Dermatomyositis with renal involvement in a Nigerian girl

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Abstract

Introduction: Juvenile dermatomyositis is a rare autoimmune muscular disorder that affects children usually less than 18 years. Common features of the disease are characteristic skin changes, calcifications within muscle groups and symmetrical proximal muscle weakness. The prevalence of this condition among Africans is unknown and only one case has been reported so far from Nigeria. We report dermatomyositis with renal involvement.
Methods: A case report of a 13 year old Nigerian girl using reports from her medical record. She presented with generalised body swelling, weakness of the limbs with inability to walk or lift herself out of bed without support and a year history of recurrent skin rashes.
Result: Findings at admission included a chronically ill girl who was mildly pale and had generalised oedema. There were multiple areas of hypopigmented macules on the face, arms, right elbow and the shoulder. Her total urine output in the first 24 hours of admission was 0.9ml/kg/hr, had a systolic and diastolic blood pressure that was above the 95th centile for her age for. Plain radiograph of the limbs showed widespread intramuscular calcifications. She was managed conservatively with oral corticosteroid, antihypertensive and analgesics and has remained fairly stable with fewer episodes flaring of symptoms.
Conclusions: Renal involvement in juvenile dermatomyositis even though rare could be a significant clinical feature in children with this condition.

Keywords: Juvenile, dermatomyositis, renal involvement, oral corticosteroid.

Highland Med Res J 2014;14(1):48-50

Introduction

Juvenile dermatomyositis is a rare systemic autoimmune muscular disorder that affects children less than 18 years.[1] It is the most common childhood inflammatory myopathy.[1] The incidence rate is 9.63 cases per million popuation and the ratio of affected girls to boys in the United State is 2-3: 1.[1,2] The prevalence of the condition among Africans is not known and there are only a handful of reported cases. To the best of our knowledge, there is only one reported case so far in Nigeria.[3] Excessive ultraviolent light exposure has recently been identified as one of the risk factors. Characteristic findings include Gottron papules, Heliotrope rash, calcinosis cutis and symmetrical proximal muscle weakness.[1] There are few reported cases of renal involvement, which may be as a result of acute renal failure from myoglobinuria or chronic glomerulonephritis.[4] We present a case of Juvenile dermatomyositis with renal involvement.

Case Report

J.N, a 13 year old female that first presented in the emergency paediatric unit of our facility 3 years ago with a one year history of recurrent skin rashes and four months of body swelling with subsequent joint stiffness. Some of the rashes which involved the face, limbs and the neck, started as erythematous lesions, became ulcerative, then healed with hypopigmented scars. Onset of the rashes was associated with a high grade fever. Stiffness affected large joints and associated weakness led to difficulty with raising the hands to comb the hair, walking, rising from a sitting position, and getting out of bed. There was also a history of dysphagia and loss of scalp hair. Body swelling which started from the lower limbs regressed with the day and was associated with oliguria. She had no dark coloured urine.

Physical examination on admission revealed a chronically ill looking girl who was mildly pale and had facial puffiness. She had multiple hypopigmented macules involving the face, fore arms, interphalangeal areas, right elbow, and the shoulders. (Figure 1)

The skin around the fingers appeared thicker and slightly paler(Gottron papules). There was an erythematous diffuse swelling on the medial aspect of the right thigh. It was tender and there was differential warmth. No subcutaneous nodules or swellings were appreciated. She had a limited range of movement across the shoulder, elbow, hip and knee joints. She had an elevated blood pressure of 140/90, which was above the 95th centile for her age and height. The diagnosis of Juvenile dermatomyositis with cellulitis and acute kidney injury was made (AKI).

Urinalysis revealed haematuria but, serum

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electrolytes, urea and creatinine values were within normal limits. Her urine output over the initial 24 hours on admission was 0.9ml/kg/hr. ESR on admission was 40mm/hr and there was marked neutrophilia of 28,800 cells/μL, with a total WBC of 36,000 cells/μL. Plain radiograph of the limbs showed widespread intramuscular calcification.(Figure 2)

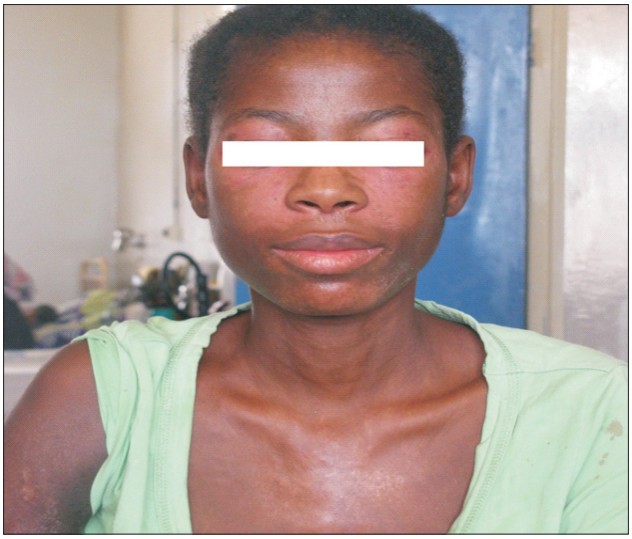

Figure 1. Multiple hypopigmented macules in a 13 year old girl with dermatomyositis

