

IMAGES IN PAEDIATRICS

Differential diagnosis of periorbital edema in sickle disease

Diagnóstico diferencial de edema periorbitario en drepanocitosis

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We present the case of an eight-year-old patient with sickle cell disease homozygous for HbS (with no family history) managed with prophylactic amoxicillin, hydroxyurea and folic acid, under annual follow-up, with a previous admis-



Figure 1 Periocular edema in the patient.

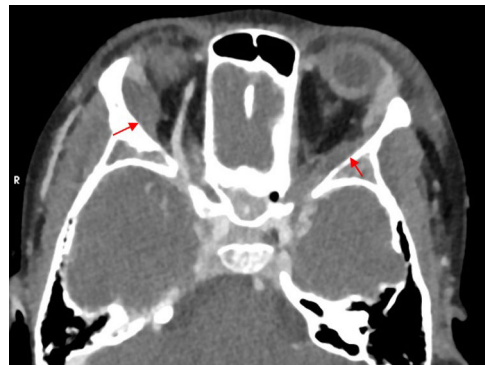


Figure 2 Axial orbital CT image showing the reported findings, marked by red arrows.

sion due to a vaso-occlusive crisis in the leg and no other relevant history. She was admitted to the sending hospital with suspected vaso-occlusive crisis in the extremities and abdominal pain and transferred to our hospital due to progressive deterioration with fever, hepatomegaly and splenomegaly extending to the iliac crest, in addition to painful bilateral periorbital edema in absence of phlogosis or changes in vision (Fig. 1). The bloodwork revealed a decreased hemoglobin concentration and platelet count and elevated hemolysis markers, with a proportion of sickle hemoglobin (HbS) of 44.3% and a C-reactive protein level of 17.7 mg/dL. Splenic sequestration and orbital cellulitis were suspected,^{1,2} leading to administration of supplemental oxy-

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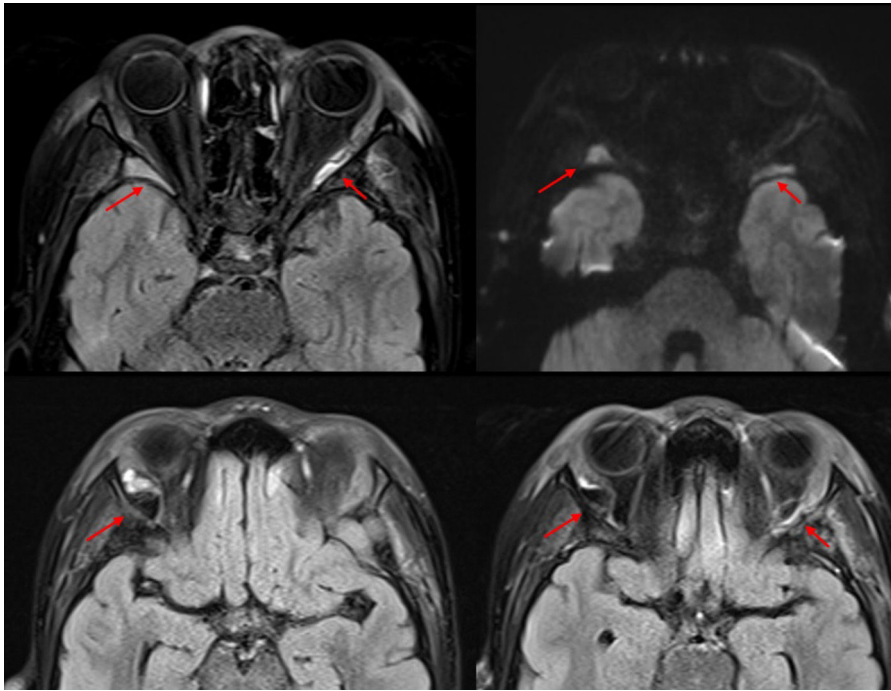


Figure 3 Multiple axial MRI images of the brain showing the reported findings, marked by red arrows.

gen, analgesia with continuous infusion of morphine, two red blood cell transfusions and intravenous antibiotherapy. The orbital CT scan (Fig. 2) showed bilateral involvement of soft tissues in the outer periorbital region, bordering the orbital rim and extending posteriorly, a finding that could be compatible with extramedullary hematopoiesis or a lymphoproliferative disorder. The head MRI (Fig. 3) ruled out nervous system involvement and showed areas suggestive of infarction in the great wings of the sphenoidal bones with secondary hemorrhagic effusion.³ The blood culture and the antibody test for Epstein-Barr virus were negative. A diagnosis of bone infarction was made and antibiotherapy discontinued, with full clinical resolution in seven days.

References

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