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CLINICAL CASE

Benign acute childhood myositis: Clinical series and literature review

Miositis aguda benigna de la infancia. Serie clínica y revisión de la literatura

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Abstract

Benign acute childhood myositis (BACM) is a rare clinical condition that mainly affects pre-school and school age-children. It is usually preceded by a viral illness, particularly influenza virus infection. **Objective:** To describe a cluster of BACM cases that were seen in a paediatric unit. **Patients and Methods:** A retrospective serie of cases that presented with a clinical picture suggestive of BACM between August and November 2012 in the paediatric emergency department of a private clinic. **Results:** Nine children, between 4 and 12 years, presented with a history of a recent febrile upper viral respiratory infection, followed by intense calf pain and claudication. They all recovered without complications. Laboratory results showed a marked increase in CK, with a mean of 4,066 IU/l. Three of the cases had influenza *B* infection and one *Mycoplasma pneumonia* infection. They were managed conservatively with hydration and non-steroidal anti-inflammatory drugs. **Conclusions:** BACM is a benign entity with a characteristic clinical presentation that can be managed most of the time in the ambulatory setting, avoiding invasive studies and unnecessary hospital admission.

Keywords:
Viral myositis;
Influenza;
Limp;
Children

Introduction

Benign acute childhood myositis (BACM) is a transient and rare inflammatory condition. It occurs mainly in school and pre-school aged children, predominantly affects males, and case outbreaks are observed in periods of respiratory virus epidemics[1–3]. It is characterized by sudden and intense pain with marked increase of sensitivity of the calf muscles, to the point of hindering or impeding walking, or bearing weight. Laboratory tests include elevated creatine kinase (CK) muscle enzyme.

BACM is of self-limiting evolution, with an excellent prognosis and no functional sequelae[2-7]. It was initially described in Sweden in the late 1950s by Lundberg as *epidemic cruris myalgia*[8]. Since then, numerous sporadic cases and some outbreaks have been described around the world, but their actual prevalence is still unknown. The review by Buss et al.[9] shows an incidence of 2.6 cases per 100,000 children under 18

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years old in epidemic times and 0.23 cases in non-epidemic times. Only sporadic cases have been reported in adults.

The etiology of this condition strongly indicates a viral origin: influenza virus (A and B) is the most frequently reported[2,4,5]. The etiologic mechanism of myositis is still controversial. Current theories support damage by direct invasion of the virus into muscle tissue, with viral particles being isolated in biopsies of gastrocnemius of children with BACM[4,7,10]. The initial infection causes necrosis of the muscle fiber, which results in elevated CK.. Muscular study has been performed infrequently, in view of the short duration of symptoms and the well-known good prognosis of this condition. The few reported muscle biopsies present normal myocytes, myositis[4], segmental rhabdomyolysis[11] or moderate muscle necrosis with interstitial inflammation[12]. When electromyographies have been performed, they have been normal or have shown patchy myopathic changes[3].

The objective of this article is to describe the clinical experience in an outbreak of patients that came to our care center, in a short period of time, with clinical features suggestive of BACM. Along with analyzing the particularities of this series, we review the topic and the elements of clinical judgment for its appropriate diagnostic study and therapeutic management. According to our review of the literature, there is no other series of BACM cases published in Latin American countries.

Patients and Method

Retrospective clinical series of patients who consulted for a clinical picture compatible with BACM in the period between August 1 and November 30, 2012, in the pediatric emergency department of a private clinic in the Metropolitan Region, Santiago, Chile. All patients who had a history of known neuromuscular and immunological disease, active bacterial infections or who were taking drugs with possible muscle toxicity (eg statins) were excluded.

Clinical records of 9 patients were reviewed, emphasizing clinical presentation characteristics, diagnostic elements, therapeutic management and initial evolution..

This study has been approved by the Institutional Ethics Committee.

Results

In a 4-month period, 9 patients with BACM-compatible clinical presentation were presented. All of them with a recent history of severe acute pain in both calves and claudication. The distribution by gender was practically equivalent (5/9 males), with a mean age of 7.3 years (4-12 years). As a clinical antecedent, all patients presented a fever prodrome associated with upper respiratory symptoms. The average onset of muscle symptoms was 4.4 days (3-5 days) after the onset of respiratory symptoms (table 1).