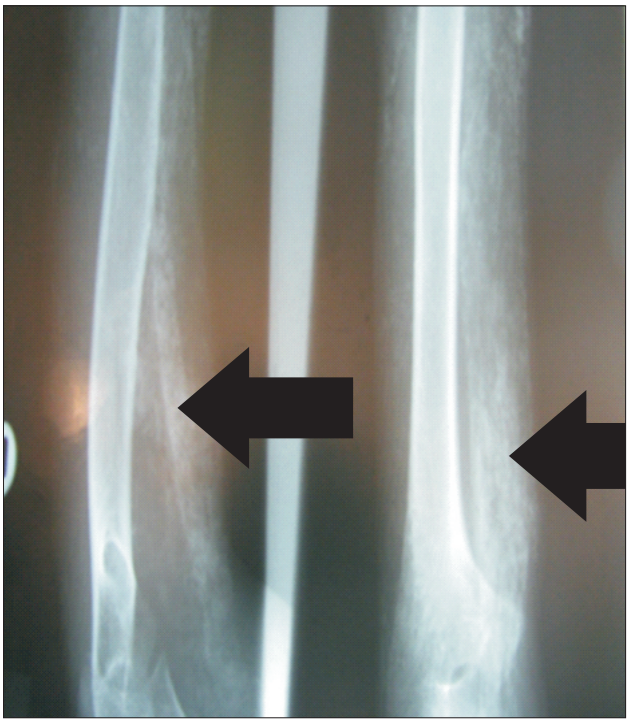

Figure 2. Radiograph showing widespread intramuscular calcifications (Dark Arrows) in a 13 year old girl with dermatomyositis

Renal ultrasound revealed bilateral loss of renal echotexture and corticomedullary differentiation. Retroviral screening was non reactive. Muscle biopsy, serological tests (ANA, Antibodies to Pm/Scl), MRI were not done as a result of absent facilities or lack of fund. She was commenced on IV antibiotics on account of the cellulitis, antihypertensives and tabs prednisolone at 2mg/kg/day. She was discharged home stable after 2 weeks of hospital stay. After the initial admission, she had 3 subsequent admissions as a result of diffuse inflammation of some muscle groups and cellulitis, occasionally accompanied with ulceration and cheesy discharge. Currently, the fequency of the flares has reduced significantly as she has only had one admission in the last 1 year. The severity of the joint stiffness has also reduced since the commencement of physiotherapy. Her current blood pressure is 120/70mmHg and renal function has been normal. She is still on antihypertensives and prednisolone.

Discussion

The lack of adequate diagnostic facilities still remains a challenge to the diagnosis of diseases such as juvenile dermatomyositis in developing countries like Nigeria. The demonstration of the five criteria suggested by Bohan and Peter[5,6] (which include progressive proximal symmetrical muscle weakness, elevated levels of muscle enzymes, abnormal electromyographic findings, abnormal finding on muscle biopsy and characteristic cutaenous changes), may not be practicable in a developing country such as ours where facilities are limited.

The diagnosis of this patient was made largely on clinical grounds. She had difficulties in rising from a siting position and carrying out activities such as combing of her hair-this is an evidence of proximal muscle weakness. This is in addition to the dysphagia, which may also be as a result of degenerative changes affecting the muscles of degluttition. The cutaenous changes she had included the Gottron papules and the macules observed in the exposed parts of the body.. There was also the history of hair loss, which has been previously documented in patients with dermatomyositis.[7]

The dermatomyositis is considered to arise from a complement mediated immune attack against muscle vessels which result in micro-infarction and susequent widespread muscle necrosis and subsequent degenerative changes.[8] The factors that may trigger this process in an individual who is already genetically predisposed may include infections with organisms such as enteroviruses and group A beta haemolytic streptococcus.[9] We were not able to establish any possible trigger in this patient. It may however be difficult to know, especially if all she had was a mild illness with vague and non specific signs, which is not

uncommon in viral illnessess.

Renal involvement in dermatomyositis is said to be rare in children.[1] The two possible pathways that have been considered are acute kidney injury from myoglobinuria due to excessive muscle breakdown and glomerulonephritis, which may arise from immune complex deposits at the basement membrane. The features of renal involvement that our patient had included the body swelling, reduced urine output, elevated blood pressure. Urinalysis also revealed haematuria with absent proteinuria, portending a diagnosis of AKI.

Calcification is commoner in children and is said to be found in 2-30% of them at diagnosis.[1] It is said to be a marker of severity and may portend poor prognosis. It often signifies a chronic illness. Such patients often have calcium deposits in muscles with occasional cheesy discharge. This creates a nidus for recurrent infections. Our patient had a similar recuring cheesy discharge with accompanying evidence of infection. We also saw evidence of widespread calcifications on plain radiograph of the long limbs (Figure 2).

The care for children with dermatomyositis is multidisciplinary and involves a paediatric rheumatologist, physiotherapists, social workers etc. Treatment essentially invovles the limitation of exposure to excessive UV rays by the use of sunscreens. Vitamin D supplements are given to reduce the risk of osteopenia, which is a known complication. Immunosuppressants such as corticosteroids and hyroxychloroquine are given to suppress the immune response. Agents such as methotrexate, cyclophosphomide and cyclosporine may be considered in patients whose laboratory parameters fail to normalize following the use of steroids. Complications such as acute kidney failure can be managed by maintaining a careful balance of fluid and electrolytes, contol of blood pressure and prompt dialysis where it is necessary. Physiotherapy plays a very vital role in reconditioning and strenghtening the muscles, especially after the inflammatory flares. The prognosis has markedly improved following the advent of steroids with mortality rate as low as 3% and period of acute inflammation reduced to less than 18 months.[9]

Conclusion

JD is a rare condition and the diagnosis may be missed, especially in the tropics where we are overwhelmed by infectious conditions. Inspite of the challenges of lack of diagnostic facilities and poverty, the diagnosis of the condition can still be made on clinical grounds, and effective therapy implemented.

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