Journal of Medical Imaging & Case Reports

https://doi.org/10.17756/micr.2020-052

Clinical Image

Open Access

"Hair-on-End" Sign on a Magnetic Resonance Imaging of the Head in a Sickle Cell Disease Patient – A Clinical Image

Lama Alomari[1] and Ayman Khushaim[2*]

[1]Emergency Medicine Department, Security Forces Hospital, Riyadh, Saudi Arabia
[2]Pediatric Radiology Department, McGill University Health Center, Montreal, Canada

***Correspondence to:**
Dr. Ayman Khushaim, MD
Pediatric Neuroradiology Fellow Pediatric Radiology Department McGill University Health Center, 1001 Decarie Blvd.
Room B02.7006 Montreal
QC, H4A 3J1, Canada
Tel: +1-514-430-8754
E-mail: ayman.khushaim@mail.mcgill.ca

Received: May 14, 2020
Accepted: July 21, 2020
Published: July 22, 2020

Citation: Alomari L, Khushaim A. 2020. "Hair-on-End" Sign on a Magnetic Resonance Imaging of the Head in a Sickle Cell Disease Patient – A Clinical Image. *J Med Imaging Case Rep* 4(2): 57-58.

Published by United Scientific Group

Abstract

An 11-year-old boy known to have sickle cell disease presented to his routine follow up with a history of new frontal headache and difficulties with his school performance. Magnetic resonance imaging of the brain depicted a hair-on-end sign, a hallmark of excessive erythropoiesis caused by severe anemia.

Keywords

"Hair-on-end" sign; Sickle cell disease; Bone marrow expansion; Periosteal pain; Magnetic resonance imaging (MRI)

List of Abbreviations

SCD: Sickle Cell Disease; MRI: Magnetic Resonance Imaging; CT: Computed Tomography

Case

An 11-year-old boy who is known to have sickle cell disease (SCD) type HbSS, which is considered the most severe type, presented to his hematologist's office for a routine follow-up. During the interview, the mother mentioned that her son had some difficulties with his school performance; in addition to an occasional frontal headache that he had never complained of before. Consequently, his physician requested a magnetic resonance imaging (MRI) of the head to rule out silent infarcts. The study was negative for silent infarcts.

However, it demonstrated marked calvarial and clival bone marrow expansion with diffuse intermediate signal intensity in keeping with red bone marrow reconversion related to sickle cell disease. There was also widening of the diplopic space with multiple linear striations seen across it giving the hair-on-end appearance (Figure 1). Additionally, axial T1-weighted and T2-weighted images demonstrated diffuse opacification and expansion of the maxillary sinuses bilaterally with diffuse intermediate signal intensity on T1/T2 and areas of peripheral high signal intensity indicative of fat component, findings are in keeping with extramedullary hematopoiesis related to SCD (Figure 2).

The hair-on-end sign of the skull is a unique trait attributed to chronic hemolysis associated with severe congenital anemias such as thalassemia and sickle cell anemia [1, 2]. This sign has been classically observed and reported on plain radiography; however, it can also be identified demonstrating the same appearance on computed tomography (CT) and MRI [3]. The literature has not

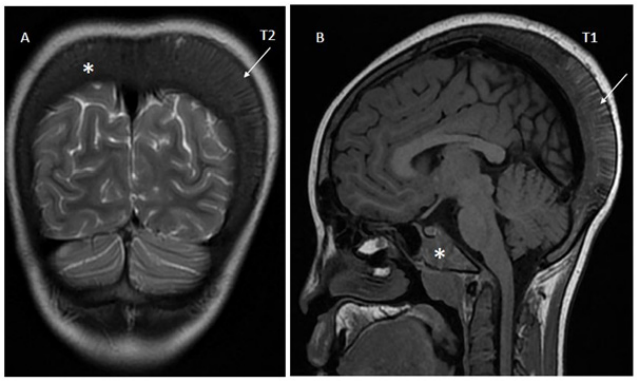

Figure 1: Coronal T2-weighted and sagittal T1-weighted images (A & B) demonstrate marked calvarial and clival bone marrow expansion with diffuse intermediate signal intensity in keeping with red bone marrow reconversion related to sickle cell disease (asterix). There is widening of the diplopic space with multiple linear striations seen across it giving the hair-on-end appearance (arrows).

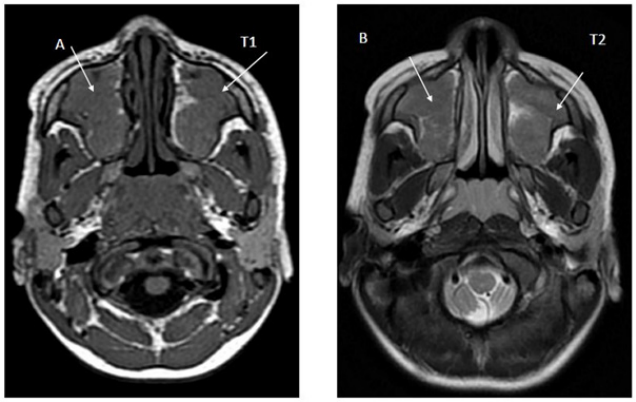

Figure 2: Axial T1-weighted and T2-weighted images (A & B) demonstrate diffuse opacification and expansion of bilateral maxillary sinuses with diffuse intermediate signal intensity on T1/T2 and areas of peripheral high signal intensity indicative of fat component, findings are in keeping with extramedullary hematopoiesis related to sickle cell disease (arrows).

described the sensitivity and specificity of different modalities for the hair-on-end sign.

The appearance is caused by excessive bone marrow proliferation in response to severe anemia causing red marrow expansion that eventually leads to widening of the diploic space, thinning of the outer table of the skull and coarse vertical trabeculation [1, 3, 4]. These vertical trabeculae are recognized on skull radiographs as long, thin vertical striations that give the distinguished appearance of hairs standing on end [4]. Patients who have this distinct radiological feature may complain of headaches that can be attributed to periosteal pain due to marrow expansion, in the absence of other radiological abnormalities, and within the context of normal neurological and ophthalmic examination [1]. In this case, given the unremarkable work up, the patient was given analgesia and was advised to maintain his regular follow up.

Disclosure statement

The authors have no conflict of interest or acknowledgements.

References

1. Albert MH, Notheis G, Wintergerst U, Born C, Schneider K. 2007. "Hair-on-end" skull induced by long-term G CSF treatment in severe congenital neutropenia. *Pediatr Radiol* 37(2): 221-224. https://doi.org/10.1007/s00247-006-0363-3
2. Amadi OP, Patel C, Tangella K, Khaliq W. 2012. 'Hair-on-end' sign: an important finding in clinical practice. *QJM* 105(9): 905-906. https://doi.org/10.1093/qjmed/hcr151
3. Martin L, Rackard F. 2016. Images in clinical medicine Hair-on-End Sign. *N Engl J Med* 374(19): e23. https://doi.org/10.1056/nejmicm1510045
4. Basu S, Kumar A. 2009. Hair-on-end appearance in radiograph of skull and facial bones in a case of beta thalassaemia. *Br J Haematol* 144(6): 807. https://doi.org/10.1111/j.1365-2141.2008.07404.x