Case Report

RHEUMATOID ARTHRITIS IN A SICKLE CELL PATIENT: A DANGEROUS ASSOCIATION?

Fatoumata Diakité[1*], Youssouf Traoré[2], Boubacari Ali Touré[2], Boureima Kodio[3], Mohomedine Touré [3], Sidi Touré[3], Fanta Sangaré[3], Seydou Diallo[3], Ibrahima Sory Pamanta[3], Idrissa Ahmadou Cissé[3]

Author information: [1]Department of Medecine, CHU Gabriel Touré, Bamako, Mali, [2]Sickle Cell Disease Research Center, Point G, Bamako, Mali, [3]Rheumatology Department, CHU du Point G, Bamako, Mali.

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Abstract: Introduction: The diagnosis of rheumatoid arthritis remains a challenge because sickle cell disease can result in various rheumatological manifestations, including joint and bone pain. The simultaneous presence of rheumatoid arthritis and sickle cell disease makes the therapeutic management of both conditions problematic.

Observation: A 24-year-old man, a nurse by profession, has been followed for ten years for homozygous sickle cell disease at the Sickle Cell Disease Research Center (CRLD) of Bamako, Mali. He has presented for eight months symmetrical polyarthritis with morning stiffness of 3 hours, distinct from the usual vaso-occlusive crisis. The Analog Visual Scale was estimated at 80/100. He reported unquantified weight loss and asthenia. The physical examination showed a deformity in bilateral ulnar deviation, flexion of the right elbow, twelve painful joints, and five swollen joints. Normochromic normocytic anemia (hemoglobin 8.3g/dl), inflammatory syndrome with C - Reactive Protein (CRP) 130.91 mg, and Sedimentation Rate (ESR) 72mm at the first hour were noted. Rheumatoid Factor was weakly positive at 21.3 IU and Anti Citrullinated Peptide Antibodies at 385.2 IU. The radiography discovered bilateral erosive carpitis without associated tarsitis and osteonecrosis of both femoral heads. The diagnosis of a very active immunopositive erosive rheumatoid arthritis meeting the criteria of ACR / EULAR 2010 was retained. A treatment based on prednisone 10 mg per day was initiated, associated with methotrexate at a dosage of 15 mg weekly in a single dose, folic acid, calcium, and vitamin D.

Conclusion: The coexistence of rheumatoid arthritis in sickle cell patients makes the diagnosis of polyarthritis difficult.

Keywords: Rheumatoid Arthritis; Sickle Cell Disease; Polyarthritis, Vaso-occlusive crisis.

INTRODUCTION Rheumatoid arthritis is the most common chronic inflammatory rheumatologic disorder. It is a complex, multifactorial condition characterized by rapid joint destruction [1]. Rheumatoid arthritis can be associated with hemoglobinopathies, including sickle cell anemia. Sickle cell disease is a genetic disorder transmitted in a recessive mendelian mode. Early diagnosis of rheumatoid arthritis in a sickle cell patient would prevent highly disabling functional complications and visceral damage that can be life-threatening. However, the diagnosis of rheumatoid arthritis remains a challenge because sickle cell disease can result in various rheumatological manifestations, including joint and bone pain. These manifestations can cause the joint damage of rheumatoid arthritis. The simultaneous presence of rheumatoid arthritis and sickle cell disease makes the therapeutic management of both conditions problematic [2]. We report a case of sickle cell disease and rheumatoid arthritis treated with prednisone and methotrexate and discuss diagnostic and therapeutic difficulties.

Corresponding Author: Fatoumata. Diakité, MD,
CHU Gabriel Touré BP 267 Bamako, Mali.
Email: fatoumatamli@yahoo.fr.

CASE PRESENTATION

A 24-year-old man, a nurse by profession, has been followed for ten years for homozygous sickle cell disease at the Sickle Cell Disease Research Center (CRLD). The patient has presented for eight months symmetrical polyarthritis of the hands, wrists, elbows, shoulders, ankles, and feet with morning stiffness of 3 hours, distinct from the usual vaso-occlusive crisis. Axially, he indicates

fluctuating neck pain and low back pain without irradiation. The Analog Visual Scale was estimated at 80/100. In addition, he manifested an unquantified weight loss and asthenia. However, the questioning did not report any visceral sign. The physical examination showed a deformity in bilateral ulnar deviation, flexion of the right elbow, twelve painful joints, and five swollen joints.

