

Extraaxial Chloroma of the Cerebellopontine Angle

Serebellopontin Açıda Yerleşen Ekstraaksiyel Kloroma

ABSTRACT

A chloroma or granulocytic sarcoma is an extramedullary leukemia. It can be encountered at any anatomical location, but until now only three cases have been reported in the cerebellopontine angle. We present an 8-year old patient with an extraaxial chloroma of the cerebellopontine angle to highlight this very rare and malignant pathology in the differential diagnosis of cerebellopontine angle tumors. The presented case, being the fourth chloroma in the cerebellopontine angle, occurred in the absence of relapse which is very unusual for these lesions. Chloroma should be remembered as a very rare and a malignant pathology in the differential diagnosis of pediatric cerebellopontine angle tumors.

KEY WORDS: Acute myeloblastic leukemia, Cerebellopontine angle, Chloroma, Tumor

ÖZ

Kloroma ya da granülositik sarkom ekstramedüller bir lösemidir. Granülositik sarkom herhangi bir anatomik lokalizasyonda yerleşebilir, ancak bugüne kadar serebellopontin açıda sadece üç vaka bildirilmiştir. Biz bu makalede serebellopontin açıda ekstraaksiyel kloroma ile gelen 8 yaşındaki bir hastayı takdim ederek serebellopontin açıda yerleşebilen bu kötü huylu patolojiye dikkat çekmek istedik. Takdim edilen vaka serebellopontin açıda yerleşen dördüncü kloroma vakasıdır ve relaps olmadan ortaya çıkmıştır bu özellikte bu tip lezyonlar için oldukça atipiktir. Kloroma serebellopontin açıda yerleşen pediyatrik yaş grubu kötü huylu patolojilerin ayırıcı tanısında hatırlanmalıdır.

ANAHTAR SÖZCÜKLER: Akut myeloblastik lösemi, Kloroma, serebellopontin açısı, Tümör

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INTRODUCTION

Chloroma or granulocytic sarcoma (GS) is an extraaxial leukemia. It can be encountered at any anatomical location, but until now only three cases have been reported in the cerebellopontine angle (CPA). We present an 8-year old patient with an extraaxial chloroma of the CPA to highlight this very rare and malignant pathology in the differential diagnosis of CPA tumors.

CASE REPORT

This 8-year old girl with a previous diagnosis of acute myeloblastic leukemia (AML) was referred to our department from a pediatric hospital with a loss of consciousness. She had received a chemotherapy (AML-BFM 93) protocol and was considered to be in remission confirmed by blood tests and bone marrow aspiration results. Cranial computerized tomography revealed a hyperdense lesion in the left CPA with a central necrotic area.

Neurologic examination disclosed a fully unconscious patient with a GCS score of 6. Magnetic resonance imaging (MRI) revealed a left extraaxial CPA lesion with dimensions of 4.5x4x3 cm. The lesion was isointense with gray matter on T1W images and minimally hyperintense on T2W images. Areas suggestive of hemorrhage were seen on T1W images as hyperintense and on T2W as hypointense regions. Marked enhancement was observed after intravenous contrast media along with the mass effect on brain stem and cerebellum (Figure 1A,B,C). The fourth ventricle was compressed resulting in a triventricular hydrocephalus. MR spectroscopy

