ELSEVIER

Canadian Association of Radiologists Journal 60 (2009) 50–52

CANADIAN ASSOCIATION OF RADIOLOGISTS JOURNAL

www.carjonline.org

Canadian Residents' Corner / Coin canadien des résidents en radiology

Answer to Case of the Month #144 Juvenile Dermatomyositis

Ryan F. MacDougall, MD[a], Suzanne E. Ramsey, MD, FRCPC[b], Matthias H. Schmidt, MD, MSc, FRCPC[a,*]

[a]*Department of Radiology, Dalhousie University, QEII Health Sciences Centre, Halifax, Nova Scotia, Canada*
[b]*Division of Rheumatology, Department of Pediatrics, IWK Health Centre, Halifax, Nova Scotia, Canada*

Clinical Presentation

A previously healthy 5-year-old boy presented with a low-grade fever, migratory myalgia, and progressive proximal muscle weakness over a period of 1 month. Physical examination revealed severe, symmetric weakness of the trunk and proximal extremities, multiple joint effusions, and a heliotrope rash over the right upper eyelid. The white blood cell count and erythrocyte sedimentation rate were normal. The serum creatine kinase level was increased at 7,900 IU/L (normal, <160 IU/L). A magnetic resonance imaging (MRI) scan was performed with sedation, and selected images are provided (Figures 1 and 2).

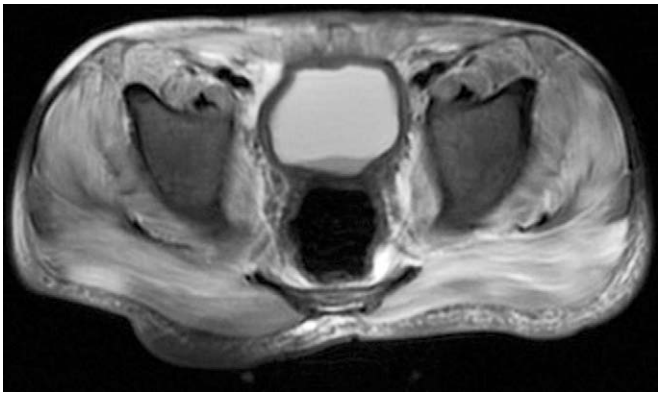

Figure 1. Axial T2-weighted image with fat saturation shows diffuse oedema of pelvic girdle muscles. There is extensive fascial oedema and dermal thickening with subcutaneous oedema.

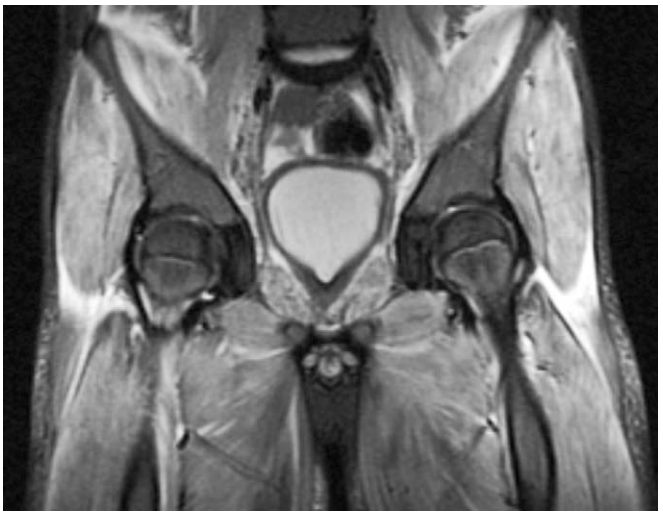

Figure 2. Coronal short tau inversion recovery image shows oedema extending into the proximal thigh muscle groups. Again, there is extensive fascial and subcutaneous oedema.

Key Words: Dermatomyositis; Magnetic resonance imaging; Child; Preschool.

* Address for correspondence: Matthias H. Schmidt, MD, MSc, Department of Radiology, Dalhousie University, QEII Health Sciences Centre, 1278 Tower Road, Halifax, Nova Scotia B3H 2Y9, Canada.

E-mail address: mhschmid@dal.ca (M.H. Schmidt).

