Multiple primary cranial granulocytic sarcoma: a case report

Multipl primer kraniyal granülositik sarkoma: olgu sunumu

Fuat Torun[1], Emel Avci[2]

[1]Harran University, Faculty of Medicine, Department of Neurosurgery, Sanliurfa, Turkey
[2]Mersin University, Faculty of Medicine, Department of Neurosurgery, Mersin, Turkey

Corresponding author: Fuat TORUN, Harran University, Faculty of Medicine, Department of Neurosurgery, Sanliurfa/TURKEY, Tel: 90 414 314 11 70, Fax: 90 414 315 11 81, E-mail: fuatorun@hotmail.com

Abstract
Intracranial primary granulocytic sarcoma is a solid tumor that occurs rarely. A 10-year-old female presented with large and multiple primary granulocytic sarcomas in the cranium. In this report, we discuss primary granulocytic sarcoma disease, including clinical presentation, diagnosis, and treatment.
Key words: Primary granulocytic sarcoma, proptosis, cranium

Özet
İntrakraniyal primer granülositik sarkoma solid bir tümor olup oldukça nadir görülür. On yaşında kız çocuğu, kranyumda büyük ve multipl yerleşim gösteren primer granülositik sarkoma ile prezante oldu. Bu olgu sunumunda primer granülositik sarkoma hastalığının klinik görüntüsü, tanısı ve tedavisi tartışılmıştır.
Anahtar Kelimeler: Primer granülositik sarkom, propitoz, kranyum

Introduction

Granulocytic sarcoma (GS) is an extramedullary tumor composed of leukemic myeloblasts and/or myeloid precursors. It is also known as chloroma because of the greenish hue secondary to the presence of intracellular myeloperoxidase. Although it can occur anywhere in the body, it most commonly has epidural, mediastinum, bone, lymph node, skin and soft tissue localization (6). GS occurs most frequently in the pediatric population in association with acute myelogenous leukemia (AML), but it may also be associated with other myeloproliferative diseases (6, 7). It may develop before, during or after the onset of systemic leukemia; GS that precedes leukemia is called primary granulocytic sarcoma (7). Herein, we present an extremely rare case of primary GS, and the clinical presentation, diagnosis, and treatment of the lesion are discussed.

Case report

A 10-year-old healthy girl presented to our department with a sudden proptosis and pain of the left eye. On examination, she had unilateral irreducible proptosis and left abducens nerve palsy. There was no congestion of the eyelids. Visual acuity and visual field were intact. Before admission, she was healthy, with a normal daily life, and had no history of anemia or mass lesion. Her physical examination was normal, and no pathological enlargement of lymph nodes was noticed. Biochemical and hematological evaluations were normal. The tumor markers β2-microglobulin and alpha-fetoprotein were negative.

Computed tomography (CT), which was performed on admission, demonstrated a large tumor extending from the medial orbital wall to the paranasal sinuses, in addition to an epidural mass at the level of the vertex. Magnetic resonance imaging (MRI) showed a mass originating from the medial wall of the orbit, which aligned towards the ethmoid sinus and pushed the bulbus oculi anterolaterally. The mass was hypointense on T2- and isointense on T1 series, and it was homogeneously contrasted in post-contrast series. MRI also showed another distinct epidural mass at the convexity level, which was hypointense on T2- and isointense on T1 series (Figure 1).

One of the masses was hemorrhagic with epidural localization and was eroding the bone of the superior sagittal sinus at the level of the vertex, and the other extended from the orbital medial region toward the ethmoid sinus and lower conchae; both were totally excised by a combined bifrontal craniotomy and left orbital osteotomy. The portion of the mass that extended toward the lower conchae was excised by transnasal endoscopy. On gross examination, the mass was greenish brown, soft and hemorrhagic. The histological investigation showed immature cells that resembled primitive cells and the pathological diagnosis was a GS (Figure 2). The patient was discharged without any additional neurological deficits on the 14th day of her surgery and has been incorporated into the chemotherapy program by the Pediatric Hematology Unit. On the follow-up visit at the 3rd post-operative month, her proptosis had completely recovered.