The physical examination on admission showed all of them in good general condition, although a third remained still feverish. They all had severe localized pain in both calves, especially triggered by foot dorsiflexion or vigorous compression of the gastrocnemius muscles, with a greater or lesser degree of claudication: 2 did not walk, 2 had equine gait and one Frankenstein gait (table 1). Muscle strength, tone, and osteotendinous reflexes of the lower extremities were uniformly conserved. The remainder of the physical examination was normal, except for the presence of upper respiratory symptoms in all of them. Although all of our patients were evaluated by a pediatric neurologist, none merited extra studies to rule out any other differential diagnosis.

Of the 9 patients, 8 were hospitalized for monito-

Table 1. Patients demographic characteristics, prodromal symptoms and presentation latency

Patient	Age (years)	Sex	Latency (days)	Prodromal symptoms
1	12	F	5	Cough, nasal congestion, headache, fever
2	8	F	5	Coryza, cough, fever
3	5	F	5	Cough, coryza, nasal congestion, fever
4	4	M	5	Cough, coryza, diarrea, fever
5	7	F	4	Cough, vomit, headache, fever
6	10	M	5	Nasal congestion, coryza, sore throat, fever
7	9	M	4	Nasal congestion, fever
8	4	M	3	Coryza, dysphonia, cough, fever
9	7	M	4	Cough, fever

Table 2. Physical Examination on admission

Patient	Fever	Physical examination	Recovery time (days)
1	+	Calf pain, tip toe walking	1
2	-	Calf pain, not bearing weigth, dolor a la dorsiflexion pie	2
3	+	Calf pain, not bearing weigth	NR
4	+	Calf pain, claudication	2
5	-	Calf pain, not bearing weigth, tip toe walking	2
6	+	Calf pain, claudication, dolor a la dorsiflexion pie	NR
7	-	Calf pain, claudication	2
8	-	Calf pain, Frankestein gait	NR
9	-	Calf pain, claudication	NR

NR: not registered

ring and study for a period of no more than 3 days. The clinical management consisted exclusively of hydration and nonsteroidal anti-inflammatory drugs, resulting in a favorable evolution in all of them

In our series, all patients evaluated with blood counts, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and CK levels. In the exams, leukopenia with neutropenia was noted in 6 of 9 patients. Four patients had mild thrombocytopenia and all had an increase in CK, with an average value of 4,066 IU/L (table 2). Both ESR and CRP were in the normal range in all of them. Transaminases were monitored in 8 patients; of these, 7 had a slight increase in their values, mainly glutamic oxalacetic transaminase (GOT), and 2 of them had a slight increase in lactate dehydrogenase (LDH).

Eight of 9 patients were monitored for renal function with serum creatinine, all in the normal range. Myoglobinemia was requested in 5 patients: in 4 of them it was elevated (table 2), although all the urinalysis were normal and without myoglobinuria. The etiological study showed associated agents in only 4 of our patients: 3 patients with influenza B and one with *Mycoplasma pneumoniae*. In the rest, the search for etiological agent was negative (tabla 2).

Our patients evolved satisfactorily during hospitalization, with progressive drop in CK levels (figure 1), without complications and with a rapid regression of

