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Muscular Atrophy and Limb-length Discrepancy Complicating Pauci-articular Juvenile Idiopathic Arthritis in a 14-year-old in Abeokuta, Nigeria: A Case Report and Review of Literature

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Abstract

Background

Juvenile idiopathic arthritis (JIA) is a chronic autoimmune inflammatory joint disease. It is the most common form of arthritis in children and adolescents. It is classified as pauci-articular, poly-articular, and systemic onset disease depending on the number of joints involved as well as presence of systemic symptoms.

Pauci-articular JIA commonly involves large joints such as the knee joint, and typically manifests clinically with pain, swelling, tenderness and joint stiffness. Promptly diagnosed and appropriately managed JIA typically has an excellent prognosis while poorly treated cases may result in debilitating sequelae such as uveitis, ipsilateral muscular atrophy, limb-length discrepancies, and flexion-contracture deformities at the involved joint(s).

Objective

To review literature and report a case of muscular atrophy and limb-length discrepancy as rare complications of poorly managed JIA.

Methodology

We report a case of JIA in a 14-year-old female who presented with a three-year history of recurrent right knee pain, asymmetry of the thighs of two years duration and abnormal gait observed 18 months prior to presentation. Physical examination revealed an antalgic gait as well as thigh circumference and lower limb-length discrepancies.

Results

Anti- nuclear antibody (ANA) was positive, rheumatoid factor (RF) was negative; ultrasound scan of the right knee joint revealed sonographic features that were in keeping with arthritic changes, while plain radiograph of the knee joint revealed right knee effusion with overlying soft tissue swelling.

Conclusion

This report highlights and reviews limb-length and thigh circumference discrepancies as complications of pauci-articular JIA in children and adolescents.

Keywords: Pauci-articular JIA, Limb-length Discrepancy, Thigh Circumference Discrepancy

Introduction

Juvenile idiopathic arthritis (JIA) is an umbrella term describing a heterogeneous group of conditions, characterized by chronic arthritis beginning before the age of 16 years, persisting for at least six weeks and having no identifiable cause [1]. It is the most common rheumatologic condition in childhood and adolescence [2]. Three major forms of JIA are typically described namely: Pauci-articular, polyarticular, and systemic onset disease [2]. The ILAR classification however includes other forms of arthritis such as psoriatic, enthesitis-related arthritis, and undefined arthritis [1]. Pauciarticular JIA is the most common form of the disease and refers to disease limited to not more than four joints in the first six months [3]. It predominantly affects the joints of the lower extremities, such as knee and ankle, with a high frequency of positivity to anti-nuclear antibody (ANA)and high risk of chronic uveitis [4].

The overall prevalence is estimated to be 1-2 per 1,000 children, with an incidence of 1 per 10,000.[2] The onset of the rheumatoid factor- (RF) negative subtype follows a bimodal distribution, with the peak occurring between 1-3 years of age, and the second peak occurring later childhood at 10-13 years [4].

Juvenile idiopathic arthritis is characterised by auto-reactive antigen specific T-cells and high titres of autoantibodies, and typically show strong associations with MHC class II alleles [4]. Association with HLA class II and presence of ANA suggests

that an adaptive immune response is predominant in the pathogenesis of pauci-articular JIA [4]. Initiation of the JIA pathophysiologic cascade includes abnormal activation of T-cells, B-cells, natural killer (NK) cells, dendritic cells (DC), macrophages and neutrophils and the production of pro-inflammatory mediators that cause joint destruction and systemic complications [4].

Clinical manifestations of JIA include intra-articular swelling, limitation of movement in association with pain, stiffness, warmth, or erythema of the joint, and an antalgic gait. Chronic problems such as uveitis, limb-length discrepancies, muscle atrophy and flexion contractures may develop if pathologies are not effectively managed [2, 5]. The diagnosis of JIA is based on history, suggestive physical examination findings, laboratory markers, and imaging modalities. Pharmaceutical therapy with non-steroidal anti-inflammatory drugs (NSAIDs), corticosteroids, disease modifying anti-rheumatic drugs (DMARD), and biological agents is the forefront gold standard approach in management [5]. Emerging evidence on non-invasive physical and mechanical therapies in JIA, suggest that allied health professionals also play important roles in the management of this condition [5]. Early clinical intervention is desirable as prognosis is generally good. Late presentation and institution of treatment are however associated with long-term sequelae such as muscle wasting and limb-length discrepancies.

We present a 14-year-old adolescent female who presented with muscle wasting and limb length discrepancy associated with belatedly diagnosed JIA to highlight these chronic complications associated with delayed diagnosis and poor management of this disorder.

