Rheumatol Int (2009) 29:565–567
DOI 10.1007/s00296-008-0702-9

CASE REPORT

Anasarca as the presenting manifestation of parvovirus B19 associated juvenile dermatomyositis

Shanmuganathan Chandrakasan · Surjit Singh · R. K. Ratho · Sharat Kumar · B. Mishra

Received: 7 March 2008 / Accepted: 7 September 2008 / Published online: 20 September 2008

Abstract Anasarca as the presenting manifestation of juvenile dermatomyositis (JDMS) is extremely rare. We report a case of a 4-year-old boy who was initially managed for nephrotic syndrome in view of anasarca and mild hypoalbuminemia. Later, at presentation to our institute, a diagnosis of severe edematous JDMS was made in view of associated profound muscle weakness and characteristic skin changes. The child responded to aggressive immunosuppressive therapy. On further evaluation, he had evidence of acute parvovirus B19 infection. Our case illustrates anasarca as an uncommon severe manifestation of JDMS and the possible role of parvovirus B19 in the onset of this autoimmune disorder.

Keywords Juvenile dermatomyositis · Anasarca · Parvovirus B19

Introduction

Juvenile dermatomyositis (JDMS) is a rare autoimmune systemic vasculopathy of childhood commonly affecting the muscles, skin and gastrointestinal tract. It classically presents as a weakness of proximal muscles, heliotrope rash and Gottron's papules [1]. Severe disease is characterized by marked muscle weakness with pharyngeal and laryngeal involvement, widespread skin changes, and gastrointestinal ulcerations [1]. Along with weakness in the affected muscles there can be a variable extent of nonpitting indurated edema of the affected limbs. Anasarca in association with JDMS is extremely rare [2, 3]. In most of the cases the edema is localized and restricted to the affected limbs. Though parvovirus B19 has been associated in cases of JDMS, its role in the pathogenesis of the disease is still not clear [4, 5]. We report the case of a child here with severe acute edematous JDMS with anasarca as the presenting manifestation, who on further evaluation was found to have evidence of parvovirus B19 infection.

S. Chandrakasan · S. Singh (✉) · R. K. Ratho · S. Kumar · B. Mishra
Postgraduate Institute of Medical Education and Research, Chandigarh, India
e-mail: surjitsinghpgi@rediffmail.com

Case report

A 4-year-old boy presented to us with complaints of fever, myalgia, profound muscle weakness, anasarca and nasal regurgitation of feeds. He had had a respiratory catarrh with flushed face and generalized erythematous rash 6 weeks ago. Following the prodromal symptoms he developed progressive muscle weakness. Over the next 2 weeks the weakness gradually progressed and he developed nasal regurgitation of feeds and dysphonia. By the end of the third week, he had developed anasarca with no associated renal or cardiac symptoms. The child was initially managed as a possible case of nephrotic syndrome. However, as the investigative workup was not corroborative and as the clinical condition was worsening, he was referred to our institute.

On examination the child had generalized, nonpitting edema with marked tenderness (Fig. 1). Neurological examination showed muscle weakness with power of grade 2/5 in all proximal muscles, inability to raise head against gravity, weak gag and cough reflex. Deep tendon reflexes were well preserved with normal sensory examination. No significant lymphadenopathy or organomegaly was noticed. He had telangiectasia in the upper eyelids, without any

