in Pediatric Acute Myeloid Leukemia Case

Hüseyin Avni Solgun, Sibel Tekgündüz, Özlem Terzi, Cengiz Bayram, Ali Ayçiçek

Department of Pediatric Hematology and Oncology, Health Sciences University, Başakşehir Çam and Sakura Training and Research Hospital, İstanbul, Turkey

Myeloid sarcoma (MS) or granulocytic sarcoma (chloroma) is a rare extramedullary tumor composed of myeloblasts or immature myeloid cells that can destroy tissues. It usually occurs together with acute myeloid leukemia (AML). In MS, skin and orbital localization are the most common sites in childhood.

Myeloid sarcoma is a rare extramedullary tumor arising from immature myeloid cells. Its relationship with acute or chronic leukemias is well known.[1] The incidence of MS is between 3% and 5% in patients with AML. When MS and AML occur together, the prognosis for MS is much worse.[2] M2, M4, and M5 types, which are subtypes of AML in some morphological classifications, are more closely related to MS.[3,4] Although MS cases usually occur together with leukemia, they can rarely be seen in the absence of leukemia. In these cases, other solid tumors such as lymphoma, neuroblastoma, or rhabdomyosarcoma may be misdiagnosed. Although the localization of MS in children is mostly in the orbital region, it can also be involved in the skin, bone, lymph nodes, and soft tissues.[5] Orbital MS is generally seen in children under 10 years of age and is responsible for 2%-6% of all pediatric orbital tumors.[6,7] In our case in this study, tumor localization was atypical; it was located in the parietotemporal region. We present this case to update the information about MS in the literature and emphasize the incidence due to its atypical clinical location and rarity.

Here, we present a 2-year-old male patient referred to our pediatric hematology oncology clinic with a left parietotemporal mass (Figure 1). The laboratory test results were for white blood cells (WBC) 71 320 × 10^9/L, hemoglobin 8.3 g/dL, and platelets 81 000 × 10^9/L, respectively. In computed tomography (CT) of the brain; a mass lesion of approximately 8 × 9 cm in size extending from the left parietotemporal lobe to the inferior, extending toward the skin/subcutaneous, causing lytic changes in the left posterior temporal bone was observed (Figure 2). A rapid peripheral blood smear (PBS) was planned to identify the leukocytosis and cytopenia. The observation of PBS reported a 5%-6% myeloid blast formation. The flow cytometric evaluation of peripheral blood showed 10% myeloblasts. A neurosurgery consultation was planned. Pulse prednisolone 40 mg/kg for the first 3 days and contiuned 30 mg/kg for 4 days have been started as a preventative treatment from brain edema. Soft tissue

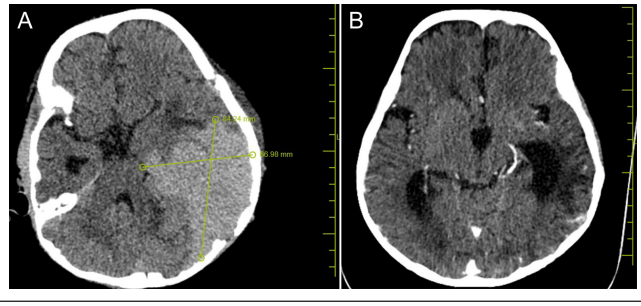

Figure 1. (A) First-day physical view of mass in parietotemporal skull. (B) The markable involution of physical mass view on the 28th day of treatment in parietotemporal skull.

Corresponding author:
Hüseyin Avni Solgun
✉ hsynavn@gmail.com
Received: September 12, 2022
Accepted: January 9, 2023
Publication Date: May 2, 2023

Cite this article as: Solgun HA, Tekgündüz S, Terzi Ö, Bayram C, Ayçiçek A. Myeloid sarcoma presenting as brain tumor in pediatric acute myeloid leukemia case. *Turk Arch Pediatr.* 2023;58(3):347-350.

tumors and bone marrow metastasis or AML and granulocytic sarcoma were the initial diagnostic features. A flowcytometric peripheral blood test was reported to be 8% myeloblast. In confirmation of diagnosis AML, 20% and a higher blast percentage was needed. In this period to decrease the WBC count to safe values cytosine arabinoside treatment of 100 mg/m^2 dosage for 24 hours was initiated. On the second day of follow-up, the WBC count was 42 000 $\times$ 10^9/L, and bone marrow aspiration (BMA) and cerebrospinal fluid were planned. The flow cytometric evaluation of BMA showed 10% myeloblasts (Figure 3). The genetic panel for AML was sent from bone marrow aspirate. On the 3rd day of treatment, the genetic tests reported t(8,21) positivity in this patient. The AML diagnosis was confirmed and AML Berlin Frankfurt Möunster (AML-BFM) 2019 protocol was initiated (Figure 4). The markable involution of tumoral mass from the first day to the 28th day of treatment of cerebral CT and their physical view are displayed in Figures 1A, 1B and 2A, 2B respectively.