Case Summary

S.H was a 14year old female who presented to the paediatric out-patient department on account of a three-year history of recurrent right knee pain and stiffness, difference in sizes of the thighs of about two years duration, and an abnormal gait observed 18 months prior to presentation. The right knee pain was insidious in onset, aching, and severe enough to disturb sleep and activity. The pain was aggravated by walking and exertion, and temporarily relieved by analgesics such as ibuprofen and diclofenac procured over the counter. The pain did not radiate to any other part of the body. There was associated stiffness of the joint with inability to full extend the right knee joint. Pain score ranged from 6-8/10 in between analgesic use. There was no preceding history of fall nor trauma.

About a year following onset of right knee pain, the parents noticed that the right thigh was progressively getting smaller in size as compared to the left thigh. Six months later she began walking with a limp, and the abnormal gait progressively worsened despite continued administration of analgesics. Since the onset of symptoms, she had presented at various private clinics and had at separate times been on ibuprofen, diclofenac, folic acid, and calcium tablets without resolution of symptoms. On account of the persistence of the pain, and the obvious reduction in the size of her right thigh muscles, her parents decided to seek alternative care with traditional bone setters. Symptoms however persisted till the change in gait was noticed, which necessitated presentation at a General Hospital from where she was referred to our Centre.

Physical examination revealed an adolescent walking with a limp; there was pain on both active and passive extension of the right knee joint, but no obvious swelling at the knee joints. The muscle bulk of the right thigh was obviously reduced compared with the left side. The circumferential measurement around the right thigh measured 10 cm from the patella [6] was 24cm, as against 28cm on the left side giving a thigh circumference discrepancy (TCD) of 4cm (see Fig 1). There was also a significant limb-length discrepancy (LLD) with three consecutive measurements taken from the anterior superior iliac spine to the medial malleolus measuring 89cm on the right side and 86cm on the left; giving a limb-length difference of 3cm. Limb-length difference was further confirmed by checking the height of the knees with the child's pelvis flat on the couch and the medial malleoli held together, and by checking the height of the iliac crests with the child standing [6]. The examinations of the arms, spine, pelvis and other systems were unremarkable.

Fig 1: Showing index patient with reduced right thigh circumference

A diagnosis of suspected pauci-articular JIA of the right knee joint with muscular atrophy and limb-length discrepancy was made. Plain radiograph and ultrasound scan of the right knee joint were ordered, while samples were also obtained for complete blood count (CBC), erythrocyte sedimentation rate (ESR), haemoglobin electrophoresis, rheumatoid factor (RF) and anti-nuclear antibody (ANA).

Results

Anti- nuclear antibody (ANA) was positive, rheumatoid factor (RF) was negative. Ultrasound scan of the right knee joint showed thickening of the right quadriceps fat pad, moderate reduction of its echogenicity indicative of inflammatory changes, minimal joint effusion, with no intra-

articular bodies noted. These sonographic features are in keeping with arthritic changes. The plain radiograph of the knee joint revealed right knee effusion with overlying soft tissue swelling. Erythrocyte sedimentation rate (ESR) was elevated, FBC parameters were normal, while haemoglobin phenotype was AS,

Treatment and outcome

She was initially commenced on oral diclofenac and oral corticosteroids, but oral methotrexate was subsequently introduced with significant improvement in symptoms. She also commenced physiotherapy and was scheduled for follow-up and further care at the rheumatology clinic. She however defaulted after about 6 months of out-patient follow-up.

Discussion

The diagnosis of pauci-articular JIA in the reported case was based on the history of right knee pain, stiffness, and swelling which began three years prior to presentation and was limited to only one joint as at the time of presentation. Plain radiograph and ultrasound scan findings of the affected joint were in keeping with JIA, while ANA was positive. The index patient also had a limb-length discrepancy (LLD) of 3cm and thigh circumference difference (TCD) of 4cm.

Juvenile idiopathic arthritis is typically characterised by pain and functional disabilities that significantly impact quality of life. In addition, several studies have also reported long-term sequelae of the disease such as uveitis as well as generalised and local growth abnormalities in children and adolescents [2, 7-12]. According to Bechtold *et al*, [7] global growth impairment resulting in reduced final height has been noted to be one of the permanent consequences in 10–20 % of children and adolescents with systemic and polyarticular JIA. It has been proposed that inflammatory cytokines, prolonged administration of glucocorticoids and undernutrition, all of which interfere with the growth hormone–insulin-like growth factor axis, are at the hub of the diminished growth in JIA [7], while growth hormone has been found to be beneficial in such children [2, 7, 8].

