

## ORIGINAL ARTICLE: CASE REPORT

### Focal myositis in the lower leg of an 11-year old girl

Marrije van Ijperen<sup>1</sup>, Paul H.G. Hogeman<sup>1</sup>, Ben G.F. Heggelman<sup>1</sup>,  
Sjoerd G. van Duinen<sup>2</sup>, and Nico M. Wulffraat<sup>3</sup>.

<sup>1</sup>Department of Pediatrics, Meander Medisch Centrum Amersfoort, the Netherlands.

<sup>2</sup>Department of Pathology, Leiden University Medical Center, the Netherlands.

<sup>3</sup>Department of Pediatric Immunology, Wilhelmina Kinderziekenhuis, University Medical Center Utrecht, the Netherlands

#### Contact:

Nico Wulffraat

Department of Pediatric Immunology

Wilhelmina Kinderziekenhuis University Medical Center

P.O.Box 85090, 3501 AB

Utrecht, the Netherlands

phone 0031-30-2504000

e-mail n.wulffraat@wkz.azu.nl

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#### ABSTRACT

An 11 ½ year old girl presented with a painful left lower leg and increased swelling of her calf. On examination there was a palpable tumor in her gastrocnemius muscle. Laboratory findings were normal. An MRI showed two similar lesions: a large one in her left musculus gastrocnemius and a smaller one in her left musculus flexor digitorum longus. The biopsy was compatible with focal myositis. This case report underlines the importance of the differential diagnosis for this unusual condition.

#### Case Report

An 11 ½ -year old girl presented with a painful left lower leg for 3 months. At the same time, she discovered a swelling of increasing size on the lateral side of her left calf. Gradually she developed a contracture of her left gastrocnemius muscle resulting in pes equinus. She was in good health, but had limited function of her left leg. She occasionally used acetaminophen or naproxen for the pain. There were no neuromuscular or autoimmune disorders in her family.

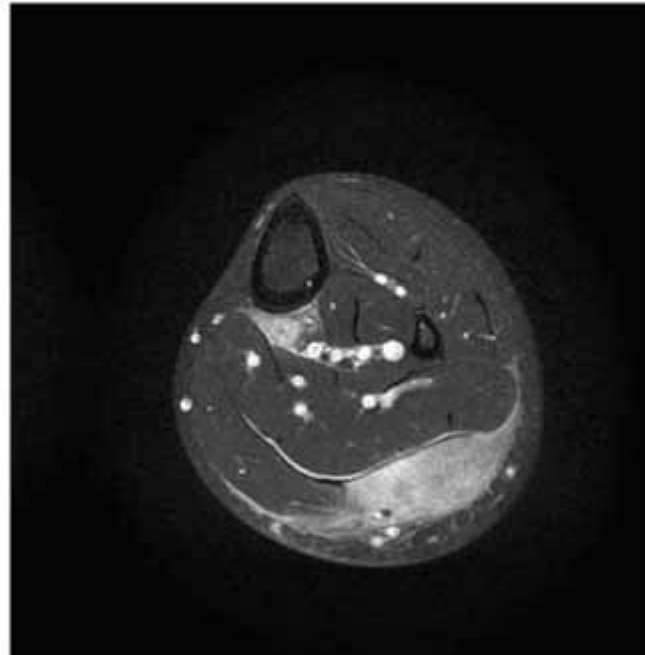
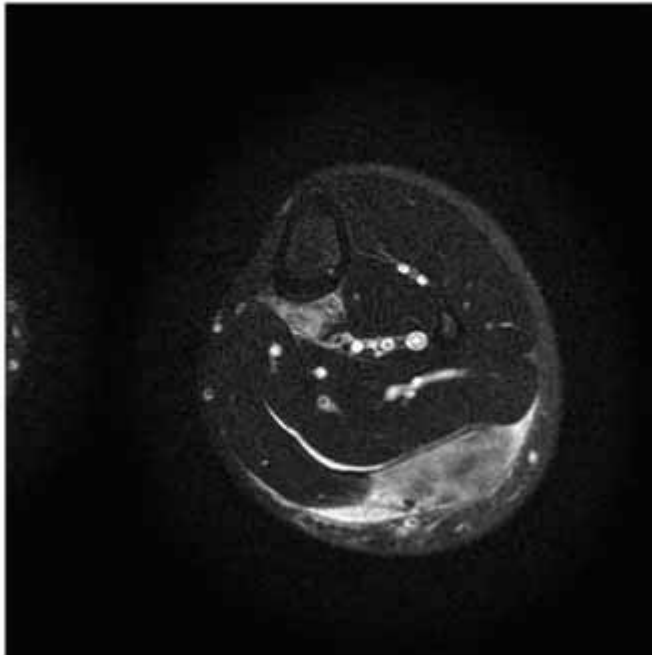
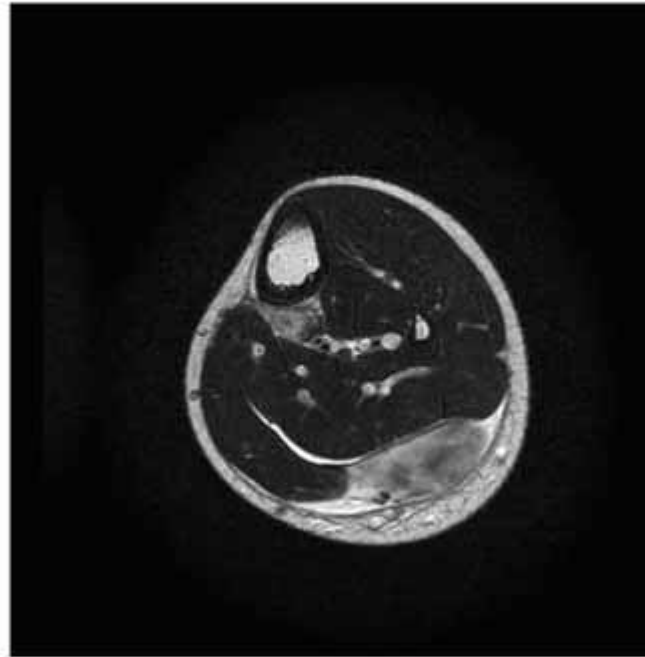
Further medical history did not reveal a recent viral infection, diarrhea, tick bite or trauma. The girl has not been vaccinated with Bacille Calmette Guerin (BCG).