0846-5371/$ - see front matter
doi:10.1016/j.carj.2009.02.013

Diagnosis

Juvenile dermatomyositis (JDM).

Radiologic Findings

An axial T2-weighted image with fat saturation (Figure 1) and a coronal short tau inversion recovery image (Figure 2) of the pelvis show similar findings. All of the visualized muscle groups of the pelvic girdle and proximal thighs show diffuse hyperintensity. There is extensive fascial oedema and subcutaneous oedema. These imaging findings, combined with the patient's age, heliotrope rash, rapidly progressive and symmetric proximal muscle weakness, and increased serum creatine kinase level, are consistent with the diagnosis of JDM.

Discussion

The inflammatory myopathies comprise 3 distinct disease entities: dermatomyositis (DM), polymyositis (PM), and sporadic inclusion-body myositis [1]. In all age groups, DM is more common than PM [1]; PM and inclusion-body myositis rarely are seen in children [1,2]. Based on the most recent studies, the incidence of JDM in Great Britain and the United States ranges from 2 to 3 children per million per year [2]. DM is a systemic disease, with inflammation often extending beyond skin and muscles [1,2]. This is apparent more readily with JDM, compared with adult DM [1,2].

The seminal articles by Bohan and Peter [3,4], written more than 30 years ago, detailed the currently accepted criteria for the diagnosis and classification of DM and PM. The diagnosis of DM requires the presence of a characteristic skin rash (heliotrope rash and Gottron's papules), along with at least 3 of the following findings: symmetric proximal muscle weakness progressing over weeks to months, increase in serum skeletal muscle enzyme levels, myopathic findings on electromyography, and typical findings on muscle biopsy, including degenerating and regenerating fibers as well as perivascular inflammation [3,4]. As noted earlier, JDM is a systemic inflammatory disorder with a wide range of clinical manifestations, including such signs and symptoms as muscle pain (73%), fever (65%), dysphagia (44%), hoarseness (43%), abdominal pain (37%), arthritis (35%), soft-tissue calcifications (23%), and melena (13%), in addition to rash and weakness [5]. The incidence of skin ulcerations, calcinosis, and vasculitis is greater in JDM, compared with adult DM [2]. Most importantly, associated malignancies in children with JDM have been reported rarely [6]. This is unlike adult DM, which has a 20% to 25% rate of associated neoplasms and is well recognized as a paraneoplastic syndrome [6].

The diagnosis of JDM can be made with high sensitivity and specificity (both >92%), using the criteria of Bohan and Peter [2]. However, many pediatric rheumatologists are reluctant to perform electromyography or muscle biopsy because of the invasive nature of these tests and the high false-negative rate of muscle biopsy early in the disease [2]. Even when muscle biopsy is undertaken, patchy disease can cause falsely negative results owing to sampling errors [2]. Therefore, the full diagnostic criteria often are not met. It has been suggested recently that magnetic resonance imaging (MRI) should be added to the list of diagnostic criteria for JDM [2]. A recent international consensus survey of the diagnostic criteria for JDM rated typical findings on MRI, along with muscle biopsy and electromyography, as the most useful diagnostic criteria after rash, proximal muscle weakness, and increased serum skeletal muscle enzyme levels [7]. Indeed, some investigators have proposed that MRI potentially could replace muscle biopsy in confirming the suspected diagnosis of JDM in many patients [2]. Certainly, MRI can guide the biopsy and thus increase the diagnostic yield by showing inflamed muscle groups [2,8].

The advantages of MRI in the diagnosis and definition of DM relate primarily to its excellent soft-tissue discrimination and high sensitivity for inflammation [8]. This is enhanced further by various fat-suppression techniques [8]. Fat-suppressed T2-weighted sequences and short tau inversion recovery sequences characteristically show a multifocal or diffuse pattern of muscle hyperintensity, most marked in the proximal limbs [8]. Dermal thickening and a reticular pattern of subcutaneous oedema are seen commonly in areas overlying inflamed muscle groups [8]. Edema can spread along myofascial planes [8]. Mildly prominent lymph nodes also are seen. Arthritis and panniculitis, if present, can be shown. The MRI findings of DM at presentation may be mimicked by infectious myofasciitis [8]. However, the clinical presentation of DM differs significantly from that of infectious myofasciitis. Muscle atrophy and fatty infiltration occur after a long course of the disease and appear on MRI as decreased muscle bulk and increased signal intensity, between muscle bundles, on both T1- and T2-weighted images without fat suppression [9]. This appearance is nonspecific and difficult to distinguish from steroid-induced muscle atrophy.

