Juvenile idiopathic arthritis in a Congolese patient with sickle cell haemoglobin C disease: Case report

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Abstract

Sickle Cell Disease (SCD) presenting with musculoskeletal manifestations may be difficult to distinguish from Juvenile Idiopathic Arthritis (JIA), especially in sub-Saharan area where SCD is endemic and share some clinical aspects with JIA.

We report a case of JIA occurring in a patient with a Sickle Cell Haemoglobin C Disease (SCHCD). The patient was diagnosed as SCHCD at the age of two years and underwent two blood transfusions for anemia at the age of 2 and 3 years. Musculoskeletal manifestations appeared at the age of 12 years and consisted of pain, swelling and deformity of the fingers, tees, wrists and ankles and were suggestive for JIA.

Keys words: Juvenile idiopathic arthritis, sickle cell disease, DR Congo

Introduction

Bones may be affected by both haemoglobin and vasocclusive processes in SCD. Joint symptoms during painful crisis generally result from pain in the juxta articular areas of bones being referred to the knees, ankles, wrists, elbows and shoulder. Occasionally, painful crisis may be associated with one or more warm, tender, swollen joints[1].

Systemic Lupus Erythematosus (SLE)[2,3], fibromyalgia[4], rheumatoid arthritis[5] chronic synovitis with profuse plasma cell infiltration and cartilage destruction[6], Juvenile idiopathic arthritis[7], increased frequencies of rheumatoid factor and antinuclear antibodies have been described[8] in association with SCD.

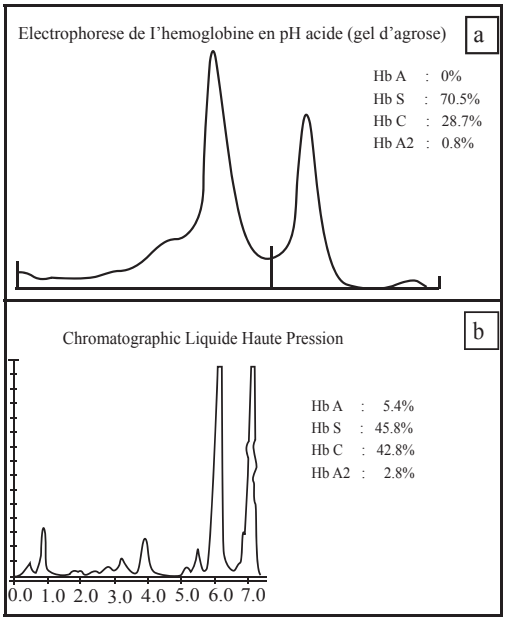
SCD is an endemic haemoglobinopathy in sub-Saharan Africa, SS haemoglobin being the most predominant and severe type in DR Congo and Central Africa while SC haemoglobin, which is less severe is confined to West African population.

In this study we report a case of a Congolese patient suffering from SCHCD with clinical manifestations suggestive for JIA.

Case report

A 16 year old patient was referred to a general practitioner in October 2008 for pain, swelling and deformity of the hands, wrists, feet and ankles. His past history revealed blood transfusions for anaemia at the age of 2 and 3 years. He was diagnosed as SCHCD (Table 1).

Table 1: Haemoglobin type of the patient performed by Agarose gel electrophoresis (Table 1a) and High Pressure Liquid Chromatography (HPLC) (Table 1b)



His mother was of Congolese (DR) origin and her haemoglobin was of AS type while his father was of West African origin (Benin) and his haemoglobin was of AC type. The physical examination revealed a normal physical growth since his weight, height, arm spam and cranial circumference were similar to the values found in a normal Congolese population of the same age and sex (Table 2).

Table 2: Patient anthropometric parameters

Parameter	In patient value	Normal values
Weight	48 kg	48 – 66 kg
Height	173 cm	171 – 173 cm
Cranial circumference	73 cm	70 – 75 cm
Arm span	173 cm	168 cm ± 93 cm

The tangential palpation of metacarpophalangeal (MCP) and metatarso phalangeal (MTP) joints was painful. The swelling of MCPS, MTPS, and of proximal interphalangeal (PIP'S), of the wrists and ankles was observed. A boutonnière deformity of the PIP'S of the fingers II and V of both hands was also noted (Figure 1).

