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Case Report

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Orbital complications in sickle cell disease: a case report presentation

Abstract

Our report describes a child with sickle-cell anemia presenting with left-sidedperiorbital swelling, headache, elevated leukocytes and inflammatory markers, discovered to have orbital bone infarction. Initial treatment included antibiotics for presumed infection. As MRI orbit suggested infarction of the orbital bone, exchange transfusion and steroid course was administered with subsequent improvement of symptoms. This case demonstrates that although infectionisa more common etiology of periorbital edema in sickle-cell disease, maintaining the differential of orbital bone infarction is crucial as patient out comes depend on prompt intervention. Caution is advised when treating these patients with steroids given risk of rebound pain.

Keywords: pediatrics, hematology, sickle cell anemia, orbital complication, vaso occlusive crisis, orbital bone infarction

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Abbreviation: HGB S, hemoglobin S; IV, intravenous; NPO, nil per os/nothing by mouth; SCD, Sickle-Cell Disease, VOC, vaso occlusive crisis

Introduction

Periorbital eye swelling is often presumed to be of infectious etiology and in the majority of cases this is an accurate assumption. However, given the potential for life-threatening consequences associated with orbital manifestations in sickle-cell disease, all possibilities should be considered at time of presentation. Ocular complications in sickle-cell disease can include retinopathies, ischemia of the anterior segment, and glaucoma.[1] Vasoocclusive crises (VOCs) involving the orbital bone can present similarly to infectious processes with periorbital edema, proptosis, eye pain and restricted eye movement. Thus, VOCs such as infarction of the orbital bone should always be regarded given high risk for compressive optic neuropathies and vision loss if not addressed in a timely manner.[2–5]

Results case presentation

A two year old African American male with Sickle Cell Disease (SCD), SS phenotype on prophylactic penicillin with good compliance, presented with a 2day history of rapidly worsening unilateral (left) orbital swelling with severe left-sided head and eye pain. He was diagnosed with sickle-cell disease as a newborn and had a prior history of Salmonella osteomyelitis with a similar orbital presentation at 1 year of age. At time of presentation he was afebrile with mild symptoms of congestion. After initial care was established, laboratory work up was sent (Complete blood count, complete metabolic panel, hemoglobin electrophoresis, and inflammatory marker) with results displayed. A blood culture was obtained and respiratory viral panel obtained was negative. He was started on a treatment plan consisting of intravenous (IV) hydration, broad spectrum antibiotics, systemic anti-inflammatory agents and opioid analgesic support per institutional standard guidelines for sickle cell pain crises. Imaging of the orbit was ordered showing mild peri-orbital soft tissue swelling with no evidence of abscess.

Due to persistence of severe paindespite analgesics (hospital day 2), imaging was re-reviewed with Radiology, revealing a high suspicion of orbital bone infarct. The patient was immediately made NPO and central-line was placed in preparation for exchange transfusion. He was given a dose of IV Solucortef pre-procedure and underwent erythrocytapheresis with goal of reduction of Hemoglobin S to <30%. After exchange transfusion the patient was transitioned from IV methylprednisoloneto oral prednisolone to complete a five day course of steroid therapy with the intent of reduction in inflammation while pain control was optimized. Additionally an analgesic regimen was prescribed to prevent rebound pain after steroid discontinuation.

The patient was monitored for an additional twenty-four hours and was noted to have significant improvement in periorbital edema and was without signs of discomfort/pain. Infectious work up remained negative throughout the hospitalization and the patient showed dramatic improvement in symptoms. The patient was thus discharged home and at follow-up examination eight days later was noted to have complete resolution of eyes welling and pain. No rebound pain from steroid discontinuation was noted.

Discussion

The pathophysiology of sickle-cell disease stems from the substitution of glutamic acid with valine at the sixth position of the hemoglobin chain and results in creation of a variant hemoglobin molecule (Hemoglobin S). Polymerization of the hemoglobin S molecule during transient changes in oxygen affinity leads to formation of sickled redblood cells that have potential to cause pain crises and vaso-occlusive events.[1,2] It is well known that the locations of vaso-occlusive crises in sickle-cell disease typically involve the long bones given the increased supply of bone marrow. Facial bones such as the orbital bones are believed to be less frequently affected by sickle-cell crisis given presence of decreased marrow space, but in younger patients there is still potential for involvement of facial bones given slightly increased marrow space and marrow activity at these locations when compared to older children[2,3,4,5].The classic presentation of orbital infarction can appear similar to orbital infectious etiologies (pre-septal and orbital cellulitis, osteomyelits) with progressive peri-orbital pain, swelling and proptosis[1, 4, 5] as was the presentation in our 2 year old patient. The inflammatory response triggered by orbital bone infarction due to distinct pathology from infection, requires immediate evaluation and intervention to prevent detrimental effects such as opthalmoplegia, orbital compression syndrome and vision loss.[6] One should be mindful of the entity of rebound pain after a course of high dose systemic steroid use in sickle cell patients while

using such agents for unusual presentations such as the one reported above.[7,8] Complications in SCD such as the one described in this case report presentation serve as an important reminder that when sickle-cell patients present with eye-swelling, the differential diagnosis should be broad irrespective of previous presentations, including both infectious and non-infectious causes such as bone infarction. Moreover, urgent intervention is required when serious pathologies are discovered in order to achieve the best outcomes for our patients who suffer from sickle-cell disease and vaso-occlusive events.

Conclusion

This case highlights the importance of identifying unique orbital complications in SCD and the appropriate clinical strategy to manage such patients with special attention to a proactive approach and judicious use of systemic steroids. Young patients with severe sickle cell phenotype can have unusual presentations of the often complicated pathophysiology of red cell sickling. Early loss of splenic function, incomplete immunization series and ongoing end organ hypoxemia put such patients at a high risk for infectious complications. When managing orbital disease in SCD, unique presentations such as orbital infarcts should always be carefully screened for, as timely intervention can prevent overuse of systemic broad spectrum antibiotic usage and potentially salvage ocular function.

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Conflicts of interest

For Dr. Akshat Jain: Advisory board for Shire, Bayer pharmaceuticals.

For Dr. Sara Jane Onyeama: None.

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