On examination the child appeared healthy. Her left knee had a flexion contracture. Her left calf circumference was 0.5 cm more than the right calf. At 5 and 10 cm under the tibial tuberosity, the circumferences were 29.5 and 26.5 cm on the right leg and 30 and 27 cm on the left leg. An elastic firm nodule of 3 x 7 cm was palpable at the lateral head of her gastrocnemius. The tumor was extremely painful on palpation. The overlying skin and the underlying tissue were mobile. There were no skin abnormalities. The muscle strength was normal. Other muscles and joints were not affected, and there were no other palpable tumors. The left foot was colder than the right, but peripheral pulsations were normal. There were no other abnormalities on examination.

Laboratory findings included a normal serum C-reactive protein (CRP) of 6mg/L, erythrocyte sedimentation rate (ESR) of 19 mm/hour, creatinine kinase (CK) of 101U/L and negative antinuclear antibodies (ANA). Myositis-specific autoantibodies (MSA) (Jo-1, Scl-70, RNP, SSA) were negative. Serum *Borrelia burgdorferi* IgM and IgG tests were negative. No *Campylobacter* or other pathogenic bacteria could be cultured in the stools. A plain radiograph of the left lower leg was normal. Ultrasound examination showed a lesion in the soft tissues with mixed increased and decreased signal intensity, and with a good vascularization. The MRI (Figure 1) demonstrated a diffuse abnormality in the lateral head of the gastrocnemius and also in the flexor digitorum longus.

Figure 1.

MRI of left lower leg. Upper left (1a): Transversal T1 weighted turbo spin echo (TSE) sequence without fat suppression and without contrast, showing slight swelling of the gastrocnemius muscle. Upper right (1b): Transversal T2 weighted TSE sequence without fat suppression, showing hyperintensity of the gastrocnemius and flexor digitorum muscles. Lower left (1c): Transversal T2 weighted TSE sequence with fat suppression, showing hyperintensity of the gastrocnemius and flexor digitorum muscles. Lower right (1d): Transversal T1 weighted TSE sequence with fat suppression and after Gadolinium, showing slightly increased enhancement of the gastrocnemius and flexor digitorum muscles.



