Myeloid Sarcoma presentetion as brain tumor in pediatric acute myeloid lekeumia

Huseyin Solgun[1], Sibel Tekgunduz[1], and Cengiz Bayram[1]

[1]Saglik Bilimleri Universitesi

April 05, 2024

Abstract

Case: We present a 2-year-old male patient who was referred to the pediatric hematology oncology clinic with left pariatotemporal mass. In computed tomography (CT) of brain; a mass lesion of approximately 8x9 cm in size extending from the left parietotemporal lobe to the inferior, extending towards the skin / subcutaneous

Myeloid Sarcoma presentetion as brain tumor in pediatric acute myeloid lekeumia

Authors: Huseyin Avni Solgun[1], Cengiz Bayram[2] , Sibel Tekgündüz[3]

Short title: Myeloid Sarcoma presentetion as brain tumor

SOLGUN Huseyin A., M.D., Department of Pediatric Hematology and Oncology, Health Sciences University, Basaksehir Cam and Sakura Training and Research Hospital /Istanbul

TEKGÜNDÜZ Sibel, M.D., Department of Pediatric Hematology and Oncology, Health Sciences University, Basaksehir Cam and Sakura Training and Research Hospital /Istanbul

BAYRAM Cengiz., Assoc. Prof. M.D., Department of Pediatric Hematology and Oncology, Health Sciences University, Basaksehir Cam and Sakura Training and Research Hospital /Istanbul

Key clinical massege

In our case the tumor localization was pariatotemporal as it is not usuall presentation in Myeloid sarcoma (MS) of childhood. By this knowledge; we presented this case report to refresh the data in the literatüre and to emphasize the atypicall clinical prensentation, rare incidence and management protocols of granulocytic sarcoma.

Absract

Background: 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 involved tissue and typically occurs concurrently with acute myeloid leukemia (AML). It can also occur in association with accelerated-phase chronic myeloid leukemia or myelodysplastic syndrome; as an extramedullary relapse of AML, including in the post-bone marrow transplant setting; and occasionally as the first presenting manifestation, even before bone marrow involvement. Bone, periosteum, skin, orbit, lymph nodes, the gastrointestinal tract, and the 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 .

Case: We present a 2-year-old male patient who was referred to the pediatric hematology oncology clinic with left pariatotemporal mass. In computed tomography (CT) of brain; a mass lesion of approximately 8x9 cm in size extending from the left parietotemporal lobe to the inferior, extending towards the skin

/ subcutaneous, causing lytic changes in the left posterior temporal bone was observed. (**figure 1a)** . A neurosurgery consultation was planned. The brain antieudema protective treatment with prednisilone 40 mgr/kg first 3 days and subsuquent to 30 mgr/kg 4 days was initiated. Soft tissue tumor and the bone marrow metastasis or AML and granulocytic sarcoma were the initiall diagnositc features. Flowcytometric peripheral blood test was reported %8 myeloblast. In this period to decrease the WBC count: 76000 10'9/L to safe values cytosine arabinoside treatment of 100mg/m2 dosage for 24 hours was iniated. The control values in treatment hours and days are in**table 1** . In the second day of follow ups, WBC count was 42000 10'9/L and bone marrow aspiration (BMA) and cerebrospinal fluid was planned. The flow cytometric evaulation of BMA showed %10 myelobalasts. The genetic panel for AML was sent form bone marrow aspirate. In the 3th day of treatment; the genetic tests reported t(8,21) positivity in this paitent. The AML diagnosis was comfirmed and AML BFM 2019 protocol was initiated to the patient. The involution of tumoral mass from first day and first month of CT and physical view are in **figure 1a-b and 2a-b** respectively.

Conclusion: The most common presentation sites in children with MS is skin and orbital localizations. Dexamethosene and conventianol chemathearphy is first steps in treatment. Our case presented exterior pariatotemporal mass extending to interior left pariatel lobe with extraordinary feature. This presentation may need differantial diagnosis even with brain tumors.

