ORIGINAL ARTICLE

Systemic Onset Juvenile Chronic Arthritis (JCA) In A Nigerian Boy-A Case Report

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ABSTRACT

Background*: Juvenile chronic arthritis (JCA) is a chronic arthritis affecting children below age of 16 years. The systemic onset subgroup is also known as Still's disease. There are several distinct subgroups. There is paucity of literature of this disease entity in our environment due to under diagnosis of the disease.*

Method*: The case note of this patient was retrieved. He was managed for 2 years in the adult rheumatology clinic after being transferred from the Paediatric unit. Relevant literature was reviewed.*

Result*: There was a good response to immunosuppressive agents and low dose prednisolone. He was back at School and able to play with his mates after long withdrawal from School.*

Conclusion*: Prompt referral of such cases to specialist centers will go a long way in determining the outcome of such patients. Prognosis is better with early presentation and appropriate management.*

Key words*:systemic onset arthritis,persistent, polyarticular arthritis*

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INTRODUCTION

Juvenile chronic arthritis (JCA) is a chronic arthritis of childhood comprising of several different subgroups[1]. It is one of the most common rheumatic diseases of childhood[1]. Because most of the majorities of children are rheumatoid factor negative, it is also called juvenile idiopathic arthritis.[1]

Although the true incidence and prevalence of JCA are unknown. Its incidence is estimated at two to 20 per 100,000 children per year worldwide based on the American College of Rheumatology (ACR) Criteria.[2] Neither the aetiology nor risk factors of JCA have been identified. It is considered to be an autoimmune disease in which both the cell mediated and/ or humoral processes are involved.[3]

Currently, there is no universal consensus on the classification of the JCA. Several distinct subgroups have been suggested. The American College of Rheumatology criteria require that the age at onset of arthritis be less than 16 years, and that arthritis be present in one or more joints for at least six weeks.[4] The subgroup type of JCA is determined by the first six months of disease and include pauci-or oligoarticular JCA, polyarticular- JCA, and systemic onset JCA.[4]

Systemic onset JCA, also known as Still's disease, is characterized by high intermittent fever, maculopapular rash and other organ system involvement.[5] It affects about 10% of children with JCA occurring equally in males and females.[5] Pauci-articular JCA describes the involvement of 4 or fewer joints and affects an estimated 40-60% of children with JCA.[5] Iridocyclitis is a dreaded complication in this subset. Polyarticular type involves more than 4 joints and affects about 35% of children with JCA.[5]

Useful investigations in JCA include markers of inflammatory diseases (Erythrocyte sedimentation rate, C- reative protein), rheumatoid factor, antinuclear antibody and X-ray of the affected joints. Joint aspiration and biopsy are particularly helpful in monoarticular arthritis for differential diagnosis.

The treatment options include non-steroidal anti-inflammatory drugs (NSAIDS), hydroxychloroquine, oral gold salts, D-penicillamine, methotrexate and anticytokines[6] (infliximab, etanercept). Glucocorticoids are useful in the short-term management of JCA.

CASE HISTORY

A 14 year old boy was referred to the adult rheumatology clinic after being managed as a case of systemic onset juvenile chronic arthritis for six years. He first presented to the paediatric unit at the age of eight years with history of being unwell for four years with high grade swinging fever, anorexia and weight loss. He also complained of pain in the knees. He was noted to be stunted and had quotidian fever pattern, lymphadenopathy but no splenomegaly. The cardiovascular examination was normal. ESR was high (ESR 120mm/hr) and had a PCV of 26%. He was managed with high dose of non-steroidal anti-inflammatory drugs and low dose prednisolone. Despite these treatments he continued to have active disease (ESR 46mm/hr).

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He subsequently developed pain in his wrists, ankles, elbows, and the neck. He was then transferred to the adult rheumatology unit at age of 14 years. He had withdrawn from primary school because of persistent pain.

At examination, he was found to be stunted (140cm tall at the time of transfer to adult care, with only 40cm gain in height from diagnosis). There was no evidence of iritis. The joints of the hands, the wrists, ankles, and the knees were swollen and boggy with varying tenderness. There was restriction of movements of the affected joints. Neck movement was also restricted. The liver and the spleen were not palpably enlarged and there was no lymphadenopathy.

ESR was elevated (ESR 76mm/hr), had normocytic normochromic anaemia (PCV 24%) with normal white cell counts and differentials. The rheumatoid factor was negative. Synovial fluid aspiration yielded no growth, but there were few inflammatory cells predominantly lymphocytes. Polarized microscopy was however not requested for. Plain radiograph showed ankylosis of the hands bilaterally.

He was managed with daily prednisolone, intraarticular methylprednisolone injection into the knees following synovial fluid aspiration. He was also placed on weekly disease modifying agent (methotrexate) and daily chloroquine (250mg). Six months after commencement of definitive treatment, he had less painful joints, and the range of movement of the affected joints improved significantly. Repeated ophthalmological examinations showed no evidence of retinopathy. He was back at school and able to play with his peers. He was followed-up for 2 years with stable clinical condition before he defaulted.