Discussion

GS, a solid tumor originating from immature granulocytes, has been frequently observed in AML patients, although it can occur in any part of the body in other types of myeloproliferative diseases. A retrospective study by Pui et al. reported the frequency of tumor localization as follows: 24% in subcutaneous tissue, 24% in the orbit, 11% in a paranasal sinus, 11% in a lymph

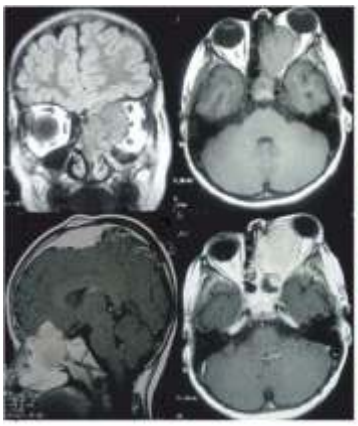
node, and 5% in bone and the periosteum (8). In case of AML, its frequent central nervous system localization is the orbit and at its peripheral region (8). Our case was a multifocal intracranial GS, which located in the orbit, ethmoidal sinus and the epidural distance at the level of the vertex.

The diagnosis of GS is important in order to achieve a complete remission by aggressive chemotherapy and/or radiotherapy. Nevertheless, due to the presence of normal hematological findings, it is difficult to make a definite diagnosis by means of radiological examination only. CT and MRI are not specific in differentiating granulocytic neoplasms from the other intracranial tumors (8). The lesion may show an increased attenuation area with normal limits and peripheral contrast enhancement on CT (3). On MRI, intracranial GS is hypo- or iso-intense on both T1- and T2-weighted images, and shows a homogeneous enhancement following contrast administration (8). The gadolinium enhancement differentiates GS from hematomas and abscesses, but it is impossible to differentiate GS from lymphoma, meningioma or pseudotumor by these imaging techniques (9). Meanwhile, these tumors could appear similar to mesoderm-derived tumors by means of their localizations. For a definite diagnosis, histological analysis is mandatory. The operation material is also used to analyze specific chromosomal abnormalities that can guide the clinician in deciding on the appropriate chemotherapy protocol (2).

The clinical findings are related to the mass effect of the tumor and the compression on the cranial nerves. As there are no hematological anomalies and no possibility for a definite radiological diagnosis in primary GS cases, biopsy or specimen obtained through excisional surgery may lead to a definitive diagnosis (7). The role of surgery for primary cranial chloromas is controversial and a diagnostic biopsy is usually recommended first (4). As in our case, surgery may be indicated in the presence of the tumoral mass effect or a progressive neurological deficit. Meanwhile, in primary GS cases, total or subtotal resection of the tumor is not crucial, but starting anti-leukemic treatment is extremely important (7). This information about the management of GS cases is based on the fact that the non-leukemic period following the diagnosis is short in patients receiving regional radiotherapy and surgical treatment (10). This period is long in patients who received chemotherapy (10). Thus, early diagnosis is important in GS cases.

GS is associated with myeloproliferative diseases, and in patients both with and without a history of a myeloproliferative disease, recognition of a GS is a poor prognostic sign (5). GS may also be a sign of relapse after bone marrow transplantation or clinical deterioration in a leukemic patient (1). Most of the primary GS cases proceed to overt leukemia within 2 to 4 years (6), in spite of an appropriate treatment.

Due to the presence of normal hematological results and lack of specific radiological findings, it is difficult to make a definite diagnosis in the early period of primary GS. Although surgical treatment may help to improve the progressive neurological impairment, the primary choice of treatment is chemotherapy in cases of GS. This pathology should be considered especially in the pediatric age group with no history of leukemia. Polychemotherapy should be started as soon as possible.



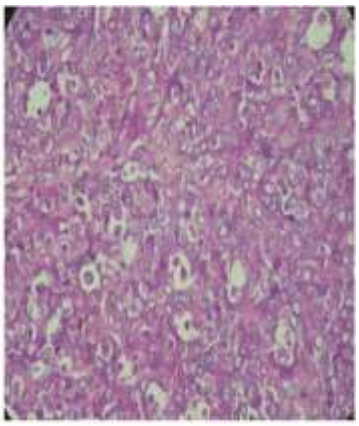


Figure 1: *The coronal and axial views of magnetic resonance imaging demonstrating isointense tumors on T1-weighted images (upper). T1-weighted images showing a diffuse gadolinium-enhancement on sagittal and axial views (lower).*
Figure 2: *Tumoral mass composed of immature myeloblastic cells showing numerous mitotic figures (Hematoxylin & Eosin x 400).*

Yazarlarla ilgili bildirilmesi gereken konular (Conflict of interest statement) : Yok (None)

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