

# CAN ANY CEREBRAL MASS OR HEMATOMA BE LEUKEMIA? CHLOROMA

Sema Yılmaz MD, İbrahim Bayram MD, Fatih Erbey MD, Derviş Aslamacı MD, Atila Tanyeli MD

Çukurova University Medical Faculty, Department of Paediatrics, Adana

## ABSTRACT

Granulocytic sarcoma (chloroma) is a malignant extra-medullary neoplasm of myeloid precursor cells. It is seen usually concurrent with or follows the diagnosis of acute myelogenous leukemia or other myeloproliferative disorders. Granulocytic sarcomas usually involve bones, periosteum, lymph nodes and skin. Involvement of the head and neck region is uncommon. Chloroma should be considered in the differential diagnosis of intracranial lesions. The computed

tomography (CT) appearance of a cranial granulocytic sarcoma can be indistinguishable from a cerebral hematoma. We present a 12 year-old girl with scalp chloroma that was initially misdiagnosed as subdural effusion and later cerebral hematoma. An accurate and early diagnosis of this rare condition can prevent treatment delay and lead to good prognosis.

**Key Words:** Granulocytic sarcoma, chloroma, acute myeloid leukemia. *Nobel Med 2012; 8(2): 118-120*

## HERHANGİ BİR SEREBRAL KİTLE YA DA HEMATOM LÖSEMİ OLABİLİR Mİ? KLOROMA

### ÖZET

Granülositik sarkoma (kloroma), myeloid öncü hücrelerinin kemik iliği dışında malign özellikte bir tümördür. Genellikle, aynı zamanda ya da takiben akut myeloid lösemi ya da diğer myeloproliferatif hastalıklar görülür. Granülositik sarkomalar, genellikle, kemik iliği, periost, lenf nodları ve deri ile ilişkilidir. Baş-boyun bölgesi ile ilişkileri yaygın değildir. Kloro-

ma, kafaiçi lezyonların ayırıcı tanısında düşünülmemelidir. Bir kranyal granülositik sarkomanın bilgisayarlı tomografisi bir serebral hematomdan ayrılabilir. Biz, başlangıçta subdural hematoma ve sonrasında serebral hematoma ile karıştıran, kafa derisinde kloroma ile başvuran 12 yaşındaki bir kız çocuğunu sunduk. Doğru ve erken tanı ile bu nadir durum, tedavide gecikme önlenir ve hastalığın seyrinde iyileşmeye sebep olabilir.

**Anahtar Kelimeler:** Granülositik sarkom, kloroma, akut myeloid lösemi. *Nobel Med 2012; 8(2): 118-120*

## INTRODUCTION

Granulocytic sarcoma (GS), also called chloroma or myeloblastoma, is a solid tumor composed of leukemic myeloblasts and/or myeloid precursors.<sup>1</sup> It rarely develops in patients with out symptoms of leukemia, either in peripheral blood or in bone marrow. Differential diagnosis is often difficult, and should include acute lymphoblastic leukemia, large cell non-Hodgkin's lymphoma, lymphoblastic lymphoma and Ewing's sarcoma. These tumors most frequently occur during the course of acute myeloid leukemia (AML) or in remission from leukemia. In most of these patients, following the occurrence of chloroma, an overt acute myeloid leukemia develops within 1 to 49 months.<sup>2</sup> Head and neck region are rarely involved. The leptomeninges, brain

parenchyma and intracranial vessels can be involved in the central nervous system. Granulocytic sarcoma is rarely seen in brain parenchyma. With central nervous system involvement, the lesions often start in the bone marrow of the calvarium and then spread to the dura. In any event, the presence of a chloroma is certainly the sign of poor prognosis.<sup>3-6</sup>

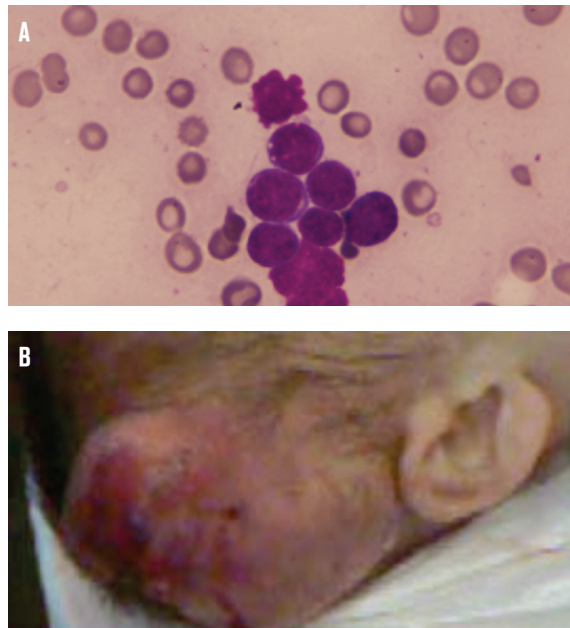
## CASE REPORT

A 12-year-old-girl was presented to the emergency department complaining of headache that had a history of gradual increase during the previous week and that the pain had been worsening recently despite her use of oral analgesic medication. She denied any head trauma. Her past medical history was unremarkable. The laboratory investigations, →