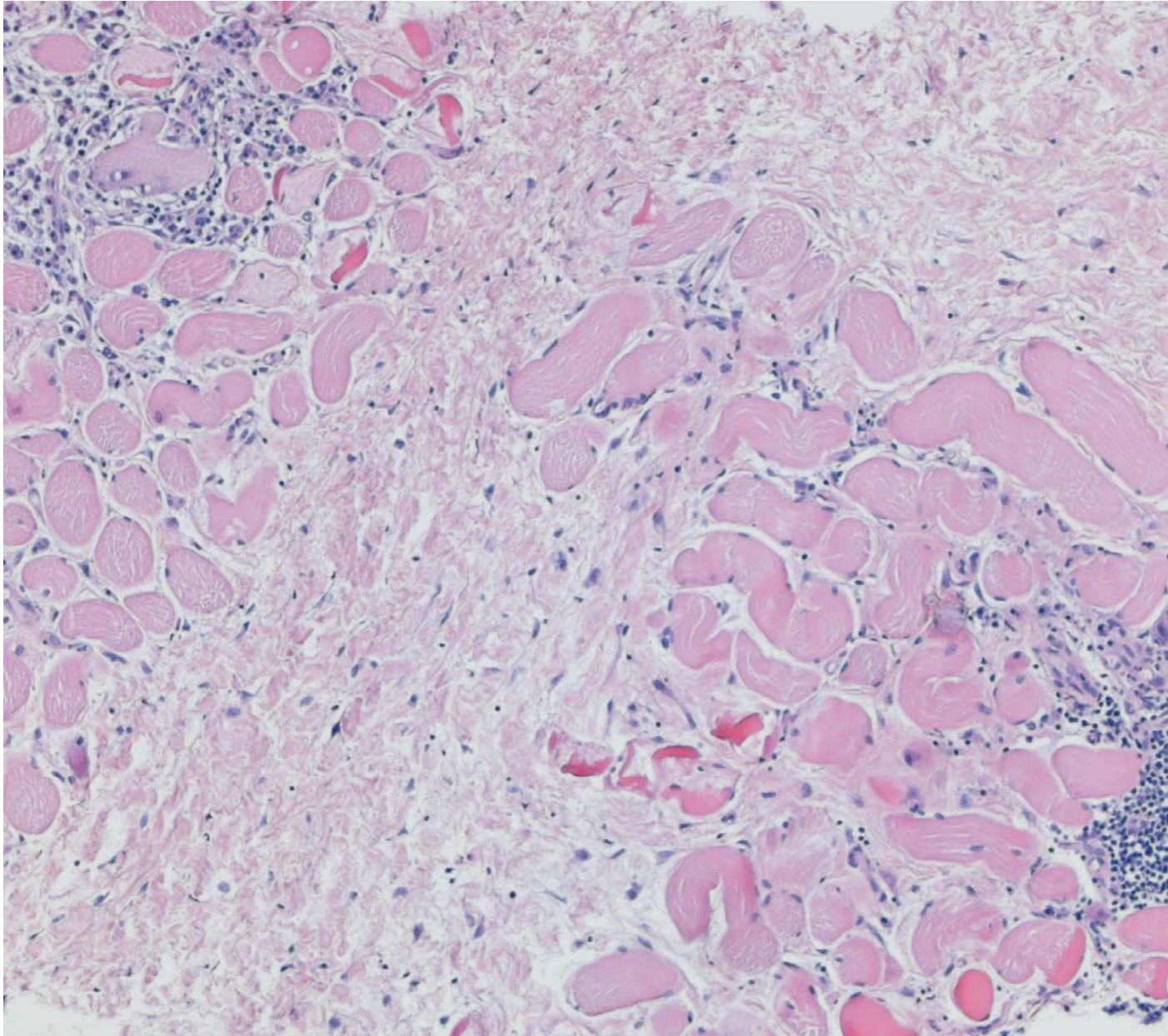
The first lesion, localized laterodorsal in the gastrocnemius, had a size of at least 17x4x5cm (longitudinal by transverse diameters). The second lesion, which was located in the flexor digitorum longus muscle and mediodorsal to the tibia, had a transverse size of 1x1.5 cm. The lesions had a markedly increased signal on T2-weighted images and were iso-intense to muscle tissue on T1-weighted images. They showed strong enhancement after adding Gadolinium. Both lesions had a steep enhancement curve with a plateau phase and no significant wash-out in the first minutes. The 2

separate lesions had exactly similar characteristics on the different sequences. On the basis of these images, a malignant disorder, though unlikely, could not be completely ruled out. Therefore a biopsy was performed.

A biopsy of the largest lesion (Figure 2) showed muscle tissue with loss of the normal fascicular architecture with prominent bands of fibrosis. The muscle fibers varied in size and shape and some were necrotic or regenerating. There was a mostly patchy inflammatory infiltrate of B-lymphocytes and mainly CD4-positive T-lymphocytes and scattered macrophages, with only few CD8-positive T-lymphocytes. There were no neoplastic cells. Based on these data, we concluded that this child had focal myositis.

Figure 2.

Biopsy of the lesion in the left musculus gastrocnemius. Muscle tissue with loss of the normal fascicular architecture caused by prominent bands of fibrosis. The muscle fibers vary in size and shape and some are necrotic or regenerating.