The mainstay of treatment for JDM is prednisone [1,2]. Second-line therapy involves immunosuppressive agents (eg, methotrexate, cyclosporine) [1,2]. Other treatments showing possible benefit include intravenous immunoglobulin therapy and immunomodulatory therapies [1,2]. The prognosis of children with JDM has improved steadily over the past several decades [10]. In a cohort of 65 children from 4 Canadian centers, diagnosed between 1984 and 1995, there was 1 death [10]. Seventy-two percent of the survivors reported no disability or minimal disability, and of the 28% with more than minimal disability, 8% had moderate-to-severe disability [10]. Sixty-three percent of the children had relapsing/remitting or chronic continuous disease, and 37% had a single disease episode [10]. None of the children in this cohort developed a malignancy over a follow-up period ranging from 3 to 14 years [10]. Conventional MRI can be used to assess treatment response by tracking the disappearance of oedema [8]. Experimental techniques such as phosphorus-31 magnetic resonance spectroscopy and quantitative assessment of T2 relaxation time have shown promise in the objective assessment of disease activity [11,12].

In summary, JDM is a systemic inflammatory condition that primarily involves skin and muscles, but frequently involves other tissues as well. Unlike adult DM, which has a high association with malignancy, JDM is typically not a paraneoplastic condition. MRI is useful in the diagnosis and follow-up evaluation of JDM, and it can guide muscle biopsy. MRI, showing characteristic findings of patchy or diffuse muscle T2 hyperintensity and fascial and subcutaneous oedema, is likely to become an increasingly important part of the diagnostic algorithm for children with suspected JDM and may entirely replace more invasive tests in the future.

References

[1] Dalakas MC, Hohlfeld R. Polymyositis and dermatomyositis. Lancet 2003;362:971–82.
[2] Wedderburn LR, Li CKC. Paediatric idiopathic inflammatory muscle disease. Best Pract Res Clin Rheumatol 2004;18:345–58.
[3] Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). N Engl J Med 1975;292:344–7.
[4] Bohan A, Peter JB. Polymyositis and dermatomyositis (second of two parts). N Engl J Med 1975;292:403–7.
[5] Pachman LM, Hayford JR, Chung A, et al. Juvenile dermatomyositis at diagnosis: clinical characteristics of 79 children. J Rheumatol 1998;25:1198–204.
[6] Callen JP. Dermatomyositis. Lancet 2000;355:53–7.
[7] Brown VE, Pilkington CA, Feldman BM, et al, on behalf of the Network for Juvenile Dermatomyositis, a working party of the Paediatric Rheumatology European Society (PreS). An international consensus survey of the diagnostic criteria for juvenile dermatomyositis (JDM). Rheumatology 2006;45:990–3.
[8] Garcia J. MRI in inflammatory myopathies. Skeletal Radiol 2000;29:425–38.
[9] Mastaglia FL, Garlepp MJ, Phillips BA, et al. Inflammatory myopathies: clinical, diagnostic and therapeutic aspects. Muscle Nerve 2003;27:407–25.
[10] Huber AM, Lang B, LeBlanc CMA, et al. Medium- and long-term functional outcomes in a multicenter cohort of children with juvenile dermatomyositis. Arthritis Rheum 2000;43:541–9.
[11] Park JH, Olsen NJ. Utility of magnetic resonance imaging in the evaluation of patients with inflammatory myopathies. Curr Rheumatol Rep 2001;3:334–45.
[12] Maillard SM, Jones R, Owens C, et al. Quantitative assessment of MRI T_2 relaxation time of thigh muscles in juvenile dermatomyositis. Rheumatology 2004;43:603–8.