Since MS localization can be in all body parts, clinical findings in MS are very variable, especially if there is no bone marrow involvement, and this may even lead to difficulties in diagnosis. In patients diagnosed with MS, secondary solid tumors such as malignant lymphoproliferative disorders, Ewing's sarcoma, thymoma, melanoma, round blue cell tumors, or poorly differentiated carcinoma have been reported in 25%-47% of cases.[8,9]

Therefore, as in our case; for differential diagnosis from other tumors and diseases; diagnostic testing for an accurate diagnosis of MS should include magnetic resonance imaging (MRI) and/or CT scanning to assess tumor size and location, morphological and flow cytometric analysis of the bone marrow, and even genetic evaluations if necessary. Especially in patients without bone marrow involvement, the diagnosis should be confirmed by effective tests such as bone marrow and peripheral blood or tumor biopsy and immunohistochemical staining.[4]

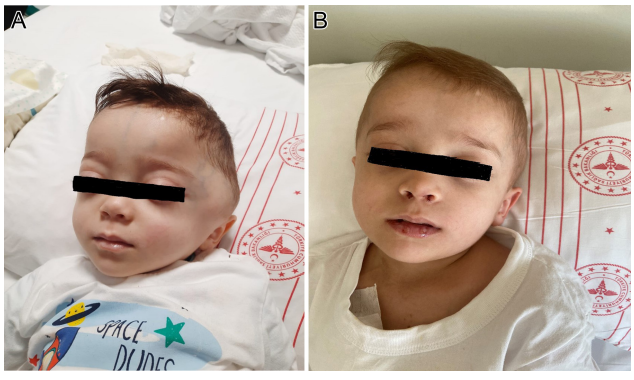

Figure 2. (A) The first day of cerebral CT imagining showed a mass lesion of approximately 8.4 $\times$ 6.6 cm in size extending from the left parietotemporal lobe to the inferior, extending toward the skin/subcutaneous, causing lytic changes in the left posterior temporal bone is observed. Perifocal edema is noticeable. The mass lesion extending toward the anterior of the left cerebellar hemisphere in the posterior fossa caused the left cerebellar hemisphere to be pushed laterally. (B) On the 28th day of treatment, cerebral CT images showed a total recovery in the mass lesion and lateral ventricle, 3rd ventricle was observed bilaterally, and there was a hyperadenosis appearance in the left temporal lobe and it was considered in favor of encephalomalaci a-glyphosate. The left lateral ventricle temporal horn was dilated compared to the symmetrical needle. Midline structures of the cerebral parenchyma were observed in the normal position. CT, computed tomography.

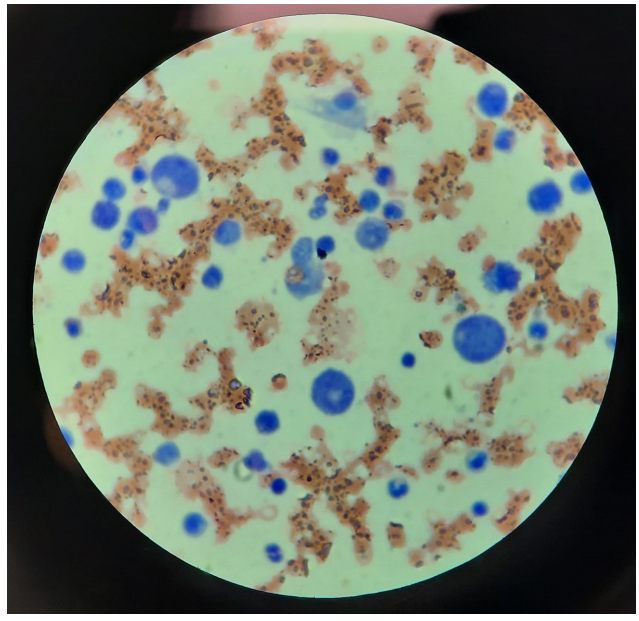

Figure 3. The immature cells have abundant, frequently basophilic cytoplasm, with variable numbers of often indistinct, sometimes coalescent granules. As such immature cells are >10% the diagnosis becomes AML-M2. AML-M2 shows significant maturation and Auer rods are frequent (promyelocytes-myelocytes). AML, acute myeloid leukemia.

Acute myeloid leukemia treatment protocols are used in the treatment of MS, and surgery and/or radiotherapy may be preferred for symptomatic lesions or tumors causing local organ dysfunction, as in our case.[10] Since the most common localizations in pediatric cases with MS are the skin and the orbita, an atypical locality of MS that is rarely seen in MS is presented in this case to emphasize this feature.

