Case Report

Juvenile Dermatomyositis in an 11 Year Old Nigerian-Boy: A Case Report and Review of Literature

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Abstract

We report the case of an 11-year-old boy with proximal myopathy, heliotrope, and Gottron papule-like rashes. Serum chemistry revealed muscle enzyme elevations, whereas muscle biopsy histology showed necrosis and inflammation, which were in keeping with juvenile dermatomyositis. Plain radiographic examination of the thigh 3 weeks after commencing treatment with prednisolone was normal. The aim of this presentation is to highlight the diagnostic challenges posed by this rare condition in a resource-limited setting and to underscore the need for prompt diagnosis and appropriate management. We hope that this report will assist physicians practicing in similar settings to make a prompt and accurate diagnosis when confronted with the same disease.

Keywords: Diagnostic challenges, juvenile, myositis dermatoses

Introduction

Juvenile dermatomyositis (JDM) is the most common inflammatory myositis in children.[1] It is a very rare but serious systemic autoimmune condition with an estimated incidence of three children per million in a year.[2,3] The underlying pathology results from inflammatory cell infiltrates, leading to vascular inflammation affecting the muscle and the overlying skin. It is distinguished by proximal muscle weakness and a characteristic rash. Evidence suggests that the etiology of JDM is multifactorial. It is thought to be based on genetic predisposition and an unknown environmental trigger.

The characteristic rashes in JDM are the heliotrope rash and the Gottron papules. Other features that can be reported at presentation include fever, dysphagia or dysphonia, arthritis, muscle tenderness, and fatigue. The presence of the characteristic rash as well as at least three signs of muscle inflammation and weakness is diagnostic of JDM. Signs of muscle inflammation can be elicited clinically, whereas evidence for muscle inflammation, however, can be obtained from serum muscle enzyme elevation. Electromyography and muscle biopsy are helpful diagnostic investigations; the former may show myopathy and the latter inflammation and necrosis.[3-6]

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The paucity of documented literature in Africans and the challenges associated with making this diagnosis in a resource-constrained setting informed the decision to publish this report. The case is discussed with a view to highlight the important clinical variations in JDM in this child and provide information that may improve both diagnostic acumen and management in children from similar settings.

Case Report

An 11 year old boy was brought by his parents to the childrens' emergency ward of Ladoke Akintola University of Technology Teaching Hospital, Osogbo, with 1-month history of multiple joint pain and body rashes and 3-week history of fever, facial swelling, and easy fatigability. Joint pains started insidiously involving both upper and lower limbs. There was an associated

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weakness of the proximal aspects of both upper and lower limbs with subsequent inability to comb the hair.

The body rashes were described as small eruptions over the knuckles of the hands and hypopigmented oval-shaped lesions over the elbow. The patient also complained of prominent hypopigmented areas located between the eyes. He had experienced a fever which was described as very high and was present throughout the day. Fever was temporarily relieved with the use of paracetamol. Facial swelling was noticed at about the time of onset of fever. Easy fatigability was noticed 3 weeks prior to presentation, which was characterized by reduced effort tolerance on walking to distances of about 100 m and more. The family and social history were not contributory.

Physical examination showed an ill-looking afebrile boy, with facial puffiness and hypopigmentation of the malar areas of the face. Dark discoloration of the upper eyelids was observed [Figure 1]. Obvious oval, macular hypopigmented symmetrical rashes were observed over the elbows, knees, and the cubital fossae. There were also rashes of varied colors from hypopigmentation to hyperpigmentation over the posterior aspect of both ears. Hypopigmented papules were noted on the knuckles of the hands [Figure 2].

Blood pressure was 90/60 mmHg supine. Normal heart sounds were elicited on auscultation, and apex beat was located in the fifth intercostal space and midclavicular line. A modified Gowers' sign on attempting to rise up from supine position was elicited. Power was 4/5 in both upper limbs and 3/5 in both lower limbs. Central nervous system and other systemic examinations were essentially normal.

A provisional diagnosis of JDM was made with differentials of acute rheumatic fever (RF) and juvenile rheumatoid arthritis (JRA). The parents and the patient were counseled and recounseled in the course of the evaluation about the working diagnosis.

Complete blood count, erythrocyte sedimentation rate, urinalysis, retroviral screening, serum muscle enzymes, and muscle biopsy were undertaken. The results of the investigations conducted are summarized in Table 1.

