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Clinical Observations

Recurrent Focal Myositis in Childhood: A Case Report and Systematic Review of the Literature

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ABSTRACT

BACKGROUND: Recurrent focal myositis in adulthood has been documented in case reports and case series. Existing textbooks and reviews do not mention or mention only in passing this entity in childhood. We present a patient with recurrent focal myositis and summarize available clinical, laboratory, management, and outcome data on this entity in the pediatric ages. **METHOD:** We describe a nine-year-old patient with recurrent myositis of the left biceps. The terms "myositis" and "relapsing" or "recurrent" or "recurrence" were searched using the United States National Library of Medicine and the Excerpta Medica Database. Pertinent secondary references were also screened. **RESULTS:** Another seven pediatric patients (five males and two females, median age ten years, interquartile range 7-14 years) with recurrent focal myositis were identified. In children, the calf was the most frequently involved muscle. Unlike adults, the myositis in children was usually painful. Episodes could be associated with normal or elevated erythrocyte sedimentation rate and blood levels of C-reactive protein, creatine kinase, and aspartate aminotransferase. Abnormalities of the creatine kinase value did not seem to be associated with a higher risk of recurrences. **CONCLUSIONS:** Focal myositis has a favorable outcome in children. Recurrent focal myositis is rare and usually benign in childhood. More data are needed to improve the understanding of this condition.

Keywords: localized myositis, muscle inflammation, pediatric, creatine kinase, inflammation, relapsing myositis

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Introduction

Focal myositis, first described by Heffner et al.,[1] is a rare entity characterized by acute onset of localized skeletal muscle inflammation of unknown origin. It usually affects only one muscle and presents a benign course without sequelae or recurrence.[2] However, adults with relapsing focal myositis have been documented.[2-5] Existing textbooks and reviews do not mention or only briefly mention forms of recurrent focal myositis in childhood. Here we describe a child with recurrent focal myositis and review the available literature to summarize clinical, laboratory, management, and outcome data of recurrent focal myositis in children.

Patient Description

This nine-year-old child was evaluated for a two-day history of swelling and increasing pain of the upper left arm. He did not claim any prolonged immobility, recent surgery, trauma, fever, rash, or drug intake during the previous month. Moreover, no personal or family history of neuromuscular, autoimmune, or connective tissue disease was reported.

Physical examination disclosed a tender, warm, and painful mass on the lateral-anterior part of the left upper arm. Movements of extension and flexion of the affected arm were limited because of the pain. The rest of his examination was otherwise unremarkable.

Initial investigations, including hemoglobin, hematocrit, white cell and platelet counts, coagulation panel, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP), were normal. Creatine kinase (CK) was 8484 U/L (reference less than 247 U/L); lactate dehydrogenase, 640 U/L (reference less than 300 U/L); aspartate aminotransferase (AST), 226

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U/L (reference less than 47 U/L); and alanine aminotransferase, 73 U/L (reference less than 39 U/L). Urinalysis was normal.

Doppler sonography of the left arm did not show any blood flow alterations, and ultrasonic imaging revealed increased echogenicity in the subcutaneous tissue and marked enlargement of the biceps. History, physical examination, and diagnostics were considered consistent with a focal myositis. An analgesic treatment was provided, and his symptoms resolved in three days. CK normalized within a week (236 U/L), as did all the remaining blood examinations.

One month later, he developed recurrent swelling and mild pain of the left arm. He had engaged in a prolonged intense sport activity in the days before, but he denied any trauma. Physical examination disclosed the same features of the previous episode except that no motion limitation was observed.

Laboratory values demonstrated only a new increase in CK (1736 U/L) and AST (62 U/L) blood levels. A subsequent diagnostic evaluation failed to reveal a recent or concurrent infectious disease (IgM for influenza A and B, parainfluenza, Coxsackie, Epstein-Barr, herpes simplex, and varicella-zoster viruses, adenovirus, echovirus, cytomegalovirus, and measles virus were within the normal range). Autoimmune screening, including anti-extractable nuclear antigen antibodies, ANCA (anti-neutrophil cytoplasmic antibodies), anti-nuclear antibodies, anti-mitochondrial antibodies, anti-smooth muscle antibodies, and endomysial antibodies, was unremarkable.