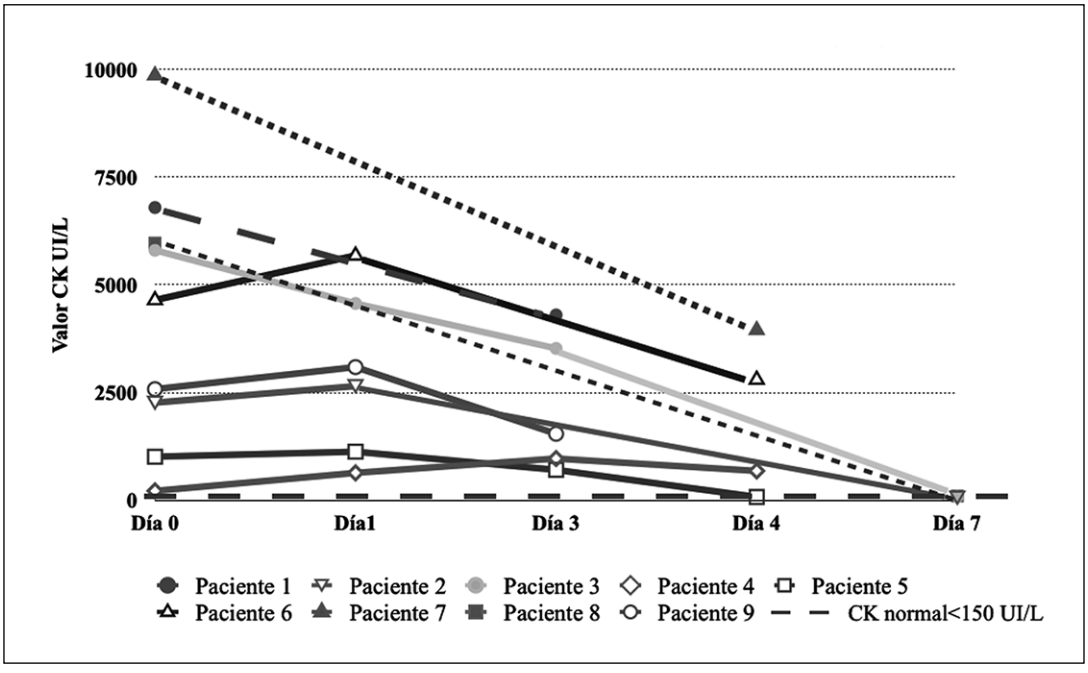

Figure 1. Evolution of creatinkinase (CK) levels in blood.

Table 3. Laboratory investigations

Patient	WBC/Neutrophils	Platelets	LDH	GOT/GPT/GGT	ESR	CRP
1	2700/915	146000	694	203/76/NR	7	< 0.1
2	4500/2597	170000	620	105/29/14	NR	0.32
3	2400/725	151000	772	157/38/10	7	0.1
4	5700 /1630	138000	590	37/15/11	7	0.6
5	3400/1054	205000	420	57/18/3	1	0.1
6	4200/1482	165000	NR	NR	2	0.1
7	3000/1146	114000	923	323/113/15	NR	0.1
8	4700/1706	199000	596	90 /32/11	6	0.1
9	3700/1428	128000	550	157/42/13	5	0.33

WBC: White blood cells, LDH: Lactic Deshidrogenase, GOT: glutamic oxaloacetic transaminase, GPT: glutamic pyruvic transaminase, GGT: Gamma Glutamyltransferase, ESR: Erythrocyte Sedimentation Rate, CRP: C Reactive Protein, NR: not registered

Table 4. Creatinekinase values

Patient	CK admission/day 0	CK day 1	CK day 3	CK day 4	CK day 7
1	6793	NR	4300	NR	NR
2	2264	2648	2000	1000	60
3	5800	4567	3523	NR	88
4	216	634	967	679	NR
5	1009	1126	704	76	NR
6	4646	5680	NR	2800	NR
7	9860	NR	NR	3950	NR
8	5967	4000	2400	NR	98
9	2578	3089	1542	NR	NR

NR: not registered.

theirs symptomatology. In the 5 patients who had ambulatory follow up, the total resolution time of symptoms from discharge was not greater than 48 h (table 1).

It is interesting to mention that one of our male patients presented a new episode of BACM one year later, also associated with previous respiratory infection without etiologic study, which was managed on an outpatient basis.

Discussion

This report describes an outbreak of BACM that occurred in our care center, coinciding with a period of higher prevalence of respiratory infections, which has been related to an increase in the incidence of this condition[8,13]. The influenza virus (with predominance of influenza B) has been the most frequently involved[1,2,4,5,14,15].However, association with other viruses such as coxsackie, adenovirus, parainfluenza, respiratory syncytial virus, among others, and bacteria such as *Mycoplasma pneumoniae* has been described[3–5]. In our series, we isolated the responsible agent in 4 patients (3 with influenza B and 1 with *Mycoplasma pneumoniae*). However, in most of the cases only direct immunofluorescence (DIF) and serological studies for *Mycoplasma pneumoniae* were available, which decreases the detection sensitivity. In our laboratory the sensitivity of the DIF for the different respiratory viruses is less than 50%, except for RSV, for which it reaches 63%.

All of our patients presented the classic clinical picture, with predominance of bilateral calf pain and claudication and with a febrile respiratory prodrome.. The age of presentation also coincided with that described in the literature[2,4,6,8], although we did not find a clear male predilection, which could be explained by the small number of patients. Of the 2 types

of gait described in BACM: a wide based, stiff legged gait (Frankenstein gait) and toe walking (gait in equine)[4], both were observed in our series. The calf muscles invariably show excruciating pain on palpation and dorsiflexion of the foot, although, exceptionally, there is compromise of other muscle groups such as thighs, arms, back and neck[7]. At the time of diagnosis, and in agreement with our series, patients are usually afebrile, in good general conditions and with a normal neurological examination. If discrete muscle weakness is perceived it is due to pain and not to neurological deficit.