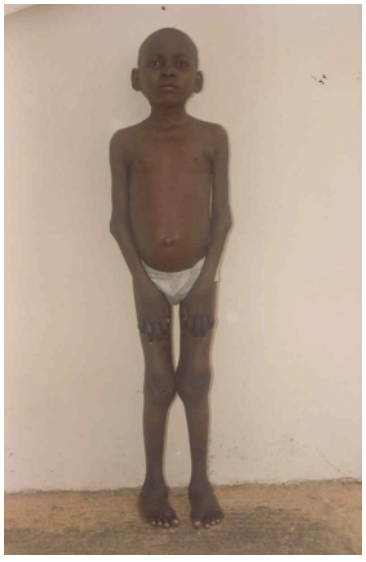

Fig 1. Stunted growth in a 16 year old with CJA.

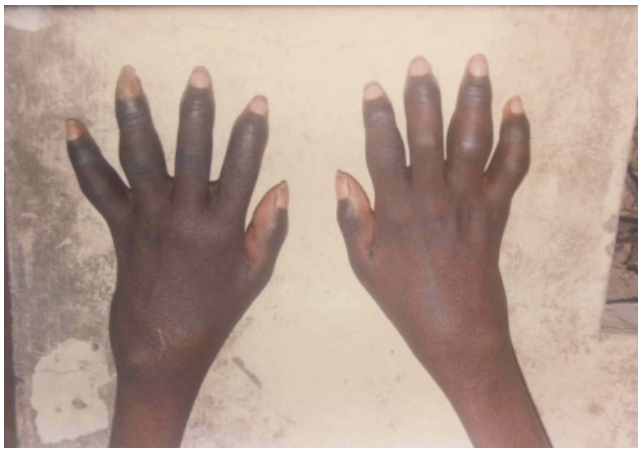

Fig 2- Ankylosed wrists and boggy digits

DISCUSSION

George Fredric Still in 1897 first recognized that there were several varieties of juvenile chronic arthritis, which seems to differ from classical, adult arthritis[7]. Of the major subtypes of juvenile chronic arthritis, the systemic onset (Still's disease) form is the one associated with the most serious short and long-term illness, as well as significant fatality following myocarditis.[8] This form is characterized more by its extra-articular features than by arthritis itself.[8] Systemic features alone may be present for weeks, months, and even years before the development of arthritis. It is often only after the development of arthritis that the syndrome is recognized as systemic- onset juvenile chronic arthritis.[8]

Still himself noticed a general arrest of growth when the disease begins in early childhood.[9] Growth hormone levels are normal in patients with systemic-onset juvenile chronic arthritis not treated with corticosteroids, but insulin-like growth factors I and II were below normal[9]. Delayed growth in affected children relates to disease activity, inadequate caloric intake and corticosteroid therapy.[9] (daily prednisolone of more than 5mg/m^2 will result in delayed growth). Ocular involvement is unusual, in contrast to the pauciarticular onset of juvenile chronic arthritis.[10] Nevertheless, asymptomatic anterior uveitis does occur in this group and these patients therefore require periodic slit-lamp examination.[10]

Rheumatoid factor negative subtype of juvenile chronic arthritis as is found in this patient, occurs in 20-30% of patients. The patient developed a persistent arthritis type with destructive arthritis of the hands with cervical spine involvement resulting in limitation of range of motion in the neck. Other complications of this subtype though not seen in our patient include micrognathia, leg length discrepancies, uveitis, angular deformities, and brachydactyly.[11]

The diagnosis of JCA depends on a comprehensive history and physical examination demonstrating the presence of chronic arthritis for at least six weeks and exclusion of other conditions, like viral arthritis, septic arthritis, reactive arthritis, and lyme disease[12]. There is no diagnostic test for JCA. Laboratory studies may reflect changes consistent with inflammation, but are not diagnostic.

The goals of therapy are to control pain and inflammation, to preserve range of motion and muscle strength, strive for normal function, growth, physical and psychosocial development, and to control systemic manifestations.[13] Patient and family understanding and participation in management are important. A multi-disciplinary team approach is essential in optimizing results. Physical therapy, occupational therapy and orthopaedic management are important. Non-steroidal anti-inflammatory drugs are important agents in the treatment of JCA.[6] Methotrexate is an effective agent in children with severe JCA and is frequently used to treat children who have failed to respond to NSAIDS.[6] Hydrochloroquine is sometimes used as an adjunct for the treatment of JCA in older children[6]. Ophthalmological examination every six months is necessary to monitor patients on hydroxychloroquine since retinopathy can occur. Glucocorticoids may be useful in the short-term treatment of severe systemic disease, severe JCA refractory to other therapies, and iridocyclitis.[14] It is desirable to avoid prolonged use because of complications such as growth retardation, osteoporosis, infection, fractures and cataracts[14]. Biologic agents targeted against TNF-alpha and IL 1 (interleukin) have also been approved for use in children and are effective.[15]

Prognosis varies with the onset type or subtype and clinical course. It also depends on the time of presentation and appropriateness of therapy. Studies have however indicated that children are without serious disability and are able to work and function normally[16]. This was the case with our patient who was back at school when an appropriate treatment was commenced.

This write-up is to heighten our index of suspicion as no diagnostic test is available. I believe this disease is present in our environment but under reported.

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