Ultrasound and Doppler flow scanning studies showed features similar to those of the previous episode. Magnetic resonance imaging of the arm revealed focal areas of increased signal intensity on T2-weighted images over the lateral and anterior margin of the left biceps. The signal enhancement suggested inflammatory edema within the muscle tissue. The clinical picture was consistent with a relapsing of focal myositis. In three days, all symptoms spontaneously resolved, and both CK (229 U/L) and blood examinations normalized in five days. The child was asked to avoid intense sport activity for three months. In the 18-month follow-up period he did not report any further episode of myositis.

Review of the literature

Study selection

Between September 2015 and January 2016, two investigators (G.P.M. and H.G.) searched the terms “myositis” and “relapsing” or “recurrent” or “recurrence” using the United States National Library of Medicine and the Excerpta Medica Database. Pertaining secondary references were also screened. For this study, we used the principles underlying the UK Economic and Social Research Council guidance on the conduct of narrative synthesis and the Preferred Reporting Items for Systematic Reviews and Meta-Analyses statement.[6] The two investigators independently screened all study titles and abstracts, and full-text articles of any potentially relevant study were retrieved. For the final analysis, they selected peer-reviewed reports published as full-length articles or letters with no limits of date. Studies reporting pediatric patients (aged $<$ 18 years at the first episode of myositis) with at least two episodes of focal myositis were retained. A diagnosis of focal myositis was reviewed using the following criteria: (1) history and clinical features consistent with a nonsuppurative localized myositis; (2) imaging studies that confirm the diagnosis; (3) spontaneous resolution or resolution with anti-inflammatory or steroids treatment within 4 weeks or histology specimen consistent with the diagnosis.[7,8] Reports published in languages other than English, Italian, Dutch, German, Portuguese, and Spanish were not considered. Reports of myositis following a trauma, pyomyositis, myositis ossificans, eosinophilic myositis, dermatomyositis, connective tissue disease, and polymyositis were also excluded.

Data extraction and analysis

From each report dealing with recurrent focal myositis, the same two investigators excerpted data on sex, age, prodromes, clinical presentation, laboratory investigation, management of the first episode, and the localization, management, follow-up, and outcome of the second episode. If data were not available, authors of any identified studies were tentatively contacted via e-mail to obtain the lacking information. Discrepancies in data interpretation were resolved by involving a third researcher (E.F.F.). The Cohen k coefficient for inter-rater reliability was used to assess agreement on article selection. Numerical data were presented as median and interquartile range.

Results

Search results

The Cohen’s k coefficient for study eligibility was 0.94, indicating an excellent agreement on study selection between the two investigators. A total of seven reports were included following the literature search process presented in Figure 1 (Supplementary file). All reports were published in English. These studies were carried out in the following countries: the United Kingdom (N = 2), Australia (N = 1), Brazil (N = 1), Japan (N = 1), Spain (N = 1), and the United States of America (N = 1).

Characteristics of the patients with recurrent focal myositis

The aforementioned reports including seven patients with focal recurrent myositis (five males and two females, median age ten years, interquartile range 7-14 years) are listed in Table 1.[5,9-14] Clinical data and localization of focal myositis are summarized in Table 2. The calf was involved in four of seven patients both at the first and at the relapsing episode of focal myositis. In only one child, the recurrence affected a different muscle (psoas) from the first episode (deltoid). The second episode of myositis followed after a period ranging from one to 36 months (median 3.5 months).

In two patients, myositis episodes were preceded by acute infections and in one by bacillus Calmette-Guérin (BCG) vaccination.[9,12,14] In four patients, episodes were not preceded by any prodromal episode.

The levels of CK were found to be elevated in two patients (12,880 and 633 U/L)[9,12] and within normal ranges in three patients.[5,10,11] In two patients, the CK value at the first episode was not listed.[13,14]

Further laboratory abnormalities were the following: AST was higher in two subjects (485 and 48 U/L)[9,12] and in one of them CRP also was slightly elevated[12]; ESR was increased in another patient (50 mm/h).[11] In no instances were autoantibodies positive. In three patients a muscle biopsy showed signs of inflammatory cell infiltrations without perivascular or vascular alterations.[9-11]