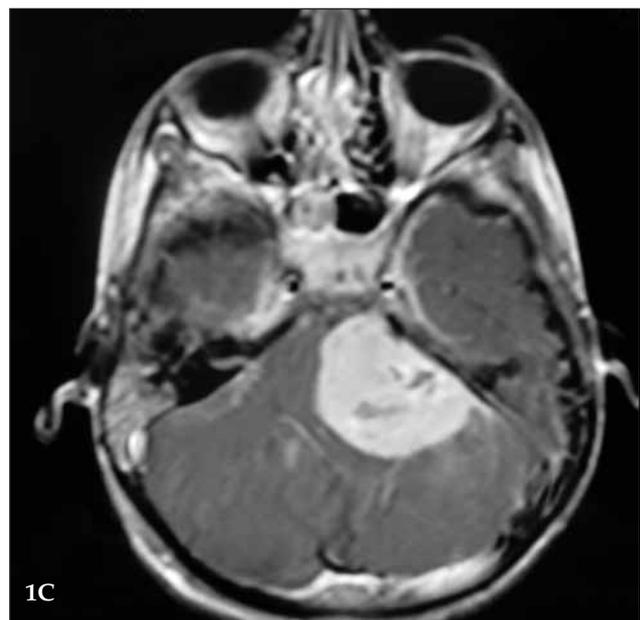
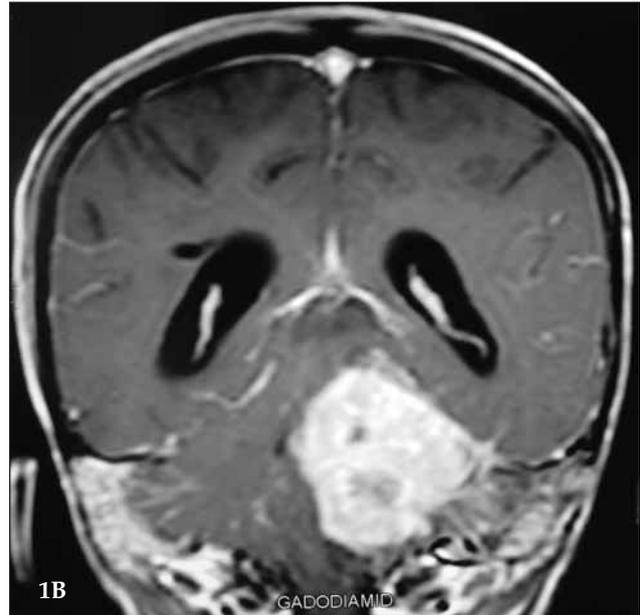
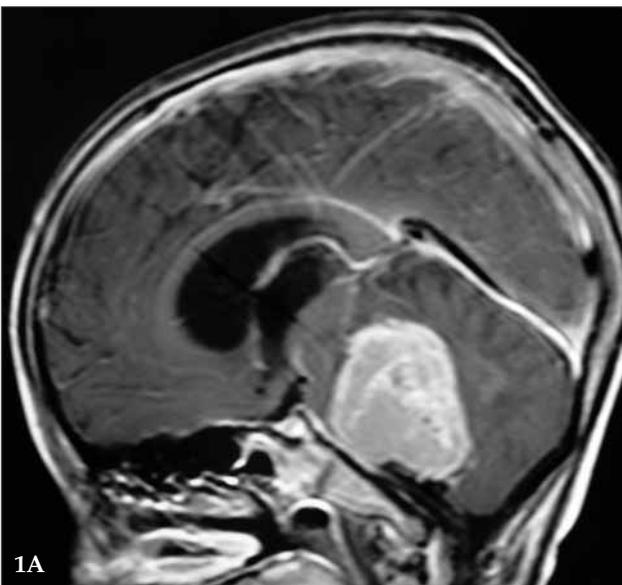


Figure 1: T1-W iv contrast sagittal (A), coronal (B) and axial (C) images showing an extra-axial mass lesion of the cerebellopontine angle with marked enhancement.

revealed an intralesional obvious choline peak with TE:135 and 270 msn values. The choline/creatinine ratio was increased.

The patient was operated on December 25, 2005 following an external ventricular drainage. The extraaxial mass was highly vascular and only subtotal decompression of the lesion could be achieved. Histopathological examination of the lesion revealed a necrotic tumoral infiltration within the cerebellum (Figure 2A). The tumor was

composed of sheets of immature myeloblasts and included numerous areas of mitosis (Figure 2B). The cells showed eosinophilic cytoplasm and nuclear irregularity in focal areas. There were multiple tingible body macrophages among the neoplastic cells. Immunohistochemical analysis of myeloperoxidase (MPO) displayed diffusely striking reactivity in the cytoplasm of the immature myeloblastic cells. The histopathologic diagnosis was consistent with granulocytic sarcoma. The patient did not show any clinical improvement and died on the 16th postoperative day.