Local growth disturbance on the other hand results in overgrowth of the inflammed joint, typically the knee, resulting in leg-length discrepancy (LLD) with the longer leg on the side of the affected joint [2]. The potential for limb overgrowth is higher when the disease begins before the age of three years as reported by Skytta *et al* and Vostrej *et al*, LLD may however also occur in older children and adolescent especially when appropriate treatment is delayed as it occurred in the reported case. Limb length discrepancies results from persistent inflammation at the growth plate which leads to increased metabolic activity, and subsequent excessive growth [3]. while localized atrophy and weakness are sometimes apparent in muscles close to active synovitis, particularly in the thigh muscle in association with knee arthritis as observed in the reported case [3, 9]. Such ipsilateral reduction in muscle bulk may be caused by disuse or reflex inhibition elicited by pain, [3].

It is generally reported that most children with pauci-articular JIA experience resolution of active synovitis regardless of whether they are treated with oral or intra-articular medications [10]. However, Oen *et al* [13] in a study involving 392 patients from 3 participating specialist rheumatology centres concluded the disease often extends into adulthood even though functional outcomes of patients have improved over the years. About half of patients with pauciarticular onset JIA evaluated by Oen *et al* [13] achieved remission by the age of 16 years, while the probability of continued active disease into the late twenties or early thirties was higher for patients who were not in remission by this age [13]. Oen *et al* also reported that remissions most often occurred within five years of onset, and that the probability of remission for patients with systemic and RF–negative polyarticular onset was low after the fifth year [13]. As for those with pauciarticular JIA, although the probability of later remissions was better, it also progressively decreases after five years [13]. This has a significant bearing on the potential development of local growth abnormalities since the degree of LLD or TCD has been shown to be related to the duration of synovitis according to Sherry *et al*. [10] The disease duration of greater than three years in the index case, therefore, increases the risk of growth abnormalities.

This report also reinforces the tendency for patients in resource poor settings to present late for orthodox care, by which time complications of diseases have usually ensued. According to a survey by Adelowo *et al*, all the 23 children with JIA evaluated in their study presented late to the rheumatology clinic with a mean disease duration of 3.7 years, and about 11% already had general or localised growth abnormalities at presentation. Most of them had been on NSAIDS and oral corticosteroids without improvement and had to be commenced on oral methotrexate or intraarticular (IA) corticosteroids. The use of intra-articular (IA) steroids in children with pauciarticular JIA is a standard therapeutic option, and its use early in the disease has been reported to prevent LLD and TCD [10]. Intra-articular steroids have been found to cause to rapid resolution of the symptoms and signs of inflammation in JIA. The optimal timing for the commencement of IA steroids, is however not known [10].

An algorithm for the assessment and management of LLD in children with JIA was proposed by Skytta *et al.* [3] This algorithm commences with regular clinical assessment for limb length equality at each paediatric rheumatology clinic visit [3]. Patients with persistent knee arthritides are to be managed with DMARD such as oral methotrexate, as well oral and intra-articular glucocorticoid injections; while synovectomy is to be considered if repeated IA injections proved ineffective after 12 months of administration [3]. Clinical limb length inequality less than 10–15mm may be managed with shoe lifts, while growth charts and periodic hand and knee radiographs are to be reviewed in order to assess growth and growth correction potential as well as the skeletal age [3]. Total arthroplasty, especially of the hip and knee, represents a successful option in the presence of severe functional impairment, and is generally delayed until growth has stopped [3]. Physiotherapy and occupational therapy are also important components of the therapeutic approach, and psychological support may also be provided.

This multi-disciplinary approach to the management of complicated JIA was proposed for the index patient but was unfortunately not actualised following the discontinuation of follow-up at the patient's instance. This is unsurprising, considering the high rates of default in care of chronic diseases in our setting owing to the high cost of out-of-pocket spending for healthcare., as well as caregiver burnout. Adelowo *et al* reported that only about a third of

patients in their review were still attending clinics at the completion of their study. The enormity of the direct and indirect costs of managing JIA cases has been evaluated by various authors [14-16]. According to Minden *et al* [14, 16] cost-driving factors in the management of JIA include disease subtype, disease activity, pain intensity, and level of disability, with higher costs being incurred by patients with more severe disease and higher disease activity [14, 16].

Conclusion

Early diagnosis and prompt treatment is desirable in children and adolescents with JIA as prognosis is generally good, while delayed presentations and institution of appropriate treatment are associated with long-term sequelae such as muscle wasting and limb-length discrepancies as well as ophthalmic complications.

Conflicts of interest

None.

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