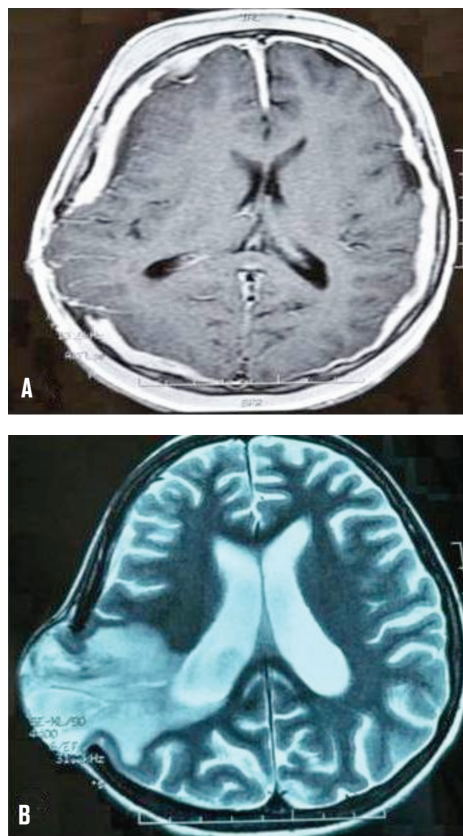
including a complete blood count, erythrocyte sedimentation rate, and basic metabolic profile were all within normal limits. She underwent CT scanning of the head, and T1- and T2-weighted images of MRI demonstrated subdural effusion in the parietooccipital region. She was treated with temporary external subdural drainage. She was discharged to home and she was able to ambulate at the time of discharge. She returned one month later with a growing floppy mass on her right parietooccipital region and complaint of fever. Brain magnetic resonance imaging (MRI) was promptly performed. Increased signal in the post-contrast images was stronger on the parameningeal region and the protrusion of cerebral parenchyma in parietooccipital region was seen. On physical examination, the patient appeared cachectic and fatigued. She was alert and oriented to her surroundings. Her examinations showed a temperature of 103.3 °F (39.6 °C), a blood pressure of 110/90 mmHg, and a heart rate of 120 beats/minute. She had no hepatosplenomegaly, and no other lymph node was palpable. Cardiovascular, respiratory and nervous system examination did not reveal any abnormality. Diagnostic laboratory studies revealed a white blood cell count of  $4.0 \times 10^3/\mu\text{L}$ , but a hematocrit of 22.5%, a hemoglobin value of 8.0 g/dL, a mean corpuscular volume of 69.7 fL, and a platelet count of  $100 \times 10^3/\mu\text{L}$ . The basic metabolic panel, liver and kidney function tests, urinalysis, and coagulation studies were in the normal range. The patient underwent bone marrow examination for further evaluation of anemia. She was diagnosed to have AML M1 according to the FAB classification (Figure 1A). Induction chemotherapy for acute leukemia was immediately started. One month later, a growing solid mass was noticed in the subdural drainage site (Figure 1B). Since her parents did not give a consent, neither fine-needle aspiration nor core biopsy of the mass could have been performed. We considered that this mass might be chloroma due to the diagnosis of AML. During chemotherapy febrile neutropenia was developed. Despite the treatment, the patient died 3 months later because of systemic dissemination of leukemia.

## DISCUSSION

Granulocytic sarcoma, also known as chloroma, is an extramedullary tumor composed of immature cells with myeloid differentiation. It occurs in only 2.9-9.1% of the cases of all acute myeloid leukemia and is more common in childhood.<sup>7,8</sup> Granulocytic sarcoma is rarely reported in the scalp and temporal bone. The tumor cells can infiltrate in the periosteum from the bone marrow. From here, the organs were



**Figure 1:** A. Myeloblasts in the bone marrow aspiration. B. Chloroma in the scalp.



**Figure 2:** The protrusion of cerebral parenchyma in parietooccipital region on MRI (A) and contrasted CT (B).

involved probably due to the anatomical proximity with the bone marrow.<sup>9-11</sup> It usually occurs either during the initial course of acute myeloid leukemia or after remission; however, it may rarely precede the clinical manifestation of acute leukemia, therefore the diagnosis might be difficult.<sup>12</sup> AML-specific chemotherapy is recommended for the →

treatment of GS.<sup>13</sup> In our case, initially subdural effusion occurred and gradual growing of a solid mass from craniectomy side was noticed during the course of acute myeloid leukemia. In retrospective assessment, a diagnosis of granulocytic sarcoma was done. It is reported that chloroma may be initially misdiagnosed such as a chronic subdural hematoma or meningioma in the literature.<sup>14</sup> Even it was not the same, subdural effusion might be a messenger for further developing of chloroma in our case. Therefore, appropriate chemotherapy could have begun early and unnecessary operation could be prevented in this case. Granulocytic sarcoma was reported as an isodense or slightly hypodense well-defined mass, before contrast media injection and homogeneously enhanced after contrast enhancement on CT images.<sup>15</sup> On MRI, isointense signal intensity to that of muscle or marrow on T1- and T2-weighted images are seen.<sup>16</sup> Therefore,

