CASE REPORT

Sickle Cell Disease Revealed by Soft Tissue Abscess: One Case Report

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Abstract:

Introduction:

Sickle cell disease is a haemoglobinopathy characterized by the occurrence of vaso-occlusive crises and osteoarticular complications.

Case-Report:

We report the case of an infant with sickle cell disease revealed by a bilateral abscess of the feet. Our patient is an 18-month-old infant who has had bilateral swelling of the feet for a week with fever of 40 °C, a CRP of 129 mg/l and a leukocytosis of 32,000 elements/mm³ together with normochromic normocytic anemia at 7.9 g/dl.

The diagnosis of abscess was taken and a puncture was made finding a purulent fluid with isolation of Salmonella. In front of the bilateral character, Salmonella isolation and normochromic normocytic anemia, electrophoresis of hemoglobin was requested confirming the diagnosis of sickle cell disease. The progress was positive with hydration and antibiotic therapy.

Conclusion:

Soft-tissue Salmonella infections must lead to thinking of sickle cell disease as a diagnosis especially with normochromic normocytic anemia combined.

Keywords: Sickle cell disease, Salmonella abscess, Normochromic normocytic anemia, Hemoglobinopathy, Osteoarticular, Electrophoresis.

1. INTRODUCTION

Sickle cell disease is an inherited autosomal recessive disorder of hemoglobin, linked to a mutation in the sixth codon of the β chain of hemoglobin with the replacement of glutamic acid by valine acid to give as a result, hemoglobin S [1]. Its frequency is high in the Mediterranean basin because of the frequency of consanguinity. Sickle cell children have many and varied complications related to the disease. Multiple abscesses of soft tissue are considered as a rare complication.

In this work, we illustrate the possibility of the accidental discovery of sickle cell disease following multiple abscesses of soft tissues and, through this case report, a brief review of the literature.

2. CASE REPORT

This is an 18-month-old infant, an only child of his parents, from a non-consanguineous marriage, well vaccinated according to the National Immunization Program (NIP), with no particular pathological history. Five days before admission, he had a hot and painful swelling on his 2 feet combined with an acute fever of 39 - 40 ° with good general status.

History taking found several episodes of acute fever, treated as an outpatient.

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On physical examination, there was a fever at 39 °C, cutaneous-mucous pallor, thrush, inflammatory edema with short pit recovery time (PRT) on the back of the 2 feet and facing the right ankle, the ankle joints were free but their active mobilization was painful.

Ankle ultrasound was indicated to take out septic arthritis, and no intra-articular effusion was found. The x-ray of both feet showed no bone involvement. Blood tests revealed predominantly neutrophilic leukocytosis at 32,000/mm$^3$ (PNN at 19,380/mm$^3$), CRP at 129 mg/l, normochromic normocytic anemia 7.9 g/dl, and sterile blood cultures.

On the second day of hospitalization, there was a changeable collection on the back of the right foot and soft-tissue ultrasound mentioned to bilateral superficial collections of the feet together with extensive edematous infiltration of the surrounding area.

An ultrasound-guided puncture was indicated and the bacteriological study of pus punctured isolated *Salmonella Typhi* susceptible to amoxicillin-clavulanic acid. In front of normochromic normocytic anemia and *Salmonella* abscess, electrophoresis of hemoglobin was performed, showing hemoglobin S at 69.4% and hemoglobin A at 2.6%. The diagnosis of sickle cell disease was therefore made.

The patient was treated with analgesic, antibiotic, blood transfusion and hyper-hydration, as recommended by the literature.

The progress was satisfactory after fifteen days of antimicrobial treatment with apyrexia and a good healing of soft tissue.

In regular outpatient follow-up, ten months later, he had never presented any other complications of sickle cell disease.

3. DISCUSSION

Sickle cell disease is an autosomal recessive, frequent and severe hemoglobinopathy related to an abnormal hemoglobin structure that results in the formation of hemoglobin S (HbS). The clinical expression of sickle cell disease is wide with many varied symptoms. This variability mainly reflects genetic and environmental influences [2].

The musculoskeletal system is the one most affected by the vaso-occlusive crisis in sickle cell patients: 80% of the affected patients suffer from it [3]. The dactylitis, described in infants between 6 months and 2 years of age, is the earliest form of involvement. It is manifested by pain combined with extensive swelling and inflammatory swelling of several metacarpals or metatarsals.

Infection is also a common complication of sickle cell disease, especially in infants and toddlers, where it can be life-threatening, with five-year mortality reaching 25-30%. It is the infection which is involved in the majority of cases [4]. These infections are almost always osteomyelitis [3]. Septic arthritis is rare, reactional effusions are more frequent. They occur very often in bone segments, the microcirculation of which has been damaged by multiple vaso-occlusive crises [3].

Infections can rarely affect soft tissues (muscle and skin), without bone involvement [5], as was the case of our patient, which makes the diagnosis of sickle cell disease difficult, leading to a delay in treatment and therefore the progress of complications including acute osteomyelitis. The dissemination is made via hematogenous way during a bacteremia [5]. *Staphylococcus aureus* is the most incriminated bacteria. *Salmonella* is not a common germ in soft-tissue infections and therefore the search for the underlying cause, including sickle cell disease, must be systematic [2].

In our patient, the isolation of salmonellosis in the punctured fluid allowed us to make the diagnosis of sickle cell disease. Treatment includes antibiotic therapy and surgical or percutaneous drainage as appropriate. There is no consensus for the duration of antibiotic treatment and this should be adapted to clinical improvement as well as biological and radiological data [5].

Studies show that three to four weeks of parenteral therapy are usually appropriate, but in patients with more complicated infections, a longer duration may be required [4].

CONCLUSION

Sickle cell disease is a disease that has various clinical symptoms. It can be revealed by superficial abscesses without bone involvement. Soft-tissue *Salmonella* infections should make us suspect sickle cell disease especially
combined with normochromic normocytic anemia.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Not applicable.

HUMAN AND ANIMAL RIGHTS

No animals/humans were used for studies that are the basis of this report.

CONSENT FOR PUBLICATION

A written informed consent was obtained from the parents.

CONFLICT OF INTEREST

The authors declare no conflict of interest, financial or otherwise.

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Declared none.

REFERENCES