Figure 1: X- Ray and photographs of the hands
Legend: Circle shows boutonniere deformity

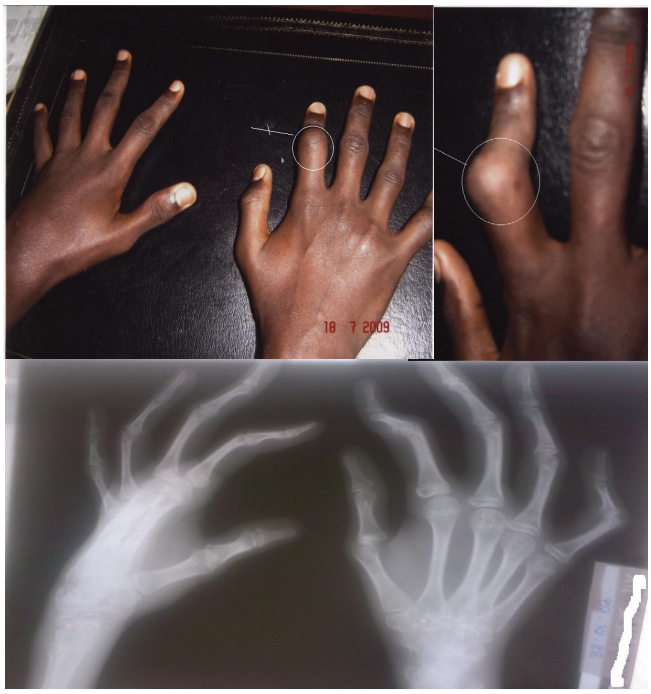


Table 3: Laboratory parameters

Parameter	In patient value	Normal values
Haemoglobin	11gr%	10 – 12 gr %
WBC	80.000/mm^3	4500 – 1100/mm^3
ESR	4 mm / Hour	0 – 15 mm/Hour
Blood sugar	104 mg%	80 – 110 mg%
Rheumatoid factor	Negative	Negative
L.E. Cells	Negative	Negative
HIV anti body	Negative	Negative

Both wrists and ankles were ankylosed and painful. Ankles remained in varus position. Except for haemoglobin level that was slightly low (11g%). Rheumatoid Factor (RF) LE cells and HIV antibody were negative (Table 3).

Discussion

Association of autoimmune disorders and SCD is still a matter of curiosity. SCD is most common in Western Africa while the homozygous type is confined to Central Africa. Its clinical features are less frequent and less severe than in homozygous sickle cell disease[9]. Juvenile Idiopathic Arthritis in this patient fulfilled the American College of Rheumatology (ACR) criteria for diagnosis: age of onset <16 years, arthritis ≥ 1 joints, duration of disease ≤ 6 weeks. The type of onset seems to be of oligo articular type.

As could be seen, hand photographs (Figure 1) showed boutonnière deformity of fingers and X-ray of hands revealed periarticular, osteoporosis. No evidence of erosive arthritis was seen. Likewise, RF and antibody to HIV-1 and 2 were absent. The absence of HIV antibody may exclude reactive arthritis to HIV. Our case highlights that JIA may be associated with SCHCD. Although this association is rare it may be the matter of confusion in endemic area for sicke cell anemia especially at the clinical point of view.

Acknowledgements

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References

1. De Ceulaer, K, Forbes, M, Serjeant GR. Non – gouty arthritis in sickle cell disease: report of 37 consecutive cases. *Ann. Rheum. Dis.* 1984; **43:** 599 – 603.
2. Wilson WA, Nichols GD, Hughes GRV, Amin, S, Alleyne GAO, Serjeant GR. Systemic lupus erythematosus and sickle - cell anaemia. *Br Med J.* 1976; **1**:813.
3. Katsanis E, Hsu E, Luke, K-H, McKee, JA. Systemic lupus erythematosus and sickle hemoglobinopathies: a report of two cases and review of the literature. *Am. J. Hemad*. 1987; **25**: 211 – 214.
4. Naomi S. Clues to pathogenesis of fibromyalgie in patients with sickle cell disease. J. *Rheumatol*. 2004; **31;** 598 – 600.
5. Marino, C, Mc Donald, E. Rheumatoid arthritis in a patient with sickle cell disease. *J. Rheumatol.* 1990; **17**: 970 –972.
6. Schumacher, HR, Dorwart BB, Bond J, Alavi A, Miller W. Chronic synovitis with early cartilage destruction in sickle cell disease. *Ann. Rheum. Dis.* 1997; **36:** 413 – 419.
7. Nistala, K, Murray JK. Coexistent sickle cell disease and juvenile rheumatoid arthritis. Two cases with delayed diagnosis and severe destructive arthropathy *J. Rheumatol.* 2001; **28:** 2125 – 2128.
8. Ofusu MD, Saunders DA, Dunston GM, Castro O, Alarif L. Association of HLA and auto antibody in transfused sickle cell disease patients. *Am. J. Hematol*. 1986; **22**; 27 – 33.
9. Serjeant GR, Serjeant BE. In: sickle cell disease 3rd ED 2001. Oxford Medical Publications pp 445 – 455.