lymphoma, hematoma or meningioma can be misdiagnosed on imaging studies. Chloroma can be differentiated from subdural hematoma with using of gadolinium on brain MRI.<sup>17</sup> In our patient, just before growing of the solid mass, imaging studies showed thick parameningeal structure, parietal subdural effusion and the protrusion of cerebral parenchyma in parietooccipital region and cerebral hematoma in herniated and adjacent paranchyma on MRI and contrasted CT, respectively (Figure 2A,2B). Even if we could not have performed the mass biopsy, chloroma was diagnosed retrospectively with the appearance of acute myeloid leukemia. In conclusion, scalp chloroma should be considered in the differential diagnosis of subdural effusion or cerebral hematoma. We aimed to report this rare type of chloroma case to pay attention not to be misdiagnosed as subdural effusion or cerebral hematoma.



<b>C</b>	<b>CORRESPONDING AUTHOR:</b> Sema Yılmaz, MD Çukurova University Medical Faculty, Pediatric Oncology, Balcalı Hospital 01330, Adana <a href="mailto:semayilmaz@hotmail.com">semayilmaz@hotmail.com</a>
<b>✓</b>	<b>DELIVERING DATE:</b> 16 / 10 / 2009 • <b>ACCEPTED DATE:</b> 28 / 05 / 2010

#### REFERENCES

- Byrd JC, Edenfield WJ, Shields DJ, Dawson NA. Extramedullary myeloid cell tumors in acute nonlymphocytic leukemia: a clinical review. *J Clin Oncol* 1995; 13: 1800-1816.
- Breccia M, Mandelli F, Petti MC, et al. Clinico-pathological characteristics of myeloid sarcoma at diagnosis and during follow-up: report of 12 cases from a single institution. *Leuk Res* 2004; 28: 1165-1169.
- Lee YH, Lee NJ, Choi EJ, Kim JH. Granulocytic sarcoma (chloroma) presenting as a lateral neck mass: initial manifestation of leukemia: a case report. *Eur Arch Otorhinolaryngol* 2006; 263: 16-18.
- Suzer T, Colakoglu N, Cirak B, et al. Intracerebellar granulocytic sarcoma complicating acute myelogenous leukemia: a case report and review of the literature. *J Clin Neurosci* 2004; 11: 914-917.
- Mansi JL, Selby PJ, Carter RL, Powles RL, McElwain. Granulocytic sarcoma: a diagnosis to be considered in unusual lymphoma syndromes. *Postgrad Med J* 1987; 63: 447-449.
- Pui MH, Fletcher BD, Langston JW. Granulocytic sarcoma in childhood leukemia: imaging features. *Radiology* 1994; 190: 698-702.
- Bulas RB, Laine FJ, Das Narla L. Bilateral orbital granulocytic sarcoma (chloroma) preceding the blast acute phase of acute myelogenous leukemia: CT findings. *Pediatr Radiol* 1995; 25: 488-489.
- Scheinberg DA, Maslak P, Weiss M. Acute leukemias. In: Devita VT, Hellman S, Rosenberg SA, editors, *Cancer: principles and practice of oncology*. Philadelphia: Lippincott Raven 1997; 2293-2316.
- Lee B, Fatterpekar GM, Kim W, Som PM. Granulocytic sarcoma of the temporal bone. *Am J Neuroradiol* 2002; 23: 1497-1499.
- Nayak DR, Balakrishnan R, Raj G, et al. Granulocytic sarcoma of the head and neck: a case report. *Am J Otolaryngol* 2001; 22: 80-83.
- Porto L, Kieslich M, Schwabe D, Zanella FE, Lanfermann H. Granulocytic sarcoma in children. *Neuroradiology* 2004; 46: 374-377.
- Neiman RS, Barcos M, Berard C, et al. Granulocytic sarcoma: a clinicopathologic study of 61 biopsied cases. *Cancer* 1981; 48: 1426-1437.
- Reinhardt D, Creutzig U. Isolated myelosarcoma in children-update and review. *Leuk Lymphoma* 2002; 43: 565-574.
- Velasco F, Ondarza R, Quiroz F, Arista J. Meningioma-like intracranial granulocytic sarcoma (chloroma). Radiologic and surgical findings. *Rev Invest Clin* 1993; 45: 473-478.
- Pomeranz SJ, Hawkins HH, Towbin R, Lisberg WN, Clark RA. Granulocytic sarcoma (chloroma): CT manifestations. *Radiology* 1985; 155:167-170.
- Kransdorf MJ, Jelinek JS, Moser RP Jr. Imaging of soft tissue tumors. *Radiol Clin North Am* 1993; 31: 359-372.
- Smidt MH, de Bruin HG, van't Veer MB, van den Bent MJ. Intracranial granulocytic sarcoma (chloroma) may mimic a subdural hematoma. *J Neurol* 2005; 252: 498-499.