Keywords: Childhood, Acute myeloid lekeumia, Myeloid Sarcoma, Brain tumors

Introduction

Granulocytic sarcoma (GS) is a rare extramedullary tumor of immature myeloid cells and is frequently associated with acute or chronic leukemias [1]. GS is seen in 3% to 5% of patients with acutemyeloid leukemia (AML).When GS is associated with AML, the prognosis is very poor [2]. Certainmorphological subtypes of AML (M2,M4, M5) are more frequently associated with extramedullary myeloid tumors [3, 4]. Although most cases of GS arise synchronously with leukemias, GS may rarely be seen prior to leukemic development, and most of these cases are falsely diagnosed as lymphoma, neuroblastoma, or rhabdomyosarcoma, among others. In the pediatric age group, GS is generally localized in the orbital area, but it may also be found in the skin, bone, lymph node, and soft tissues [5]. Orbital GS, primarily a childhood disease and rarely seen in adults, is mostly observed in children under 10 years of age and is responsible for 2% to 6% of childhood orbital tumors [6–7].

In our case the tumor localization was pariatotemporal as it is not usuall presentation. By this knowledge; we presented this case report to refresh the data of granulocytic sarcoma in the literatüre and to emphasize the atypicall clinical prensentation and rare incidence.

Case Presentation

We present a 2-year-old male patient who was referred to the pediatric hematology oncology clinic with left pariatotemporal mass. The laborautary test results were for white blood cell (WBC) 71320 10'9/L, hemaglobuline 8,3 g/dl and platalet 81000 10'9/L respectevily. In computed tomography (CT) of brain; a mass lesion of approximately 8x9 cm in size extending from the left parietotemporal lobe to the inferior, extending towards the skin / subcutaneous, causing lytic changes in the left posterior temporal bone was observed. (**figure 1a)** A rapid peripheral blood smear (PBS) planned to identify the leukocytosis and cytopenia.The observation of PBS reported %5-6 myeloid blast formation. The flow cytometric evaulation of peripheral blood showed %10 myelobalasts. A neurosurgery consultation was planned. The brain antieudema protective treatment with prednisilone 40 mgr/kg first 3 days and subsuquent to 30 mgr/kg 4 days was initiated. Soft tissue tumor and the bone marrow metastasis or AML and granulocytic sarcoma were the initiall diagnositc features. Flowcytometric peripheral blood test was reported %8 myeloblast. In confirmation of diagnosis AML, %20 and higher blast percentage was needed. In this period to decrease the WBC count to safe values cytosine arabinoside treatment of 100mg/m2 dosage for 24 hours was iniated. In the second day of follow ups, WBC count was 42000 10'9/L and bone marrow aspiration (BMA) and cerebrospinal fluid was planned. The flow cytometric evaulation of BMA showed %10 myeloblasts. The genetic panel for AML was sent form bone marrow aspirate. In the 3th day of treatment; the genetic tests reported t(8,21)

positivity in this paitent. The AML diagnosis was comfirmed and AML BFM 2019 protocol was initiated to the patient.The markable involution of tumoral mass from first day to first month serebral CT and physical view are in **figure 1a-b and figure 2a-b** respectively.

Discussion

MS may occur at any site of the body, and therefore clinicalmanifestations of MS exhibit diversity depending on the specificlocation and size, which leads to significant diagnostic challenges,in particular in patients without initial bone marrow involvement. Malignant lymphoproliferative disorders, Ewing's sarcoma, thymoma, melanoma, round blue cell tumors,or poorly differentiated carcinoma has been reported at a

rate of 25%-47% in patients subsequently diagnosed with MS. [8,9].

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 the size and location of the tumor and for distinguishing the tumor from other lesions, morphological and flow cytometric analysis of bone marrow and peripheral blood, or biopsy of the tumor 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 [10]. 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.

Pamir Isık and et al. Reported; 2 extramedullary orbital granulocytic sarcoma (GS) cases without bone marrow involvement in view of their rarity and also to reevaluate the treatment approach in this disease. Seven days of high-dose methyl prednisolone (HDMP) treatment (3 days 30 mg/kg/day and 4 days 20 mg/kg/day)was administered initially, and subsequently AcuteMyeloid Leukaemia–Berlin Frankfurt M¨ unster (AML-BFM) 2004 treatment protocol was continued for 2 cases. Eye findings of the cases resolved considerably with HDMP treatment. In our case after chematheraphy initiated there was markable involution in pariatofrantal mass in first week as Pamir study decleraed. *

Aslantaş and et al reported a 4-year-old boy admitted with the complaint of hemiparesis and a subsequent thoracolumbar mass was detected by magnetic resonance imaging (MRI) Bone marrow aspiration showed 30% blasts compatible with AML. The pathology of the mass revealed MS. [11]. 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 the AML-Berlin Frankfurt Münster 2012 protocol and achieved both remission and mass reduction following AML induction chemotherapy. The patient is still in remission without any residual tumor on follow-up MRI. In our case we iniated dexamethasone treatment reveresly radiothearphy was not preferred.

