Benign acute childhood myositis in an Accident and Emergency setting

L M Rennie, N F Hallam, T F Beattie

Four children presented to the Accident and Emergency department of the Royal Hospital for Sick Children in Edinburgh during seven days in February 2003. They shared a strikingly similar clinical picture with debilitating muscle pain in their calves. This paper discusses their clinical course and presents a literature review of the problem.

CASE 1
A 5 year old girl presented with a two day history of fever and headache, followed by a 24 hour history of painful calves. She had first experienced the pain when she got out of bed and had progressive difficulty walking. On examination she was apyrexial, coryzal, and had palpable cervical lymph nodes. She was walking with an antalgic gait bilaterally. Both calves were tender but no swelling, erythema, or wasting was noted. Her feet were unusually cold but capillary return was less than two seconds.

Her urine dipstick was normal. She had a mild leucopenia but platelets and erythrocyte sedimentation rate (ESR) were normal. Her creatinine phosphokinase (CPK) was impressively raised (see table 1).

She was discharged with dipsticks to check her urine and became asymptomatic within 24 hours. When she was seen in the review clinic two weeks later her CPK had returned to normal. Her convalescent serum did not show evidence of recent influenza A, influenza B, adenovirus, RSV, CMV, EBV, or mycoplasma infections. On examination he was apyrexial and had no palpable lymph nodes. He had tender calves and was walking with extended legs and a broad based gait. His neurological examination revealed no evidence of pathology. His feet were cold but capillary return was less than two seconds.

Again he had mild leucopenia, borderline low platelets, but a normal ESR. His CPK was very elevated (see table 1).

He was asymptomatic when seen in clinic two weeks later and his CPK had returned to normal. His acute and convalescent sera suggested a RSV infection in the recent past. However there was no evidence of recent influenza A, influenza B, adenovirus, CMV, EBV, or mycoplasma infections.

CASE 4
Four days later a 9 year old boy presented with what had been described as a “flu-like” illness with headache, vomiting, abdominal pain, and diarrhoea. On recovery from this he developed pain in both calves. The pain had been worse on awakening the day before attendance at hospital. On examination he was apyrexial. His calves were tender on palpation and on stretching. There was no swelling or erythema.

He had a leucopenia with low platelets but his ESR was normal. He had a moderately raised CPK (see table 1). His urine dipstick was negative for blood but positive for protein.

He was seen a week later in the review clinic and was asymptomatic. His blood results had returned to normal. His convalescent serum suggested a recent influenza B infection. There was no evidence of recent influenza A, RSV, CMV, EBV, or mycoplasma infections.

Abbreviations: BACM, benign acute childhood myositis; CMV, cytomegalovirus; CPK, creatinine phosphokinase; EBV, Epstein-Barr virus; ESR, erythrocyte sedimentation rate; RSV, respiratory syncitial virus.
DISCUSSION

Many patients complain of muscle pain in association with a viral illness. However, in 1957 Lundberg published the first report of "myalgia cruris epidemica" which seemed to be a newly identified syndrome of muscle pain, predominantly affecting the calves and occurring in school age children. The condition followed a prodrome of febrile illness, striking in a week. Lundberg felt that in view of the well described activity before the onset of pain. All children recovered within the first few months of the year. The patients were typically children and it was noted that:

- The onset of pain tended to occur after a period of rest— that is, on awakening.2–4
- The vast majority of children tested had a raised CPK.2–10 In many cases this was massively elevated, however none of the children tested had evidence of rhabdomyolysis—that is, myoglobin in urine.
- Other less consistent laboratory findings are leucopenia,13–19 thrombocytopenia,2 and elevated serum glutamic oxaloacetic transaminase (SGOT).4–8
- Neurology is reported as normal2–4 8 and it has been suggested that mild calf weakness is due to muscle pain rather than true inability of muscle to generate power.9
- Muscle studies have been performed relatively infrequently in view of the short duration of symptoms and the benign prognosis. When electromyograms have been recorded they have either been normal10 or suggested patchy myopathic changes.6–7 The few muscles biopsies taken have been reported as normal,16 or said to demonstrate myositis,11 segmental rhabdomyolysis,4–9 or moderate muscle necrosis with interstitial inflammation.7

Various descriptions have been given to the disorder and despite some debate about the actual muscle pathology, the term myositis seems to be generally used. This is a reflection of the assumption that there must be at least an element of muscle inflammation to account for the release of CPK. Many authors have termed the syndrome benign acute childhood myositis (BACM).

It is interesting to note that two of our patients had unusually cold feet—a sign which, to the best of our knowledge, has not been associated with BACM previously. This may suggest the pathogenesis involves a vascular component although in both these children the capillary return was normal.

The majority of reports have associated the illness with influenza viruses, particularly influenza B.2,4 7–9 11 Although only one of our children had evidence of recent influenza B infection there was evidence of significant influenza B activity in the community at the time our cases presented.12

Our viral studies however were limited to serology and no specimens were sent for direct detection or culture.