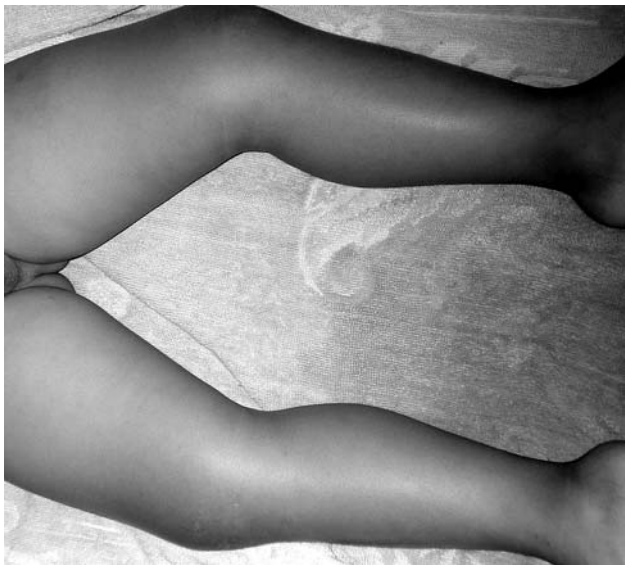


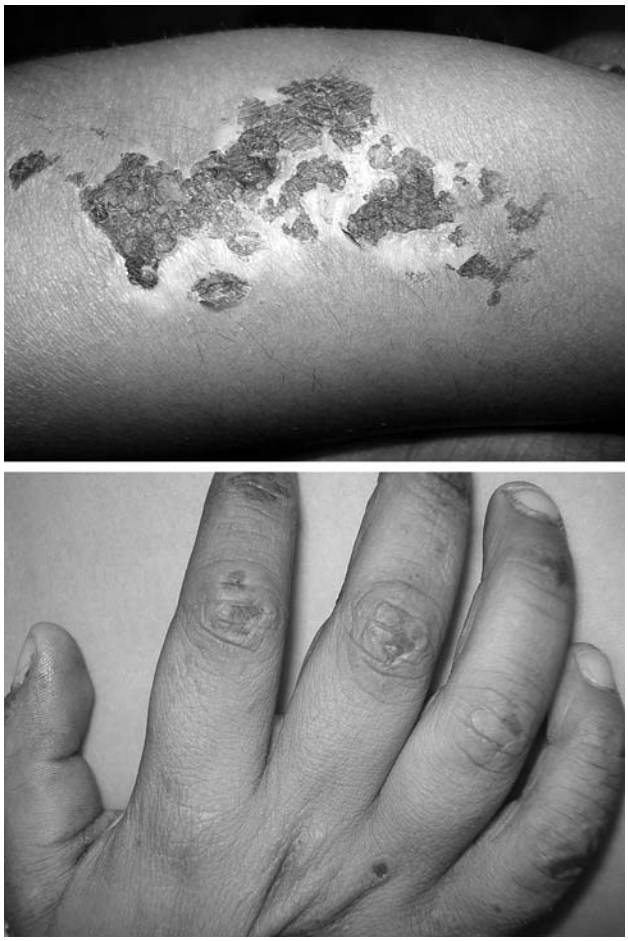
Fig. 1 Generalised edema involving the lowerlimbs

periorbital erythema or edema. He also had Gottron's papules and ulceraration over the elbow, dorsal interphalangeal and metacarpophalangeal joints (Figs. 2, 3).

Investigations showed hemoglobin of 108 g/L, polymorphonuclear leucocytosis ($21.4 \times 10^9\ L^{-1}$ with polymorphs of 72%) and thrombocytosis ($867 \times 10^9\ L^{-1}$). Erythrocyte sedimentation rate was 47 mm in the first hour with a significant increase in levels of C reactive protein of 13.4 mg/dl (normal < 6 mg/dl). Muscle enzymes assayed, showed creatine kinase level of 1,108 U/L (normal range 55–170 U/L), aspartate aminotransferase 514 U/L (normal 17–59 U/L), alanine aminotransferase 187 U/L (normal 21–72 U/L), aldolase 4.8 U/L (normal 1.0–3.5 U/L) and serum albumin of 25 g/L. Significant cardiac, renal and hepatic causes of anasarca were ruled out by appropriate investigations.

Magnetic resonance imaging of the thigh done during the second week of hospital stay showed symmetrical bilateral increased signal intensity in thigh muscles on T2 W IR images with predominant involvement of quadriceps femoris and muscles of adductor region. There were no localized collections in soft tissues of thigh. Electromyography done on bilateral deltoid and vastus lateralis muscles showed polyphasic pattern with decreased amplitude and duration, typical of myopathic pattern. Spontaneous and insertional fibrillations and high-frequency repetitive discharges were also noted.

Tests for antinuclear antibody and rheumatoid factor (RF) were noncontributory, and complement levels (C3 and C4) were within normal limits. Tests for streptococcus (antistreptolysin O titer, throat swab culture), toxoplasma (IgM, IgG ELISA), Epstein–Barr virus (IgM VCA), and cytomegalovirus (IgM ELISA and pp65 antigen) infection were negative. Both parvovirus B19 IgM serology (IBL, Hamburg, Germany) and PCR for parvovirus B19 DNA



Figs. 2, 3 Healing superficial ulcers and Gottron's papules over the elbow, dorsal interphalangeal and metacarpophalangeal joints, with wrinkling of skin seen at the resolution of edema

were positive, and IgG serology was negative in the blood sample taken during disease activity.

Based on the clinical examination and investigations, a diagnosis of acute edematous JDMS was made and he was started on pulse methylprednisolone of 30 mg/kg/day for 5 days followed by oral prednisolone of 2 mg/kg/day, and weekly oral methotrexate of 15 mg/m^2. He was initially given nasogastric feeds and these had to be continued for 4 weeks because of significant pharyngeal and laryngeal muscle weakness. Antimicrobials were added in view of a clinical suspicion of aspiration pneumonia. The weakness and edema improved gradually and by 2 weeks he was able to raise the limbs against gravity and hold his head, but his gag was still weak. By 6 months after discharge there was no edema or weakness, he was able to walk without support and had good gag and cough reflexes. His muscle enzymes and hematological parameters were now within normal limits, and repeat parvovirus B19 IgM serology and parvovirus B19 DNA PCR were negative and IgG serology was positive.