In conclusion; AML-based protocols are stil remians the main course reatment of MS and in indivudal cases, surgery and/or radiotherapy may be indicated for symptomatic lesions or tumoral mass causing local organ dysfunction. The most common presentation sites in children with MS is skin and orbital localizations. Dexamethosene and conventianol chemathearphy is first steps in treatment. Our case presented exterior pariatotemporal mass extending to interior left pariatel lobe with extraordinary feature. This presentation may need differantial diagnosis even with brain tumors.

References

1- Inoue D, Nagai Y, Kimura T. Refractory de novo myeloid sarcoma: a case report and therapeutic strategy based on bone marrow minimal residual disease. Int J Hematol. 2009;90:120–123.

2- Maka E, Lukats O, T ´ oth J. Orbital tumour as initial manifestation of acute myeloid leukemia: gran- ´ ulocytic sarcoma: case report. Pathol Oncol Res. 2008;14:209–211.

3- Porto L, Kieslich M, Schwabe D. Granulocytic sarcoma in children. Neuroradiology. 2004;46:374–377.

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. 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;185:51–54.

6- Hmidi K, Zaouali S, Messaoud R. Bilateral orbital myeloid sarcoma as initial manifestation of acute myeloid leukemia. Int Ophthalmol. 2007;27:373–377.

7- Chung EM, Murphey MD, Specht CS. From the archives of the AFIP. Pediatric orbit tumors and tumorlike lesions: osseous lesions of the orbit. Radiographics. 2008;28:1193–1214.

8. Bakst RL, Tallman MS, Douer D, Yahalom J. How I treat extramedullary acute myeloid leukemia. Blood 2011;118:3785-3793.

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;20:444-448. 5. Almond LM, Charalampakis M, Ford SJ, Gourevitch D, Desai A. Myeloid sarcoma: presentation, diagnosis, and treatment. Clin Lymphoma Myeloma Leuk 2017;17:263-267

11. Esra Arslantaş , Cengiz Bayram , Işık Odaman and et al. A Rare Cause of Paraplegia: Myeloid Sarcoma. Turk J Hematol 2018;35:200-216

Figure legends

Figure 1a: The first day of serebral CT imagining showed a mass lesion of approximately 8,4x6,6 cm in size extending from the left parietotemporal lobe to the inferior, extending towards the skin / subcutaneous, causing lytic changes in the left posterior temporal bone is observed. Perifocal edema is noticeable. The mass lesion extending towards the anterior of the left cerebellar hemisphere in the posterior fossa caused the left cerebellar hemisphere to be pushed laterally.

