Child Health Update

Viral myositis in children

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Abstract

**Question**
I recently evaluated a child in my clinic after an emergency department visit where she presented having woken up that morning refusing to walk and was crawling around the house. The parents reported she was getting over a cold, and I recall similar cases of myositis during the H1N1 influenza epidemic a few years ago. What are the key features of myositis that I should recognize? Which investigations are needed to confirm the diagnosis and how should affected patients be managed?

**Answer**
Benign acute childhood myositis is a mild and self-limited sudden onset of lower extremity pain during or following recovery from a viral illness. Presentation can include tiptoe gait or refusal to walk, secondary to symmetric bilateral lower extremity pain that resolves quickly, usually within 3 days. In general, no investigation is needed except in severe cases for which screening bloodwork and a urine myoglobin test can confirm the diagnosis and rule out complications. Myoglobinuria and highly elevated creatine phosphokinase levels are rare but should be a consideration for admission to hospital. Prognosis is excellent and management might include rest and analgesia.

A child complaining of leg pain or refusing to walk is of concern to both parents and health care providers. Benign acute childhood myositis (BACM) is a common, self-limited condition and a frequently missed cause of lower leg pain in children. This muscle-related condition is also known as influenza-associated myositis, viral myositis, and acute myositis.

It was first documented in 1905 by Leichtenstern as tender thigh and calf pain, suggested as a complication of influenza. In 1957, Lunderberg formally recognized the condition among school-aged children who developed a high-grade fever and catarrhal prodrome that subsided after 4 days, only for severe pain to develop in the calf muscles that caused difficulty walking. He named the syndrome *myalgia cruris epidemica.*

The exact incidence and prevalence remains undetermined, and no clear management guidelines exist. The characteristic clinical and laboratory features of BACM allow for rapid diagnosis, without further investigation or admission to hospital for most children.

Distinguishing BACM from serious disease processes

The primary complaint of pain in the calves in a child presenting with a history of recent viral illness should bring BACM to the forefront of clinicians’ minds. The reliability of BACM’s presentation enables physicians to make a clinical diagnosis while excluding other, more sinister, conditions (Box 1). Clinicians need to differentiate reluctance to walk secondary to pain in BACM from muscle weakness associated with other conditions.

Benign acute childhood myositis predominantly occurs in school-aged children, at a median age of 8.3 years (range 7.3 to 10.3 years) with a male-to-female ratio of 2:1. A history of contact with other sick individuals is frequently not evident in BACM, nor is a previous history or family history of a similar presentation. Providers should inquire about any family history of neuromuscular disease, recent vigorous exercise or trauma, or ovarian and testicular torsion.

**Box 1. Differential diagnoses to consider in BACM**

Consider the following:
- Trauma or nonaccidental injury
- Guillain-Barré syndrome
- Rhabdomyolysis
- Osteomyelitis
- Deep vein thrombosis
- Juvenile rheumatoid arthritis
- Malignancy
- Dermatomyositis
- Polymyositis
- Muscular dystrophy
- Intracranial pathology

BACM—benign acute childhood myositis.
Data from Tippett and Clark, Jain and Kolber, and Mackay et al.

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A thorough physical examination will further support the clinical diagnosis of BACM. Patients might be afebrile or have a mildly elevated temperature but otherwise should have normal vital signs. The distinguishing feature with BACM is the findings of bilaterally symmetric lower extremity examination. On inspection, the child’s gait might appear wide based (tiptoe walking or stiff legged); no common external signs are seen other than occasional mild swelling, and on palpation there can be exquisite tenderness, which usually resolves after 2 to 4 days. The child will have normal range of motion of the knee and hip joints but might hold the feet in plantar flexion, often refusing active and even passive dorsiflexion. There should be no sensory or motor deficits, altered deep tendon reflexes, or change in plantar reflexes.

Investigations
Laboratory testing should be limited to children who will not walk at all. An elevated creatine phosphokinase (CPK) level is one of the most common laboratory findings in BACM. In one study, 95% of patients demonstrated elevated CPK levels at a median of 4100 U/L, and CPK levels rose as high as 20 times the upper limit of normal in another study. High CPK values with a protracted elevation are associated with muscular dystrophy, while in BACM elevated CPK levels will peak after 2 weeks. Patients can have normal or decreased white blood cell and platelet counts and elevated aminotransferase levels. Viral studies and muscle biopsies or myelography should not be done routinely.

Imaging studies should only be used to exclude alternative diagnoses. Indications to perform imaging include concern about trauma, osteomyelitis, malignancy, or deep vein thrombosis. Magnetic resonance imaging might serve as a noninvasive confirmatory tool but is not currently recommended.

There are no clear guidelines for investigation in children with suspected BACM. Based on 5 cases in a retrospective series and review of the literature, Agyeman et al suggested confirming BACM using CPK level and
viral studies. In children with a rapidly worsening condition or no symptomatic resolution after a few days, the investigators suggested urine and renal function studies in order to rule out rhabdomyolysis and renal failure. Tippett and Clark recommended that workup include routine complete blood count, C-reactive protein levels, creatine kinase levels, liver function tests, and urine myoglobin measurement—all in an effort to rule out other, more ominous, disease processes.

**Management**

Clinical recovery is expected at a median of 3 days and few patients, if any, require admission to hospital. In a large Canadian prospective study, over 2 influenza seasons, 5 of 26 children between the ages of 5 and 15 were admitted. Only 1 patient had a prolonged illness with intermittent leg pain and gait abnormalities for 7 weeks, while all others fully recovered after 2 to 3 weeks. Moon et al reported complete recovery of 100% of patients, as did Agyeman and colleagues in their retrospective analysis of 5 patients. In a case series of 4 children with H1N1-associated BACM none of them was admitted to hospital, with full recovery after 4 days on average, despite influenza A syndrome notoriously being more severe than other viruses.

Rhabdomyolysis is an infrequently reported complication of BACM. In one review, 10 of 316 patients developed rhabdomyolysis, 8 of whom had renal failure. All but 1 patient, who had predisposing familial carnitine palmityl transferase deficiency, recovered. Rhabdomyolysis was 4 times more likely to occur in girls than boys, and 86% of all cases were associated with influenza A. Compartment syndrome was reported in 2 of the 311 patients reviewed, and in 1 patient a permanent disability was documented. For children with rhabdomyolysis, admission to hospital for monitoring of renal function is warranted in order to ensure timely diagnosis of acute renal failure, electrolyte abnormalities, or compartment syndrome.

Once an ominous diagnosis has been excluded, children with BACM can be managed as outpatients with analgesia and appropriate clinical and laboratory follow-up in 2 to 3 weeks. Recurrence is uncommon but was identified in 10 of 311 cases in one report. Antivirals are unlikely to be beneficial. A relationship between influenza vaccinations and BACM has yet to be established.

**Conclusion**

Benign acute childhood myositis is a mild and self-limited process that can be diagnosed clinically. If there is any muscle weakness or abnormal neurologic findings, any signs of inflammation, a lack of improvement after 3 days, or asymmetrical lower extremity pain, BACM is unlikely and an alternative diagnosis should be sought. Patients should have screening bloodwork urine myoglobin measured to confirm the diagnosis and rule out more harmful alternatives and complications. Myoglobinuria is rare and when it occasionally occurs these patients should be admitted to hospital for monitoring. Parents and providers should be reassured that prognosis for BACM is excellent and patients can be effectively managed with simple analgesia at home. Clinical follow-up can be arranged with full clinical and laboratory recovery expected at 2 weeks.

**Competing interests**

None declared

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**References**