Figure 1: Darkly pigmented eyelids of the patient

The patient was subsequently commenced on tablet prednisolone at 2 mg/kg. Plain radiographic examination of the right thigh, hip, and knee joints was requested 3 weeks after the commencement of prednisolone because of pain and tenderness in the limb; paracetamol was administered for the management of the limb pains. The skin lesions and joint pains slowly resolved over the weeks of treatment with steroids and paracetamol. Oral steroid was tapered off after 10 weeks of the commencement of therapy. The blood pressure was monitored throughout the course of his steroid therapy, and it remained within the normal range for age, sex, and height of the patient. He made a slow uneventful recovery over the months of outpatient treatment and follow-up.

Discussion

The incidence of JDM in Nigerians is unknown. The present case is the first of its kind on our hospital records. Previous reports on dermatomyositis document a gender and age group predilection, with the disease occurring more commonly in females and two peak age groups of affectation between 4 and 10 years, in childhood (juvenile), and 45–64 years in adults. The child in the present report showed clinical variation in presentation from the peak age of onset and gender predilection.

Skin lesions are a prerequisite for the diagnosis of JDM.[5-8] The characteristic rash is the heliotrope rash, which is a violet discoloration of the eyelids that may be associated with periorbital edema. In the case presented, the patient lacked the violet discoloration of the eyelids [Figure 1]. The Gottron's papules is another classic rash associated with JDM, and it is a bright pink or pale shiny thickened or atrophic plaque over the proximal and distal interphalangeal joints. The Gottron's papules in our patient were hypopigmented papules located around the proximal interphalangeal joints [Figure 2]. The presence of melanin pigment in the black-/dark-skinned individuals and its absence in Caucasians could account for the differences in color across races. Physicians managing black- or dark-skinned children with proximal muscle

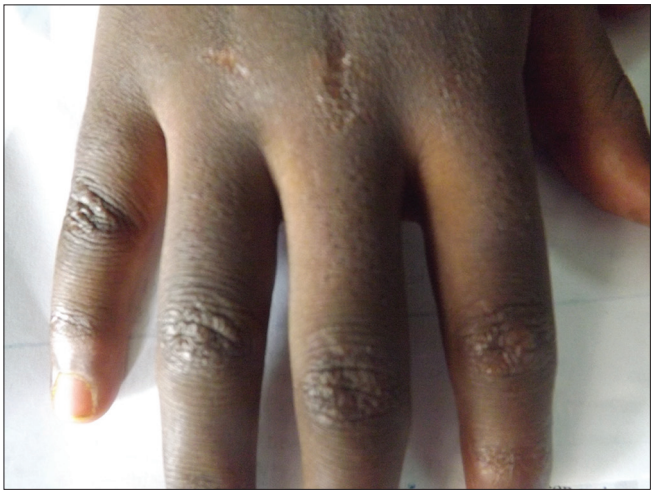

Figure 2: Hypopigmented papules in the proximal and interphalangeal joints of the hand (Gottrons papules)

Table 1: Results of investigations

Parameter	Results	Normal reference range
Serum muscle enzyme assays (µ/l)		
Muscle aspartate	69	1535
Lactate dehydrogenase	361	110-295
Creatine kinase	457	15-195
Aldolase	7.9	3.0-12.0
Hematological results and retroviral screening		
Packed cell volume (%)	32	36-41
ESR (mm/h)	11	<30 Westergreen
Retroviral screening	Negative	
Urine chemistry		
Urinalysis	Normal	
Muscle biopsy		
First skeletal muscle biopsy conducted around the thigh	Normal	
Second skeletal muscle biopsy conducted around the forearm	Moderate-to-severe perifascicular atrophy with reduction in some of the skeletal muscle sizes. Few blood vessels seen and some were surrounded by inflammatory cells, mostly lymphocytes	
Radiology		
Plain radiograph of the right thigh, hip, and knee joints	Normal	

ESR – Erythrocyte sedimentation rate

weakness and skin changes over the eyelids, face, or joints should have a high index of suspicion and consider the diagnosis of JDM.

The characteristic cutaneous findings plus three of the four muscle features make a definitive diagnosis of JDM based on the Bohan and Peter criteria.[8] Our patient had elevated serum levels of muscle-related enzymes. These aforementioned cutaneous changes and muscle enzymes' elevation plus a muscle biopsy that showed inflammatory cells with features of myositis confirmed our diagnosis of JDM. It is pertinent to state that we made an initial mistake by obtaining our muscle biopsy specimen from a normal muscle group without overlying skin changes. This initially confounded the diagnosis. A repeat biopsy from a muscle site with overlying skin changes, confirmed the diagnosis, which further underscores the importance of obtaining the muscle biopsy from sites closest to the affected skin.