## Discussion

Focal myositis (FM) is a rare benign inflammatory pseudotumour of a skeletal muscle. This was first described by Heffner et al. [1] It can affect adults as well as children. The typical presentation is a localized painful swelling within the soft tissue of an extremity. Focal myositis often affects the lower limb musculature, but may occur almost anywhere. [2-6] The muscle mass increases in days to months. There is no general muscle weakness and no joint involvement. FM is a self-limiting disease, which resolves spontaneously in a couple of months to 3 years. [7] Recurrence, local and in other muscles, with spontaneous remission has been described. [8]

The etiology of focal myositis is unknown. Possible causes include viral infection, denervation processes, and ischemic necrosis of muscle, but these are not established.[9] Cases have been reported in which focal myositis has been associated with *Campylobacter*

infection, *Borrelia Burgdorferi* and BCG vaccination (10). We found no evidence of these associations in our case.

Most importantly, FM should be differentiated from a malignant soft tissue tumour, such as sarcoma. Other conditions such as localized nodular myositis (LNM) can also initially mimic FM. LNM presents as polymyositis with a focal onset, but progresses in time to involve other muscles and cause muscle weakness. This characteristically does not occur in FM. [8, 10] Characteristics to differentiate between most common inflammatory myopathies are shown in Table 1 and Table 2. [8, 11] This observed clinical presentation also differs from the well known benign childhood myositis, often viral, that usually resolves within weeks. [12]

Table 1.

Characteristics to differentiate between inflammatory myopathies

Characteristics	Polymyositis	Inclusion body myositis	Dermatomyositis adult/juvenile	Focal myositis
Age of onset	>18 yr	>50 yr	all ages	all ages
Muscle weakness	starts proximal symmetric. late distal	starts proximal asymmetric early distal	starts proximal symmetric late distal	focal asymmetric
Progression	weeks-months, 50% full recovery, 5 yr MR 20%	Years, stabilization, remission rare, MR unknown	weeks-months, 20% spontaneous remission in adults, MR 5% in both	self-limiting <4 years rarely progressive or spreading to another area
Characteristics	None specific	knee extensor weakness, wrist/finger flexor weakness, not responsive to treatment, inclusion bodies on	heliotrope rash on eyelids, Gottron papules on knuckles, vasculitis in juvenile cases	focal, no general weakness

		muscle biopsy (EM)		
Associated	other rheumatic diseases and infections	rheumatic diseases and infections	other rheumatic diseases and infections	infections?

Abbreviation: MR: mortality rate

Table 2

Laboratory Manifestations that help distinguish these myopathies

Laboratory Tests	Polymyositis	Inclusion body myositis	Dermatomyositis adult/juvenile	Focal myositis
CK (creatine kinase)	elevated	normal - mildly elevated	elevated	normal
ANA (antinuclear antibodies)	<50% positive	15% positive	60-80% positive	negative
MSA (myositis specific antibodies)	38% positive	rare	41% positive in adults, 10% in children	negative

To diagnose focal myositis, laboratory evaluation is crucial including CT or MRI, electromyography (EMG), and muscle biopsy. The evaluation typically reveals normal or mildly elevated serum CK and other muscle enzymes in FM. Markedly elevated values may indicate progression to a diffuse muscle disease and may indicate recurrence. The ANA and muscle-specific antibodies are negative in FM. The EMG can be performed to distinguish between focal or multifocal involvement. In FM EMG will show isolated irritative myopathy in the affected muscle only. [8] An MRI of the affected muscle may show muscle enlargement, fatty infiltration, increased T2 signal suggesting muscle inflammation and edema, with poorly homogeneous enhancement on T1 images after gadolinium administration. Computed Tomography can also demonstrate enlargement of the muscle and fatty infiltration. [13]

A muscle biopsy should usually be performed to exclude a malignant soft tissue tumor. The typical changes of FM compared to other myopathies are muscle fiber hypertrophy with particularly large size of fiber nests forming tightly packed nodules, enveloped by substantial fibrosis. The inflammatory infiltrate is comprised of mostly T-cells and macrophages. There are no immunohistochemical features to differentiate FM from other inflammatory myopathies. [8] In our case laboratory findings were normal. As the MRI and muscle biopsy were typical for focal myositis, we elected not to perform an EMG.

Treatment is usually not necessary, except for symptomatic treatment with analgesics or nonsteroidal anti-inflammatory drugs (NSAIDs). If FM does not resolve or recurs, in the presence of inflammatory laboratory indices, stronger anti-inflammatory and immunosuppressive therapy such as corticosteroids should be considered. Physiotherapy is often necessary to reduce a contracture.

### **Conclusion**

Focal myositis is unusual in children but must be considered if a child's muscle complaints are limited to one muscle group and are more severe than the typical benign myositis. Diagnosis is made by MRI and muscle biopsy. A muscle sarcoma must be kept in mind. Treatment is symptomatic and the course self-limited.

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