Pamir et al[11] in their work presented the rarity and treatment approaches of 2 extramedullary orbital MS cases without bone marrow involvement. In these cases, a total of 7 days of high-dose methylprednisolone (HDMP) therapy (3 days 30 mg/kg/day and 4 days 20 mg/kg/day) followed by AML-BFM conventional chemotherapy has been applied. For 2 cases, then, the 2004 treatment protocol was continued. As a result of HDMP treatment, we noted a significant improvement regarding eye complaints. In our case, a significant involution was detected in the parietotemporal mass in the first week after chemotherapy was started, as reported by the Pamir et al.[11]

In another study, Aslantaş et al[12] reported that a thoracolumbar mass was detected by MRI in a 4-year-old boy who presented with a complaint of hemiparesis. In this patient, 30% of blasts consistent with AML were detected in bone marrow aspiration. The diagnosis of MS was made by a biopsy performed from the mass and pathological evaluation.[12] In this case, complete neurologic recovery was achieved after 2 weeks of fractionated 18 Gy radiotherapy and additional dexamethasone treatment. Conventional chemotherapy was continued with the AML-Berlin Frankfurt Münster 2012 protocol. Following AML induction chemotherapy, it provided both remission and significant

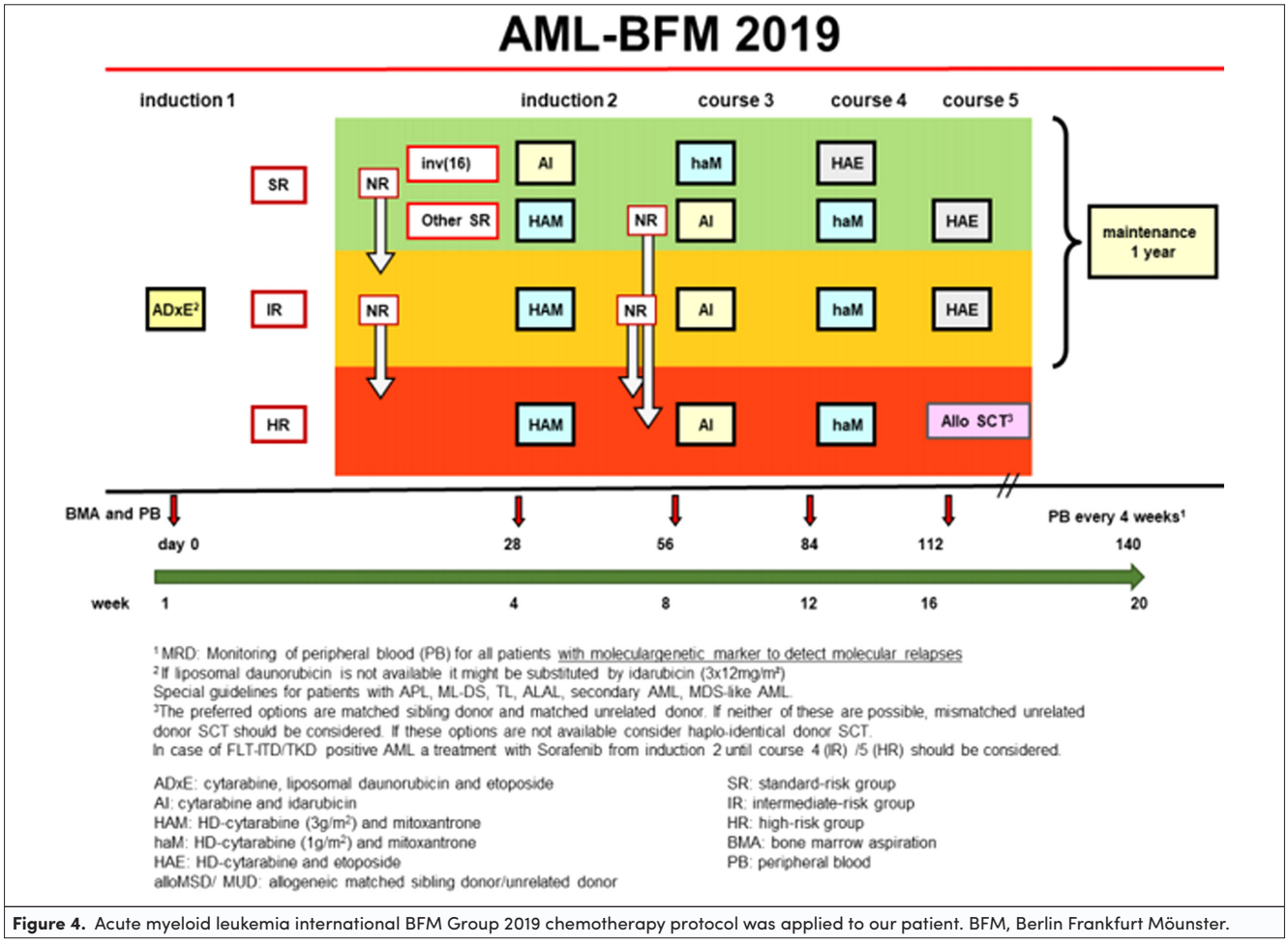

Figure 4. Acute myeloid leukemia international BFM Group 2019 chemotherapy protocol was applied to our patient. BFM, Berlin Frankfurt Möunster.

