Case 16167

Orbital Granulocytic Sarcoma
(Chloroma)
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Section: Head & neck imaging
Area of Interest: Head and neck Neuroradiology brain
Procedure: Education
Imaging Technique: MR-Diffusion/Perfusion
Imaging Technique: MR
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 6 months, female

Clinical History:
Systemic neutropenia in study. Exophthalmos with displacement of the right eyeball in direction inferonasal of ten days of evolution. Suspected orbital mass right.

Imaging Findings:
A solid mass is identified in the upper outer of the right orbit, extraconal location, well defined edges and approximate dimensions of 21 x 9 x 30 mm, extending anteroposteriorly to the vertex of the orbit. It conditions the displacement of the orbital structures in the inferointernal and anterior sense, observing secondary exophthalmos. The mass is homogeneously isointense in T1 and hypointense in T2, with a marked restriction of diffusion (ADC of 0.4), which indicates high cellularity of the lesion. After administration of intravenous contrast, there is a homogenous enhancement of the mass.

Discussion:
Granulocytic sarcoma is a rare solid neoplasm derived from precursor cells of the granulocytic series. It was first described by Burns in 1811. It was later named Cloroma by King 1853 due to its high level of myeloperoxidases that make it look green. Later it was called Sarcoma Granulocitico (Rappaport, 1966).[1, 2]

Clinical Perspective: Leukemia is the most frequent malignancy in pediatric age. Mainly patients with myelogenic precursor leukemias develop solid tumors (Granulocytic Sarcoma). They can be localized in any part of the body, however they occur more frequently in the orbit and subcutaneous tissues. Clinically the proctosis is the most common findings in the orbit, followed by cellulitis and mass in the lacrimal gland. The great majority occur in children before 15 years of age, with a peak of age between 5 and 6 years, without presenting gender predilection(M: F = 1: 1). It can occur in association with: acute myeloid leukemia (AML), chronic myeloid leukemia (CML), myelofibrosis with myeloid metaplasia, hypereosinophilic syndrome and polycythemia vera (5-10% of patients with AML, 1-2% with CML). They can develop during a haematological disease or may precede it for
months or years. Most are asymptomatic and when it occurs in patients without known hematologic disease, it is a sign of poor prognosis. The diagnosis of granulocytic sarcoma is highly suspicious in a patient with a history of leukemia and rapid progression proptosis. It is important to emphasize that it can be anatomopathologically and radiologically indistinguishable from lymphoma[1, 2, 3]

Imaging Perspective: Findings in the CT. It presents as a solid tumor, homogeneous slightly hyperintense, with slight homogenous enhancement after the administration of intravenous contrast. With possible extension towards intra- and extracranal fat, the extraocular musculature and the eyeball.
Magnetic Resonance Findings: In T1-weighted images, they are isointense or hypointense and in T2-weighted sequences they are isointense or hyperintense with homogeneous enhancement after intravenous contrast administration. We can see restriction in DWI/ADC. [1, 2, 3, 4]

Treatment; It is important to diagnose it by responding better to focal radiotherapy than to chemotherapy. However, early and aggressive chemotherapy offers the best possibility, although the final prognosis is still bad.[1, 2, 3, 4]

"Written informed patient consent for publication has been obtained"


Final Diagnosis: Granulocytic Sarcoma (Chloroma)

References:

Description: In the right orbit a homogenous tumor is seen. Isointense in T1-weighted sequences (1a) and hypointense in T2-weighted sequences (1b), with homogenous enhancement after administration of intravenous contrast (1c,d,e). Origin: Department of Radiology, Hospital Universitario Rio Hortega, Valladolid, Spain.
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**Description:** The tumor presents a marked restriction in the diffusion sequences. hyperintense on DWI, hypointense on ADC (0.4). **Origin:** Department of Radiology, Hospital Universitario Rio Hortega, Valladolid, Spain.
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