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SHORT REPORT

Anasarca: not a nephrotic syndrome but dermatomyositis

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Abstract Juvenile dermatomyositis (JDM) is a rare autoimmune disease characterized by inflammation of the muscle, connective tissue, skin, gastrointestinal tract, and small nerves. Periorbital and facial edema may also be associated. Although localized edema is a common feature of JDM, generalized edema has rarely been reported. Here, we report a 3.5-year-old boy with JDM presenting with generalized edema. The diagnostic criteria of JDM rely on typical clinical manifestations that include: severe symmetric weakness of the proximal musculature, characteristic cutaneous changes, elevated serum skeletal muscle enzymes, and myopathic electromyographic pattern. Our patient initially received methylprednisolone and intravenous immunoglobulin (IVIG) without significant improvement, so he was given azathioprine and a prolonged course of oral prednisolone. We conclude that JDM should be suspected in patients presenting with anasarca in the absence of laboratory parameters of other causes of generalized edema and an appearance of heliotrope rash with muscle weakness. Also, we suggest that muscle magnetic resonance imaging (MRI) should be considered among the diagnostic tools of JDM.

Keywords Juvenile dermatomyositis · Generalized anasarca · Case report

Abbreviations

EMG	electromyography
IVIG	intravenous immunoglobulin
JDM	juvenile dermatomyositis
MRI	magnetic resonance imaging

Introduction

Juvenile dermatomyositis (JDM) is a multisystem disease characterized by the non-suppurative inflammation of striated muscles with characteristic cutaneous findings in the form of erythematous (heliotrope) periorbital rash [2]. The classical features of the disease include proximal muscle weakness, dermatitis associated with constitutional symptoms, and edematous and indurated muscles. The localized edema of muscle is a common feature, but anasarca as a presenting feature is rare [9].

Case report

A 3.5-year-old previously healthy male presented 1 month ago with a generalized edema (pitting edema of upper and lower limbs and periorbital edema), preceded by a history

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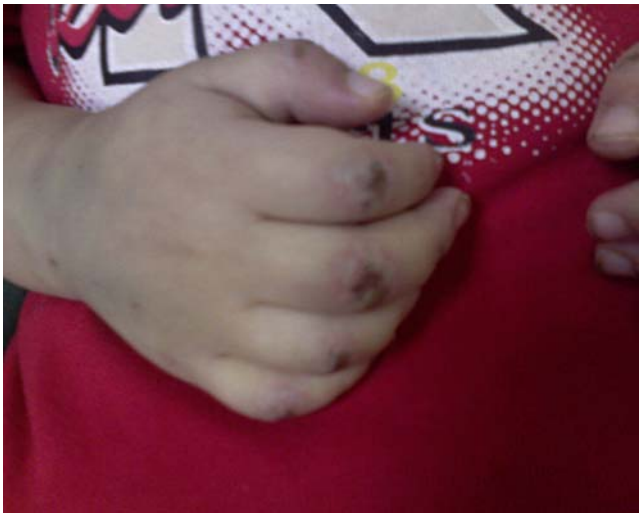

Fig. 1 The right hand of the patient shows Gottron's papules

Table 1 Laboratory results of the investigations performed

Investigation	Patient value	Reference value
Hemoglobin	13	12.5–15 gm/dl
White blood cell count	8,500	5,000–11,000/mm^3
Platelets	150,000	150,000–450,000/mm^3
Aspartate aminotransferase	1,437	Up to 40 U/ml
Alanine aminotransferase	494	Up to 45 U/ml
Creatine kinase, total	12,004	Up to 175 U/ml
Creatine kinase, MM	11,530	Up to 174 U/ml
Creatine kinase, MB	484	Up to 24 U/ml
Aldolase	7.9	Up to 7.6 U/ml
Anti-nuclear antibodies	Negative	
Anti-ds DNA ab	29	Up to 26 IU/ml
C3 of the complement	1.73	0.9- 2.1 gm/L

Creatine kinase MM=creatine kinase muscle-type isozyme
Creatine kinase MB=creatine kinase myocardial band

of upper respiratory tract infection. The patient was misdiagnosed as nephrotic syndrome and received oral steroids for a few days with partial improvement of edema, but urine analysis revealed no proteinuria. After 2 weeks, skin lesions appeared at the extensor surface of the hands (Gottron's papules) (Fig. 1) and heliotropic upper eyelids (Fig. 2) and dysphagia occurred, as well as excess drooling (gastrointestinal dysmotility) and chest secretions. It was associated with fever, severe symmetric proximal and axial muscle weakness, with mild distal muscle weakness, without affection of the antigravity muscle power.