involution of the mass. The patient was reported to be still in remission with no residual tumor on the follow-up MRI. In our case, radiotherapy was not preferred because of the significant involution response in the tumor with dexamethasone treatment.

In conclusion, conventional chemotherapy protocols for AML are still the mainstay of treatment in MS, and surgery and/or radiotherapy for symptomatic lesions or tumoral mass causing local organ dysfunction may be considered. The most common sites of application in children with MS are the skin and orbital sites. Dexamethasone and conventional chemotherapy are the first line of treatment. In our case, we presented an atypical location, an external parietotemporal mass extending to the inner part of the left parietal lobe. This settlement may require differential diagnosis even from brain tumors.

Informed Consent: The patient's parents gave informed written consent for their personal or clinical details along with any identifying images to be published in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – H.S., S.T., O.T., C.B., A.A.; Design – H.S., S.T., O.T.; Supervision – H.S., A.A.; Materials – H.S., S.T., O.T., C.B., A.A.; Data Collection and/or Processing – H.S., S.T., O.T.; Analysis and/or Interpretation – H.S., O.T. – Literature Review – H.S., S.T., O.T.; Writing – H.S., S.T.; Critical Review – H.S., S.T., O.T., C.B., A.A.

Acknowledgment: The authors are thankful to all individuals who have contributed to this study.

Declaration of Interests: The authors have no conflict of interest to declare.

Funding: This study received no funding.

REFERENCES

1. Inoue D, Nagai Y, Kimura T, et al. Refractory de novo myeloid sarcoma: a case report and therapeutic strategy based on bone marrow minimal residual disease. *Int J Hematol.* 2009;90(1):120-123. [CrossRef]
2. Maka E, Lukats O, Tóth J. Orbital tumour as initial manifestation of acute myeloid leukemia: granulocytic sarcoma: case report. *Pathol Oncol Res.* 2008;14:209-211.
3. Porto L, Kieslich M, Schwabe D, Zanella FE, Lanfermann H. Granulocytic sarcoma in children. *Neuroradiology.* 2004;46(5):374-377. [CrossRef]
4. Slavcheva V, Lukanov T, Tzvetkov N. Two cases of extramedullary myeloid tumor in patients with continuous remission of acute myeloblastic leukemia. *J Buon.* 2008;13:589-592.
5. Lee SG, Park TS, Cheong JW, et al. Preceding orbital granulocytic sarcoma in an adult patient with acute myelogenous leukemia with t(8;21): a case study and review of the literature. *Cancer Genet Cytogenet.* 2008;185(1):51-54. [CrossRef]

6. Hmidi K, Zaouali S, Messaoud R, et al. Bilateral orbital myeloid sarcoma as initial manifestation of acute myeloid leukemia. *Int Ophthalmol*. 2007;27(6):373-377. [CrossRef]
7. Chung EM, Murphey MD, Specht CS, Cube R, Smirniotopoulos JG. From the archives of the AFIP. Pediatric orbit tumors and tumorlike lesions: osseous lesions of the orbit. *RadioGraphics*. 2008;28(4): 1193-1214. [CrossRef]
8. Bakst RL, Tallman MS, Douer D, Yahalom J. How I treat extramedullary acute myeloid leukemia. *Blood*. 2011;118(14):3785-3793. [CrossRef]
9. Redner A, Kessel R. Acute myeloid leukemia. In: Lanzkowsky P, Lipton JM, Fish JD, eds. *Lanzkowsky's Manual of Pediatric Hematology and Oncology*. 6th ed. Amsterdam, Elsevier; 2016.
10. Samborska M, Derwich K, Skalska-Sadowska J, Kurzawa P, Wachowiak J. Myeloid sarcoma in children - diagnostic and therapeutic difficulties. *Contemp Oncol (Pozn)*. 2016;20(6):444-448. [CrossRef]
11. Pamir I, Tavil Betul T, Yaralı B, Demir Nese, Mualla AÇ. Extramedullary orbital granulocytic sarcoma without bone marrow involvement: a report of two cases. *Pediatr Hematol Oncol*. 2011;28(1): 65-70.
12. Arslantaş E, Bayram C, Odaman I, et al. A rare cause of paraplegia: myeloid sarcoma. *Turk J Hematol*. 2018;35:200-216.