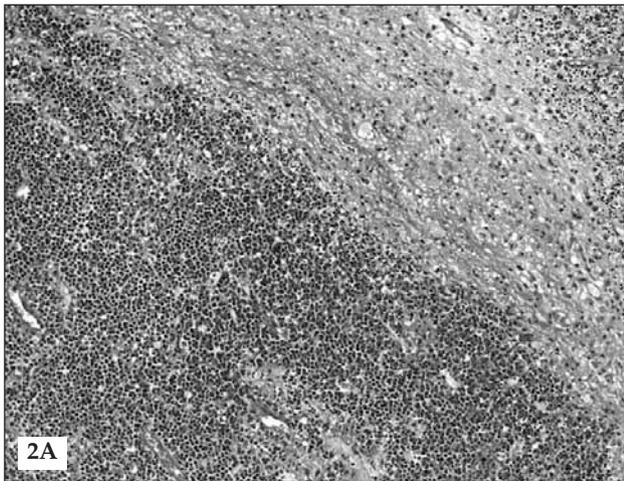


Figure 2A: Sections included necrotic tumoral infiltration within the cerebellum (H&E x 100).

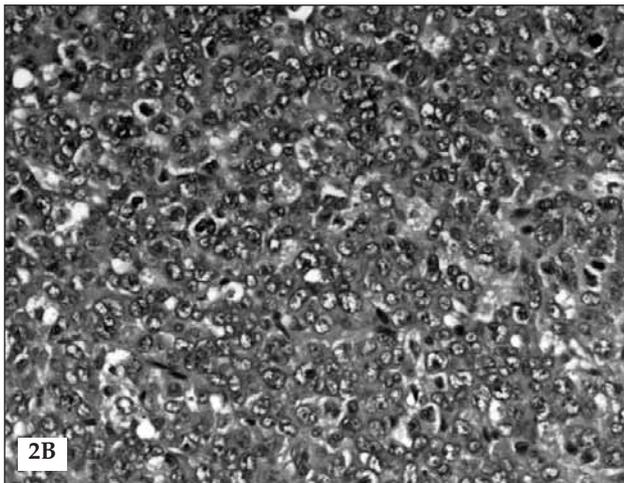


Figure 2B: The tumor consisted of sheets of immature myeloid cells and included numerous areas of mitosis (arrows) (H&E x 400).

DISCUSSION

Chloroma or GS is an extramedullary leukemia. It can be encountered at any anatomical location, the common sites being the orbit, skin, bones, paranasal sinuses and epidural areas (4). Extracranial and intracranial (2) and spinal (4) involvement have been observed and reported, but until now only three cases of GS occurring in CPA (1) and four cases localized in the cerebellum have been reported (3).

The presented case, being the fourth chloroma in the CPA, occurred in the absence of relapse. Isolated recurrence of AML during bone marrow remission, especially presenting as a solid intracranial mass, is a rare entity. The treatment for intracranial chloroma is either radiotherapy, since chloromas are radiosensitive, or surgery for large lesions (5). In our case we have chosen surgery due to the life-threatening clinical status of the patient.

Chloroma should be remembered as a very rare and a malignant pathology in the differential diagnosis of pediatric CPA tumors.

REFERENCES

1. Gokcan MK, Batikhan H, Calguner M, Tataragasi AI. Unilateral hearing loss as a presenting manifestation of granulocytic sarcoma (chloroma). *Otol Neurotol* 2006; 27:106-9.
2. Nishimura S, Kyuma Y, Kamijo A, Maruta A. Isolated recurrence of granulocytic sarcoma manifesting as extra- and intracranial masses--case report. *Neurol Med Chir (Tokyo)* 2004; 44:311-16.
3. Psiachou-Leonard E, Paterakis G, Stefanaki K, Mikraki-Christou V, Haidas S. Cerebellar granulocytic sarcoma in an infant with CD56+ acute monoblastic leukemia. *Leuk Res* 2001; 25:1019-1021.
4. Seo HY, Kim SJ, Choi JW, Lee JB, Kang CH, Seo BK, Kim BS. Unusual relapse of acute myeloid leukemia: granulocytic sarcoma presenting as an acute paraplegia. *Ann Hematol* 2006; 85:196-7.
5. Sham RL, Phatak PD, Kouides PA, Janas JA, Marder VJ: Hematologic neoplasia and the central nervous system. *Am J Hematol* 1999; 62:234-8.