JBNC Brazilian Journal of Neurosurgery
Jornal Brasileiro de Neurocirurgia

Case Report

Granulocytic Sarcoma (Chloroma) as a Cause of Acute Hydrocephalus in a Child with Acute Myeloid Leukemia

Sarcoma Granulocítico (Chloroma) como Causa de Hidrocefalia Aguda em Crianças com Leucemia Mielóide Aguda

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ABSTRACT

Chloromas or granulocytic sarcomas represent the extramedullary form of leukemia, which can be located anywhere in the body. Nonetheless, as of yet there is no case of this neoplasia reported as a cause of acute hydrocephalus in children secondary to cerebellar compression.

We report on a case of posterior fossa chloroma manifested as hydrocephalus, and discuss differential diagnosis, disease evolution and treatment.

Key words: *Acute Myeloid Leukemia, Chloroma, Granulocytic Sarcoma, Hydrocephalus*

RESUMO

Cloromas ou sarcomas granulocíticos constituem na forma extramedular da leucemia e podem ser encontrados em qualquer localização anatômica.

Apresentamos o caso de criança com cloroma da fossa posterior para destacar essa rara patologia maligna no diagnóstico diferencial dos tumores da fossa posterior.

Palavras-chave: *Leucemia Mielóide Aguda, Sarcoma mielóide, Hidrocefalia*

Introduction

Leukemia account for the most common neoplasias found in pediatric patients, corresponding to about 30% of all malignant disorders in patients younger than 15 years of age. Acute Lymphocytic Leukemia (ALL) represents about 75% of all the cases in children. The acute myeloid forms (AML) represent 20% and the chronic myeloid form approximately 5% of the cases[1].

Granulocytic sarcomas are the extramedullary form of the disease – also called chloromas, because of their greenish macroscopic aspect (because of myeloperoxidase, produced by the myeloid cells), of solid masses made of immature granulocytic cells, which occur in 2 to 14% of the acute leukemias[6-10]. They may affect multiple body systems, and the most commonly reported are: bones, paranasal sinuses, skin, gastrointestinal tract, uterus, ovaries and breast. In the central nervous system (CNS), it frequently involves the dura matter and orbit with bad prognosis[9,14,15]. In the present investigation, we report a case of a child with granulocytic sarcoma of the posterior fossa with intracranial hypertension due to hydrocephalus as the first manifestation of leukemia.

Case Report

A 1 year-and-10-month-old, previously healthy male child, started severe holocranial headache, which lasted for three days, associated with frequent vomiting. At hospital admission the child was drowsy and with Glasgow Coma Scale (GCS) score 11 (E3, V3, M5) He did not have any cranial nerve abnormality and was in opisthotonos (Fig. 1). His axial mobility was unaltered; however presenting global hyperreflexia with bilateral Babinski's reflex.

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Received Jul 13, 2013. Accepted Oct 27, 2013

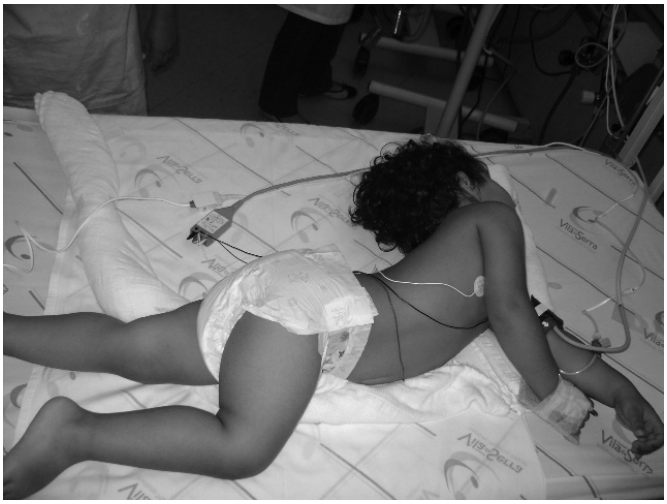

Figura 1: Opisthotonos at admission.

CT-scan revealed supratentorial hydrocephalus and an expansive retrocerebellar lesion, apparently extradural, mildly hyperdense. There were still some nodular lesions on the posterior neck surface, which were palpable in the back of his neck - painless and without skin alterations (Fig. 2).