The most striking laboratory result is the marked elevation of the CK muscle enzyme (20-30 times higher than normal values) which typically normalizes in a couple of weeks[4,5]. Even in cases where CK has been massively elevated it is seldom associated with myoglobinuria and significant rhabdomyolysis[16]. Other findings are leucocytopenia with moderate neutropenia, thrombocytopenia and a mild and transient elevation of transaminases and LDH[6,16,17]. Inflammatory markers are generally normal, although ESR may be slightly increased[2,4]. This coincides with the laboratory parameters observed in our patient group.

The clinical evolution described in the literature is similar to that recorded by us, with a marked improvement at 24 h of evolution[18]. The process is self-limited with complete recovery between the third and tenth days, complications are infrequent and do not leave functional sequelae[4,16]. Acute renal failure secondary to myoglobinuria due to massive rhabdomyolysis is anecdotal in patients with BACM and has not been observed in our patients[16,19]. Recurrences are rare - just one case detected in our group - and caused by different viruses at each opportunity[5,17]. It has been shown serologically that children with myositis associated with influenza virus are susceptible to the strain involved, and those who presented a second episode did so with a strain different from that of the initial infection. This disorder appears to be present only at the first exposure to a particular virus, which may explain the few cases reported in adulthood[7,17].

The symptoms of BCAM are alarming and can cause concern and confusion both in parents and health professionals. On the other hand, there is a wide differential diagnosis made up of a spectrum of diseases that present with claudication and/or muscle pain. These include infectious, muscular and neurological diseases such as acute myositis associated with other infectious diseases (eg dengue), toxic myoglobinuria, rhabdomyolysis, Guillain-Barré syndrome, ataxia, transverse myelitis, muscular dystrophies, polymyositis, juvenile dermatomyositis, trichinosis , osteomyelitis, arthritis and deep vein thrombosis, among others[16,20,21].

However, the clinical presentation of BCAM is characteristic and musculoskeletal and neurological examination in these patients is normal, except for pain on palpation of the affected muscle group and discrete secondary weakness[16].

Therefore, on suspicion of this entity, it is very important to obtain an exhaustive medical history and a detailed physical examination, where the musculoskeletal or neurological involvement suggestive of more ominous differential diagnoses is excluded. Findings that guide other diagnoses include: family history of neuromuscular disease, history of recent trauma, persistent high fever, dark urine, subacute or chronic progression, rash, frank muscle weakness or other neurological examination abnormalities[16].

Laboratory tests should be dimensioned and oriented to establish the diagnosis (elevation of CK). Exceptionally, and on suspicion of rhabdomyolysis due to excessive CK increase, renal function could be monitored. However, it is important to be aware of this rare but severe complication, suggested by dark urine and positive blood on urine dipstick in the absence of red blood cells on microscopic examination of urine.. An accurate diagnosis prevents invasive studies and unnecessary hospitalizations, while avoiding greater anxiety in the patient and his family.

Management of BCAM is symptomatic[18,22]. The use of antivirals in the case of influenza infection is of little benefit, since in most cases the acute respiratory infection is already in resolution[1,20,21]. Influenza vaccine has proven to reduce influenza complications, which could theoretically reduce the incidence of BCAM, which is not proven[4,21].

This study has some limitations, such as the small number of patients included, which does not allow generalization of the conclusions. Likewise, as a retrospective study, data recording in the clinical file by the treating physicians was not done in a protocolized way, so it was not always complete. For this same reason, the clinical management of these patients was not necessarily uniform. Since the etiological study was performed in most cases with less sensitive tests than is currently available, there may have been an etiological underdiagnosis of our patients. Finally, we only have the follow-up data of those patients who were controlled in our outpatient clinic, which does not allow us to know with greater certainty eventual recurrences or long term complications.

Conclusion

Our work, besides presenting the clinical series, fulfills the objective of emphasizing that BCAM is a benign, self-limited entity with an excellent prognosis, with a characteristic clinical presentation and that in

most cases it can be managed on an outpatient basis. For this reason, the study should be limited and oriented towards diagnostic confirmation, and avoid invasive studies and unnecessary hospitalizations.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Financial Disclosure

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Conflicts of Interest

Authors state that any conflict of interest exists regards the present study.

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