It has been suggested that susceptible children only become ill on the primary exposure to a particular strain of influenza, which may account for the lack of cases seen in adults. It has been shown from serological testing that children with myositis in association with influenza were naïve to the infecting strain.3 7 Two children have been recorded as developing a second episode of myositis in response to a different strain of influenza, but when re-exposed to a previously encountered strain did not develop myositis.13

Some authors have not derived any association with influenza either in terms of virus isolation or serology.7 It is interesting to note that two of our patients had send a serum sample to virology. Even though the virus associated with influenza, the results were negative. It is possible that the virus identified was not influenza but another virus such as a respiratory syncytial virus (RSV).

In our series, all patients were tested for influenza A, B, RSV, adenovirus, HSV, and EBV. It is possible that other viruses may also be involved in the pathogenesis of myositis. It is important to rule out rhabdomyolysis by performing urinalysis for evidence of myoglobin. It is prudent if drawing blood to perform a full blood count, basic biochemistry, and send a serum sample to virology.

Features (see table 2) not associated with BACM should prompt further investigation and referral.

SUGGESTIONS FOR MANAGEMENT BY EMERGENCY PHYSICIANS

A large number of conditions can cause muscle pain in children and it is important to distinguish BACM from other more severe illnesses.

Firstly the history is generally consistent: a viral prodrome followed by acute onset of severe muscle pain, usually confined to the legs. The resolution of symptoms is swift—many patients are markedly improved within 24 hours.

A full and thorough examination should be performed to include a detailed neurological examination. Neurological signs other than weakness as a result of muscle pain may herald more severe pathology.

The most consistent laboratory result is a raised CPK but it is important to rule out rhabdomyolysis by performing urinalysis for evidence of myoglobin. It is prudent if drawing blood to perform a full blood count, basic biochemistry, and send a serum sample to virology.

Features (see table 2) not associated with BACM should prompt further investigation and referral.

<table>
<thead>
<tr>
<th>Feature</th>
<th>Disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td>Erythematous plaques on the face and eczema-like patches on flexor surfaces</td>
<td>Juvenile dermatomyositis</td>
</tr>
<tr>
<td>Severe systemic upset with myoglobinuria</td>
<td>Rhabdomyolysis</td>
</tr>
<tr>
<td>Recent travel to or residence in tropical area with fever, headache, eyes/pain/muscle pain, and a rash on extremities</td>
<td>Dengue fever</td>
</tr>
<tr>
<td>Insidious onset of painless weakness</td>
<td>Muscular dystrophy</td>
</tr>
<tr>
<td>Ascending symmetrical weakness often associated with pain and diminished reflexes</td>
<td>Guillain-Barre syndrome</td>
</tr>
</tbody>
</table>

Table 1 Blood results

<table>
<thead>
<tr>
<th>WCC (x10^3/l)</th>
<th>Platelets</th>
<th>CPK</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reference range: 5–17</td>
<td>Reference range: 150–400</td>
<td>Reference range: 0–170</td>
</tr>
<tr>
<td>Case 1</td>
<td>3.6</td>
<td>250</td>
</tr>
<tr>
<td>Case 2</td>
<td>3.2</td>
<td>150</td>
</tr>
<tr>
<td>Case 3</td>
<td>3.5</td>
<td>147</td>
</tr>
<tr>
<td>Case 4</td>
<td>2.2</td>
<td>123</td>
</tr>
</tbody>
</table>

CPK, creatine phosphokinase; RSV, respiratory syncitial virus; WCC, white cell count.

Table 2 Features not associated with BACM

<table>
<thead>
<tr>
<th>Feature</th>
<th>Disorder</th>
</tr>
</thead>
<tbody>
<tr>
<td>Erythematous plaques on the face and eczema-like patches on flexor surfaces</td>
<td>Juvenile dermatomyositis</td>
</tr>
<tr>
<td>Severe systemic upset with myoglobinuria</td>
<td>Rhabdomyolysis</td>
</tr>
<tr>
<td>Recent travel to or residence in tropical area with fever, headache, eyes/pain/muscle pain, and a rash on extremities</td>
<td>Dengue fever</td>
</tr>
<tr>
<td>Ascending symmetrical weakness often associated with pain and diminished reflexes</td>
<td>Muscular dystrophy</td>
</tr>
</tbody>
</table>

www.emjonline.com
If the presentation is of classical BACM then the A&E clinician should feel comfortable discharging the patient. However we would recommend arranging to review the patient after a couple of weeks to ensure complete resolution of symptoms. At this stage it would be useful from an epidemiological point of view to draw blood for convalescent viral titres.

CONCLUSION
In publishing this case series we hope to raise awareness of an infrequently encountered condition among A&E clinicians. It is important to recognise BACM as a self-limiting condition which could however be mistaken for a more sinister disease and unnecessary tests instigated.

As clusters of cases occur, children are likely to self present to A&E because the symptoms are alarming and parents may feel inclined to bypass their GP. However the syndrome can be managed safely in an A&E setting without admission or referral, if there is capacity for review in an A&E clinic.

In highlighting this condition it may be that more thorough viral studies are performed in future cases, bringing us closer to understanding the infective aetiology of this syndrome.

REFERENCES