The patient's laboratory investigations showed marked elevation of muscle-derived enzymes (Table 1).

Electromyography (EMG) showed myopathic changes. Calcinosis was not detected in the plain radiographs of the

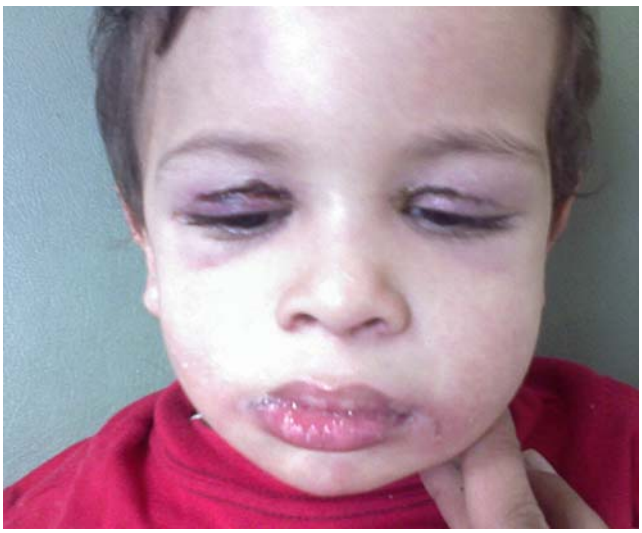

Fig. 2 The face of the patient shows violaceous upper eyelids, swollen lips, and excess drooling

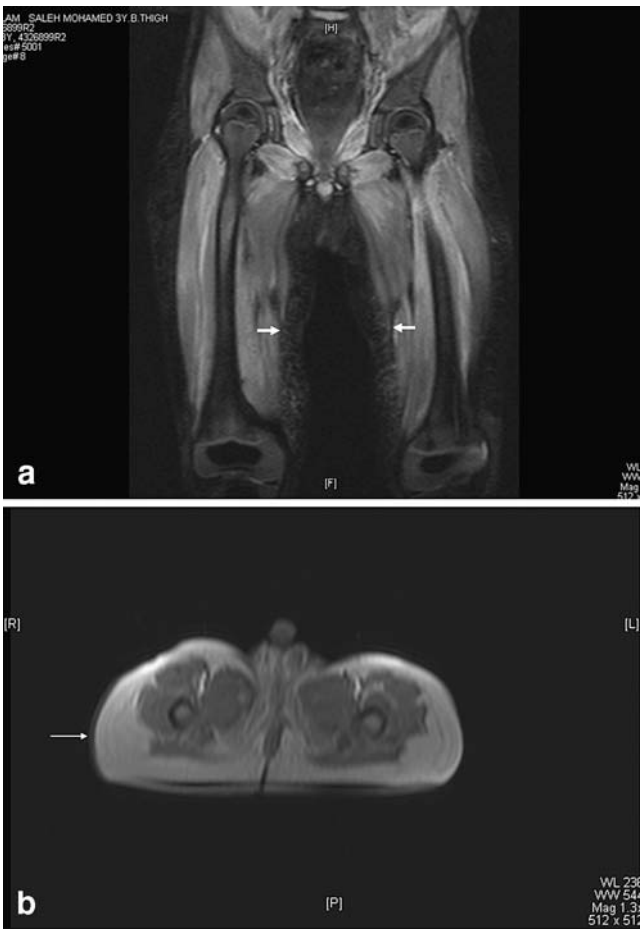

Fig. 3 Coronal (**a**) and axial sections (**b**) of magnetic resonance imaging (MRI) shows: (**a**) diffuse marked subcutaneous edema (*white arrows*) is present and hyperintense signal in T1W1 in obturator internus, the insertion of ileopsoas, and the anterior group of muscles of the upper and middle thirds of both thighs on coronal section; (**b**) subcutaneous edema is diffuse (*white arrow*), but prominent laterally and posteriorly

extremities. Magnetic resonance imaging (MRI) of the lower extremities and pelvis showed marked diffuse edema in the subcutaneous tissue and muscles (Fig. 3a,b).