The diagnostic dilemma of JDM is often due to similarities it shares with other connective tissue diseases (CTDs). Fever, arthritis, and arthralgia are all common features of both JDM and CTD. Both RF and JRA were considered in this patient, but these were invalidated by the patient not meeting the Duckett Jones or other criteria necessary for diagnosing these other CTDs of childhood.[9,10]

Steroid therapy represents the mainstay of treatment in JDM. They are given at a dose as low as 1 to 2 mg/kg depending on the severity of skin lesions. Children with extensive vasculopathy evidenced by nail fold capillary dropout may have poor oral steroid absorption, thereby requiring high dose of intermittent intravenous methylprednisolone at 30 mg/kg (maximum of 1 g/day).[1-6] Furthermore, weekly parenteral methotrexate with oral folic acid is usually combined with methylprednisolone for patients requiring intravenous steroids.

Calcinosis of the soft tissue is a late development of poorly managed cases of JDM. The prompt and appropriate management of JDM probably prevented or delayed this complication. Oral prednisolone can be tapered off as the skin lesion of the disease wanes. Extensive immunosuppression from the disease condition and steroid use may require replacement immunoglobulin to help prevent infections.

JDM is a chronic illness that is distressing to both parents and the children. Our patient comes from a family of low economic status which affected the extent of our investigations, limiting us to request for the very basic and most essential ones. The patient also had physiotherapy with counseling and recounseling sessions. Psychotherapy was offered to the patient and parents at different periods during admission and follow-up.

Conclusion

A high index of suspicion is required to diagnose and manage the disease promptly. Proximal weakness with dermatological changes over the joints and the face should be reasons to entertain a differential of JDM. Prognosis of JDM appears promising with prompt treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Robinson AB, Reed AM. Juvenile dermatomyositis. In: Kliegmann RM, Berhrman RE, Jenson HB, Stanton BF, editors. Nelson Textbook of Paediatrics. 20th ed. Philadelphia, USA: Saunders; 2014;1630-5.
2. Martin N, Li CL, Wedderburn LR. Juvenile dermatomyositis: New insights and new treatment strategies. J sage 2011;4:41-50.
3. Bingham A, Mamyrova G, Rother KI, Oral E, Cochran E, Premkumar A, *et al.* Predictors of acquired lipodystrophy in juvenile-onset dermatomyositis and a gradient of severity. Medicine (Baltimore) 2008;87:70-86.
4. Eimer MJ, Brickman WJ, Seshadri R, Ramsey-Goldman R, McPherson DD, Smulevitz B, *et al.* Clinical status and cardiovascular risk profile of adults with a history of juvenile dermatomyositis. J Pediatr 2011;159:795-801.
5. Adelowo O, Nwankwo M, Olaosebikan H. Juvenile dermatomyositis in a Nigerian girl. BMJ Case Rep 2014;2014. pii: bcr2013202132.
6. Martin N, Krol P, Smith S, Murray K, Pilkington CA, Davidson JE, *et al.* A national registry for juvenile dermatomyositis and other paediatric idiopathic inflammatory myopathies: 10 years' experience; the juvenile dermatomyositis national (UK and Ireland) cohort biomarker study and repository for idiopathic inflammatory myopathies. Rheumatology (Oxford) 2011;50:137-45.
7. Mustapha MG, Ashir MG, Mayun AA, Machoco Y, Ibrahim AB. Juvenile dermatomyositis in a Nigerian girl: A case report. Nig J Paed 2011;38:182-5.
8. Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). N Engl J Med 1975;292:344-7.
9. Shulman ST. Acute rheumatic fever. In: Kliegmann RM, Berhrman RE, Jenson HB, Stanton BF, editors. Nelson Textbook of Paediatrics. 20th ed. Philadelphia, USA: Saunders; 2014. p. 1331-7.
10. Rabinovich CE. Rheumatic diseases of childhood. In: Kliegmann RM, Berhrman RE, Jenson HB, Stanton BF, editors. Nelson Textbook of Paediatrics. 20th ed. Philadelphia, USA: Saunders; 2014. p. 1150-3.