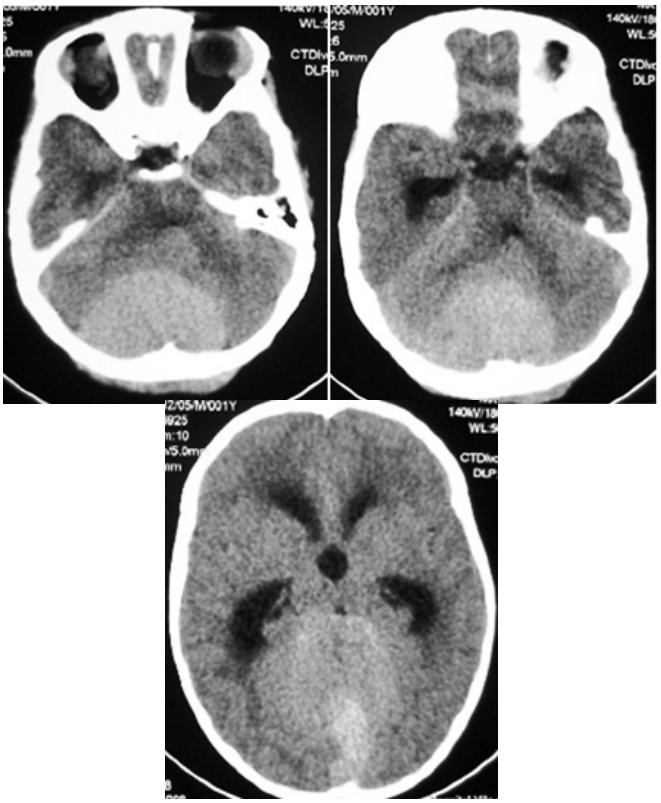

Figura 2: CT-scan without contrast showing an hyperdense infratentorial expansive mass and supratentorial hydrocephalus.

In a few hours he had symptoms progression (GCS-8) and required orotracheal intubation. Following that, he underwent emergency external ventricular shunting with intracranial pressure monitoring.

In Magnetic Resonance Imaging (MRI), the mass had an intense gadolinium uptake, showing significant compression over neural structures and fourth ventricle dislocation (Fig 3).

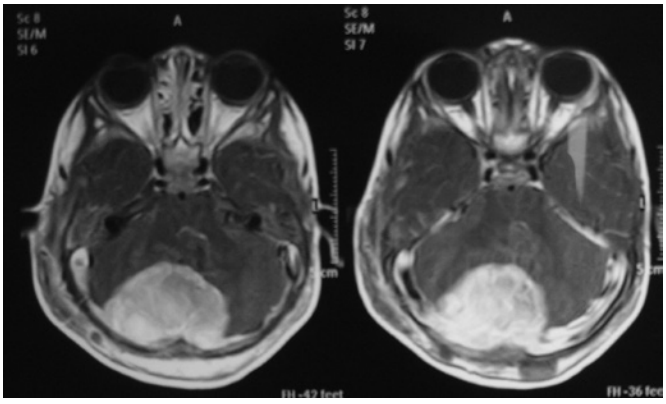

Figura 3: MRI showing intense contrast uptake and encephalic compression.

Because of the case severity, clinical instability and atypical mass appearance, we initially chose to perform a biopsy of the posterior neck lesions. Initial morphology suggested rhabdomyosarcoma, and the patient was scheduled to oncological treatment.

Concurrently, the child had a progressive raise in leucocytes, which reached a peak at the tenth day of hospital stay (134,000 leukocytes, 2% metamyelocytes, 1% myelocytes). After a myelogram and immunophenotyping, we found positive common leukocyte antigens and positive myeloperoxidase in 30% of the cells. He was diagnosed with type M5 AML.

Hystopathological examination cconfirmed the diagnosis.

We started chemotherapy with thioguanine (7 days), cytarabine and etoposide (4 days).

He had a quick leukocyte count drop (Table 1) and tumor volume shrink (Fig. 3).

However, despite this improvement, the child developed pancytopenia, meningitis by pseudomonas and subarachnoid hemorrhage, and he died at the 30th day of hospital stay by septic shock.