The diagnosis of JDM was based on Bohan and Peter's criteria [1] based on: characteristic rash and three of the following criteria: symmetric proximal muscle weakness, elevated muscle-derived enzymes, and EMG findings of inflammatory myopathy.

The inpatient treatment continued for 5 weeks and consisted of: weekly pulse methylprednisolone (30 mg/kg/dose) every other day, alternating with oral prednisolone (2 mg/kg/day), intravenous immunoglobulin (IVIG) was given in the third week of admission (400 mg/kg/dose) for five consecutive days; no significant clinical improvement was observed, but the skin rash completely faded out.

The patient was discharged on a nasogastric feeding, rehabilitation program, with daily oral prednisolone (2 mg/kg/day) and azathioprine (2 mg/kg/day) for two weeks. On the eighth week from the diagnosis, clinical improvement was observed in the form of improved gastrointestinal dysmotility symptoms, partial regain of the motor power of the extremities, and the significant decrement of muscle enzymes (total creatine kinase 270 IU/L, creatine kinase MM 194 IU/L). Steroid therapy was tapered by 0.5 mg/kg/month. On the tenth week from the diagnosis, the patient started full oral feeding alongside a steroid withdrawal regimen and rehabilitation program. Drug therapy is planned to be completed in two years, with monthly follow up of the patient to reach the least steroid dose that maintains control.

Discussion

Heliotrope rash, symmetrical proximal muscle weakness, elevated serum muscle enzymes, and typical findings of inflammatory myopathy by electromyogram in our case were compatible with JDM. In our case, generalized pitting edema was the presenting feature misdiagnosed as nephrotic syndrome and later muscle weakness and skin lesions evolved. The absence of clinical features and laboratory parameters that might be associated with generalized edema, such as nephrotic syndrome, hypothyroidism, cardiac failure, liver disease, and malignancy strongly suggested that the anasarca, in this case, was due to dermatomyositis. The rapid improvement of generalized edema over the course of prednisolone treatment without diuretics strongly supported our speculation.

Generalized edema in association with JDM is infrequently reported [9]. A review of the English literature available revealed only 21 children reported with JDM presented by generalized anasarca (Table 2). Such patients respond poorly to oral steroid therapy. The exact mechanism of generalized edema in JDM in not known. Autoantibodies are believed to play an important role in the pathogenesis. The pathological hallmark is an immune complex vasculitis—the damage to the endothelium caused by complement C5b-9 attack complex [3, 11]. It is suggested that, as a sequel to diffuse and widespread capillary endothelial damage, there is an increase in capillary permeability in muscles, leading to generalized edema [8].

Table 2 The clinical features of 22 patients with juvenile dermatomyositis (JDM) presenting with anasarca

Reference	Age (years)/sex	Severe weakness	Myalgia	Dysphonia	Dysphagia	Calcinosis	Response to steroids
Our case	3.5/M	+/BB	+	+	+	–	Poor
[4]	3.6/M	+ (function NA)	+	+	+	–	– (Corticotropin, testosterone)
[5]	2.2/1M, 1/F	2/2, 2/2 BB	1+, 1 NA	NA	NA	2/2	NA
[6]	14/M	+	+	–	–	–	Good
[7]	10/F	+, BB	+	NA	+	+	NA
[8]	8/F	+/BB	+	–	–	–	Initially good
[9]	7/F	+, BB/WC	+	+	+	–	–
[10]*	6/M	–	+	–	–	–	Good
[12]	6/F	+, BB	+	NA	NA	–	NA
[13]	7.1/F	+, BB	+	+	+	NA	NA
[14]**	17/M	+, BB	+	+	+	–	NA
[15]	3.5/F	+, BB	+	NA	NA	–	NA
[16]***	Mean 7.5 (2.3–10.6) 5M, 4F	9/9 (function NA)	8/9	NA	5/9	4/9	1/4+, 5/9 NA (cortisone, corticotropin)

BB=bed-bound; WC=wheel-chair-bound; NA=not available
*This case of JDM was associated with hereditary angioneurotic oedema
**This case of JDM was associated with a mediastinal tumor
***In this series, 9 of 26 cases of JDM presented with generalized edema

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