Discussion

Juvenile dermatomyositis presenting as anasarca is a rare clinical presentation and only a few cases have been reported in recent literature [2, 3]. The presence of anasarca in cases of JDMS has been suggested as a clinical marker of severe disease activity [2]. Marked muscle weakness, cutaneous ulcers, laryngeal and pharyngeal muscle involvement suggested severe disease activity in our case. In most cases of JDMS and other inflammatory myopathies, anasarca was due to subcutaneous edema and edema of the involved muscles [2, 6, 7]. Though the exact pathophysiology of anasarca is not known, it is believed to be due to severe systemic vasculopathy, leading to capillary damage and increased permeability [2]. In the absence of significant renal, hepatic or cardiac dysfunction, mild hypoalbuminemia may have had a role to play in the pathogenesis of anasarca in our case. As anasarca is otherwise a relatively common clinical finding in pediatric practice, this presentation of JDMS is liable to be confused with a primary renal or cardiac disease if the clinician is not careful. However, the profound muscle weakness and characteristic skin changes are usually clinical giveaways.

Both epidemiological and laboratory data suggest the pathogenesis of JDMS is infection-driven [8, 9]. Many viral and bacterial antigens have been identified as important triggers in the pathogenesis of JDMS [1], but the cause and effect relationship is still not clear [4, 5]. Parvovirus B19 has been associated with manifestations ranging from myositis [10], myofasciitis [11] and dermatomyositis [4]. Lewkonia et al. [4] first reported the association of parvovirus B19 with the onset of JDMS. Similarly, Chevrel et al. [12] reported the presence of parvovirus DNA in the muscle biopsy specimens of patients of dermatomyositis. It appears, therefore, that the occurrence of parvovirus infection in our patient may not have been entirely coincidental.

Both the cases of parvovirus-associated myositis [10] and myofasciitis [11] reported earlier had a self-limiting course and recovered with supportive therapy. However, in our case the severity of the disease mandated aggressive immunosuppressive therapy, and this resulted in prompt clinical improvement. The child remains well on follow-up.

References

1. Cassidy JT, Lindsley CB. Juvenile dermatomyositis. In: Cassidy JT, Lindsley CB (eds) Textbook of pediatric rheumatology. Elsevier Saunders, Philadelphia, pp 407–434
2. Mitchell JP, Dennis GJ, Rider LG (2001) Juvenile dermatomyositis presenting with anasarca: a possible indicator of severe disease activity. J Pediatr 138:942–945
3. Mehndiratta S, Banerjee P (2004) Juvenile dermatomyositis presenting with anasarca. Indian Pediatr 41:752–753
4. Lewkonia RM, Horne D, Dawood MR (1995) Juvenile dermatomyositis in a child infected with human parvovirus b19. Clin Infect Dis 21:430–432
5. Mamyrova G, Rider LG, Haagenson L, Wong S, Brown KE (2005) Parvovirus b19 and onset of juvenile dermatomyositis. JAMA 294:2170–2171
6. Smyth AE, Bell AL, Crone M (2000) Acute oedematous dermatomyositis. Ann Rheum Dis 59:575
7. Gorelik O, Almoznino-Sarafian D, Alon I (2001) Acute inflammatory myopathy with severe subcutaneous edema, a new variant? Report of two cases and review of the literature. Rheumatol Int 20:163–166
8. Pachman LM, Lipton R, Ramsey-Goldman R (2005) History of infection before the onset of juvenile dermatomyositis: Results from the national institute of arthritis and musculoskeletal and skin diseases research registry. Arthritis Rheum 53:166–172
9. Tezak Z, Hoffman EP, Lutz JL (2002) Gene expression profiling in dqa1*0501+ children with untreated dermatomyositis: a novel model of pathogenesis. J Immunol 168:4154–4163
10. Kishore J, Singh J (2006) Detection of parvovirus b19 in a case of erythema infectiosum with myositis. Indian Pediatr 43:814–817
11. Ichinose K, Migita K, Sekita T (2004) Parvovirus b19 infection and myofasciitis. Clin Rheumatol 23:184–185
12. Chevrel G, Calvet A, Belin V, Miossec P (2000) Dermatomyositis associated with the presence of parvovirus b19 DNA in muscle. Rheumatol (Oxf) 39:1037–1039