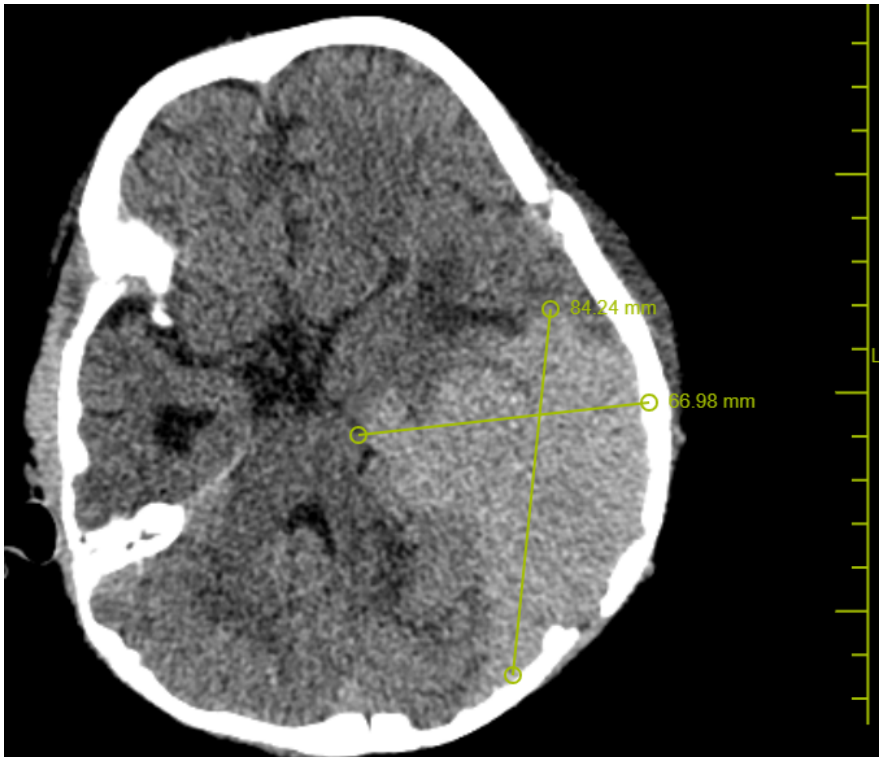

Figure 1b: In the 28th day of treatment serebral CT imagining showed a total recovery in the mass lesion and lateral ventricle, 3rd ventricle was observed dilarerally, there was a hypodenosis appearance in the left temporal lobe and it was considered in favor of encephalomalacia-glyphosis.The left lateral ventricle temporal horn was dilated compared to the symmetrical needle.Midline structures of the cerebral parenchyma were observed in the normal position.

Hosted file

`image2.tmp` available at https://authorea.com/users/373217/articles/710332-myeloid-sarcoma-presentetion-as-brain-tumor-in-pediatric-acute-myeloid-lekeumia

Figure 2a: First day physical view of mass in parietotemporal skull.

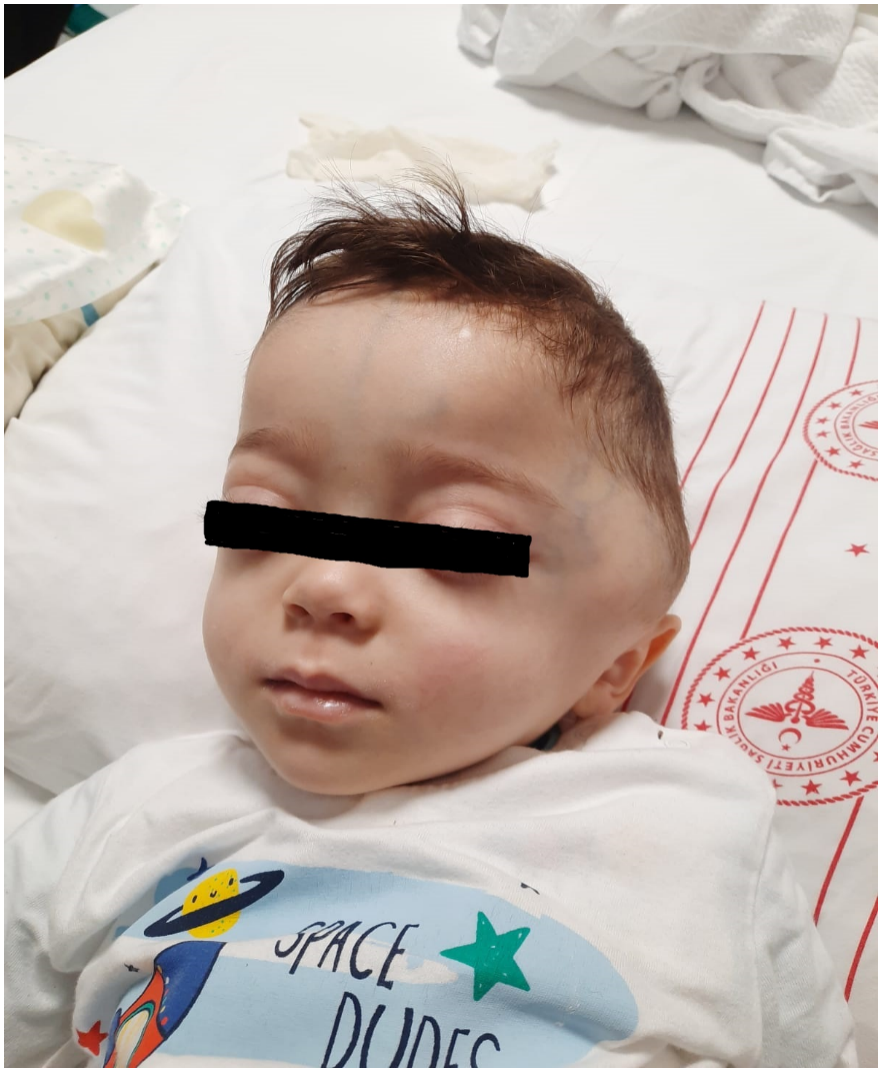

Figure 2b: The markable involution of physical mass view in 28th day of treatment in parietotemporal skull.