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Table 1. Complete Blood Count during evolution on Chemotherapy

	1st Day	10th Day	18th Day	28th Day
Leucometry (cel/mm³)	13,800	134,000	3,900	1,400
Hemoglobin (g/dl)	10.4.	8.7	8.1	8.0
Hematocrit (%)	30.2	23.4	25.2	21.9
Platelet count (cel/mm³)	208,000	119,000	14,000	10,000
PCR	2.0	2.0	71.3	262.5
		Chemotherapy		

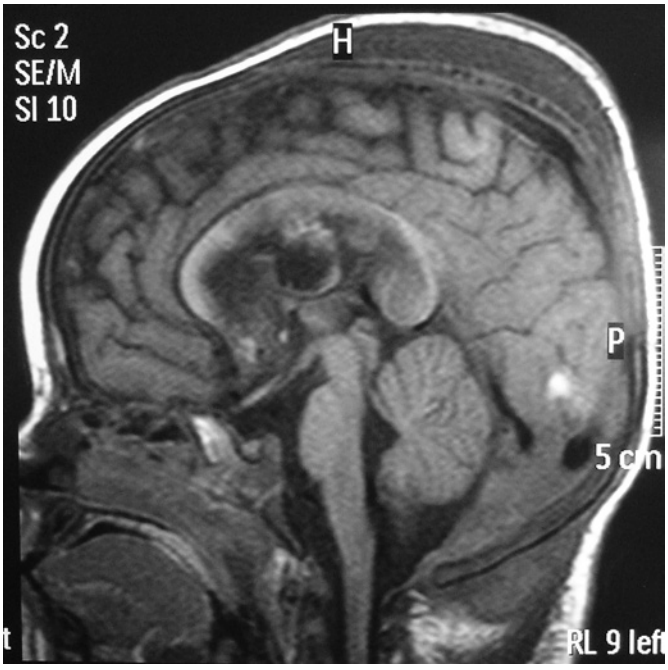

Figura 4: MRI shows an important mass involution after chemotherapy.

Discussion

The leucemia's etiology remains obscure. In its pathogenesis, there seems to be genetic and environmental mechanisms involved. Its clonal origin, stemming from the disorganized proliferation of a single progenitor cell is observed in most of the forms of human leukemia[2,11,25,26].

Among environmental risk factors associated with the development of leukemia, we list: exposure to radiation, use of topoisomerase II inhibitor drugs and the consumption of alcohol during pregnancy. Among genetic factors we especially stress: Down syndrome, Fanconi anemia, ataxia-telangiectasia, neurofibromatosis, Bloom syndrome, Noonan syndrome and first degree relatives of leukemic patients[3].

The AML most used classification system is the one developed by the FAB group (French-American-British classification), which classifies AML in nine distinctive subtypes according to the myeloid strain involved and the level of leukemia cells differentiation[4,27,32].

In his first description, Burns in 1811 reported a case of this tumor in the lachrymal gland of a young man[5]. The first to use the term Chloroma was King, in 1853[16].

In a review of 17 cases, Dock showed an association between leukemia and chloroma[10].

Since not all of these tumors have a green color, the term granulocytic sarcoma is the most indicated[15]. In its incidence, there does not seem to be any difference between genders[16,33].

Some authors report a mild predilection for the posterior fossa[12,17,22,28].

Neurologic complications of leukemia usually happen by direct invasion of the central nervous system (SNC) by leukemia cells or by changes in blood factors which cause blood excessive viscosity and cerebrovascular disease[18,29,30].

Intracranial granulocytic sarcoma is a rare condition. It is stated that the leukemia cells, from the bone marrow penetrate the periosteum, cross the skull through the Harver's canals and thus reach the dura matter, where they proliferate and form the granulocytic sarcoma[24]. Until very recently, it was believed that these tumors did not invade the brain parenchyma[19,20]. Current studies have shown that these leukemia granulocytic cells do invade the brain[23,31.]

As far as the clinical manifestation is concerned, there are reports of chloromas involving the intracranial compartment and they are radiologically very similar to meningiomas[29,30] and, also, lymphomas[31]. Chelly et al. described the intracerebellar form of the tumor in adults[6]. In an unprecedented case, reported by Obara et al., a 41-year old male developed a right-side granulocytic sarcoma after complete remission of his myeloid leukemia [23].

Granulocytic sarcomas are very severe manifestations, especially when involving the skull and the CNS. Colović et al. reiterate the major mortality associated with this neoplasia as they reported the case of an adult patient who died on the second day after surgery for a partial resection of a cerebral

granulocytic sarcoma[7].

Regardless of its radiosensitivity, intracranial granulocytic sarcomas have a bad prognosis because of its systemic disease severity and blastic transformations[13,17].

The initial diagnostic doubt, based on morphology is also reported in the literature. In a review of 61 granulocytic sarcoma cases, most were originally diagnosed as lymphomas[21].

The description of the present case aims at drawing attention to this rare pathology and its unusual presentation..

Conclusion

Granulocytic sarcomas are rare and can mimic other posterior fossa tumors when appear in cerebellum. Careful must be made for accomplish a correct diagnosis and treatment.

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