The cardiopulmonary, abdominal, and mucocutaneous examinations were all normal. The patient weighed 47 kg with a height of 161 cm. Furthermore, normochromic normocytic anemia (hemoglobin 8.3g/dl), inflammatory syndrome with C-Reactive Protein (CRP) 130.91 mg, and Sedimentation Rate (ESR) 72mm at the first hour were noted. The Rheumatoid Factor (RF) was weakly positive at 21.3 IU (Standard <14 IU) and Anti Citrullinated Peptide Antibodies (ACPA) at 385.2 IU (Standard <17 IU). The radiography revealed bilateral erosive carpitis without an association with tarsitis and osteonecrosis of both femoral heads. The cardiac ultrasound revealed a tricuspid leak. A diagnosis of a very active immunopositive erosive rheumatoid arthritis (DAS 28 CRP = 6.40) meeting the criteria of ACR / EULAR 2010 was concluded.

A treatment based on prednisone 10 mg per day was initiated, associated with methotrexate at a dosage of 15 mg weekly in a single dose, folic acid, calcium, and vitamin D. On the eleventh day of treatment, the patient reported low back pain without irradiation, which was treated by the CRLD with analgesics. Two days later, dental pain and temporomandibular joints appeared with the inability to open the mouth and chew, which prompted a consultation at the Odontology and Stomatology University Center. Hence a diagnosis of dental caries was observed, and a broad-spectrum antibiotic therapy was prescribed based on amoxicillin, clavulanic acid, metronidazole, and decayed tooth extraction was performed. The evaluation of the disease at the sixth week of treatment was marked by the improvement in clinical symptoms. The Visual Analogue Scale (VAS) was estimated at 30/100, morning stiffness at 1 hour, the presence of two pain joints: right wrist and the second metacarpophalangeal, and synovitis of the right wrist, regression of the inflammatory syndrome (CRP: 13.22 mg). The disease activity was moderate, with a CRP SAR of 3.40. The hemoglobin level was 10.1 g/dl. In view of these observations, a new posology was proposed by increasing the dose of methotrexate to 20 mg per week and gradually reduced the dose of prednisone to 5 mg per day in one week. At the twelfth week, the VAS and morning stiffness remained stable, but the painful joint count became three (right wrist and both elbows) with persistent synovitis of the right wrist. Biology monitoring was not performed for lack of funds.

DISCUSSION A case of rheumatoid arthritis in a homozygous sickle cell patient was observed. The coexistence of rheumatoid arthritis and sickle cell disease remains rare. However, cases of the association have been reported in the literature [3, 4]. The patient presented an erosive seropositive rheumatoid arthritis meeting the ACR / EULAR 2010 criteria in this present case. The combination of rheumatoid arthritis and sickle cell disease poses a diagnostic and therapeutic problem. These two pathologies can have similar clinical presentations. The patient's age is superimposable to those reported by some authors [3]; however, he is significantly younger compared to the patients in the McFarlane series [5]. The diagnostic delay was eight months, which is lower than that reported in the literature [2, 5]. Also, the patient presents peripheral and axial topography arthralgias. Sickle cell disease is characterized by sudden intense pain affecting the bones (long bones, pelvis, spine, ribs) and peripheral joints, which may be migratory [6]. A contributing factor is the most found [7, 8]. The presence of inflammatory arthralgia with synovitis, which is characteristic of rheumatoid arthritis, supported the diagnosis. The clinical picture was confirmed by synovitis in the patient and gusty ulnar deformity, which are pathognomonic signs of RA [9]. This unusual clinical spectrum in the sickle cell patient pointed to rheumatoid arthritis.

Axial involvement is usually not described in rheumatoid arthritis except in cases of arthritis of the cervico-occipital hinge or atloid-axoid dislocation [6]. Polyarthralgia, synovitis, and deformities have been reported in the literature; however, axial involvement has not been described [3, 4]. Biologically, an important inflammatory syndrome was noted, and rheumatoid arthritis observed is strongly positive for ACPA and weakly positive for RF. Zomalhèto reported a positivity of 90.5% and 80.9%, respectively, for RF and ACPA. The radiological aspect highlighted an erosion present in 85.7% in his series [3]. If corticosteroid therapy is indicated in rheumatoid arthritis, however, it is contraindicated in sickle cell disease because it can cause a vaso-occlusive crisis [10, 11]. We were confronted with diagnostic difficulties with the presence of sickle cell anemia and its consequences, which are osteonecrosis of the femoral head and anemia in our patient. The short-term course was marked by improvement in disease activity, but it was interspersed with infection. It is recognized that corticosteroid therapy and sickle cell anemia increase the risk of infection and osteonecrosis [10, 12]. According to the literature,

methotrexate is the anchoring treatment for rheumatoid arthritis [13]. Despite the anemia in our patient, treatment with methotrexate was started. An improvement in the anemia was observed. According to Brandalize, the use of methotrexate resulted in an improvement in pain and joint functionality; however, a disturbance in the platelet count was observed [14].

CONCLUSION The coexistence of rheumatoid arthritis in sickle cell patients makes the diagnosis of polyarthritis difficult. The usual treatment for each of the associated conditions is nonetheless effective.

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