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International Journal of Clinical Biochemistry and Research

Journal homepage: https://www.ijcbr.in/

Letter to Editor

Septic arthritis in sickle cell anemia

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ARTICLE INFO

Article history:
Received 14-04-2023
Accepted 22-04-2023
Available online 25-10-2023

1. Letter

It is estimated that the global rate of birth with homozygous children with sickle cell anemia (SCA) is around 112 per 100000 and this figure rises to 1125 per 100000.[1] The hotspots of SCA prevalence are sub-Saharan and North-East Africa, India and the Middle-East region. The mortality rate are much higher in Africa and India due mainly to low-resource concerns.[2] Over 75% of the SCA-born children often do not attain their 5^{th} birthday.[3] Therefore, SCA is considered the most common, fatal hemoglobinopathy.

2. Biochemical Basis of SCA

SCA is a genetic mutation that substitutes valine for glutamate in the 6^{th} position of β-chain of HbA. This genetic mutation is accompanied by phenotype transformation from HbA to HbS (A for Adult while F for sickled). As a result of the missing repulsing negative charge imposed by glutamate, the valine forms hydrophobic interactions with the other chains, clustering and stacking with one another forming the crescent shape seen in sickled red blood cells (RBC).[4] However, the deleterious consequences taken place within RBC are extended into the general circulation. That is, impaired biorheology of sickled RBCs, distributed vaso-occlusion, hemolysis and inflammation ensues[5] as illustrated in Figure 1.

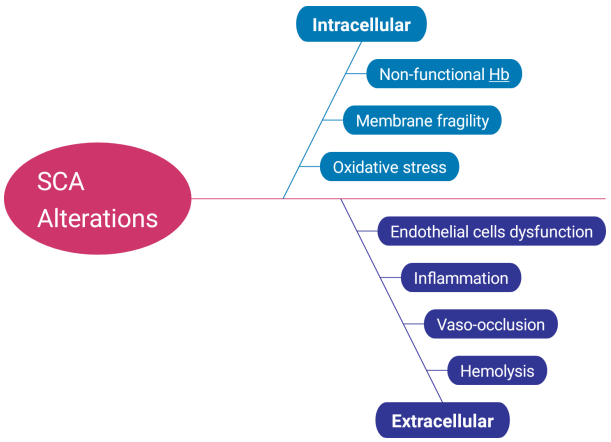

Fig. 1: Intracellular and extracellular alterations present in patients with SCA

3. SCA and Septic Arthritis

Patients with SCA develops fast progression of significant complications, more notably painful vaso-occlusive crisis (VOC) affecting joints, bones, spleen, kidneys and brain leading to end-organ failure.[6] Septic arthritis is also documented in SCA patients albeit rare. Approximately 3% of all SCA patients experience septic arthritis with frequent pain, swelling, fever, leukocyte count above 15000/mm^3 and

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https://doi.org/10.18231/j.ijcbr.2023.046

c-reactive protein exceeds 20 mg/L. The causative pathogen is predominantly *Staphylococcus aureus* where the aspirated synovial fluid culture is positive in > 96% of cases.[7] The disease affects primarily long bones, i.e. the hip (femoral head) and shoulder (humeral head) although vertebrae and ribs can be effected interfering with the respiratory functions of the lung.[8] The implicated pathogenesis can be interpreted as follows: the inflammation induced by VOC represent a promoting environment for bacterial pathogens growth. Furthermore, VOC damages the spleen, an essential immune organ that in normal circumstances simultaneously filters RBC and engulf pathogens.[9] After confirming the diagnosis of septic arthritis, empirical management should be introduced as soon as possible. The treatment-of-choice depends on the antibiotics sensitivity findings. In other words, Ceftriaxone 50-75 mg/kg up to 2 g/dose is prescribed for Cephalosporin-sensitive culture whereas Clindamycin 10-15 mg/kg/dose for Cephalosporin-resistant ones. Notably, Vancomycin 15 mg/kg/dose is indicated if there is systemic sepsis or Clindamycin-side reactions experienced.[10]

4. Conclusion

The autosomal recessive disease SCA is caused by a single substitution (point mutation) leading to catastrophic consequences within as well as outside the RBC. The painful arhthritis caused by VOC should be distinguished from septic arthritis caused by *S. aureus*. As soon as the diagnosis has been confirmed with synovial fluid culture, either Ceftriaxone or Clindamycin should be introduced to deal with the burden sepsis.

5. Source of Funding

None.

6. Conflict of Interest

None.

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Cite this article: AlMadhagi HA, Tarabishy AA. Septic arthritis in sickle cell anemia. *Int J Clin Biochem Res* 2023;10(3):258-259.