American Journal of Medical Case Reports, 2018, Vol. 6, No. 4, 72-74
Available online at http://pubs.sciepub.com/ajmcr/6/4/5

DOI:10.12691/ajmcr-6-4-5

An Unusual Case of Juvenile Polymyositis Triggered by *Bartonella henselae* Infection

Maria Pereira[1,2,*], Catherine Mary Healy[1,3], Carrie A. Mohila[4], Monica Marcus[1,2]

[1]Department of Pediatrics, Baylor College of Medicine, Texas Children's Hospital, Houston, TX
[2]Division of Allergy, Immunology and Rheumatology, Baylor College of Medicine, Texas Children's Hospital, Houston, TX
[3]Division of Infectious Disease, Baylor College of Medicine, Texas Children's Hospital, Houston, TX
[4]Department of Pathology & Immunology, Baylor College of Medicine, Texas Children's Hospital, Houston, TX
*Corresponding author: maria.pereira2@bcm.edu

Abstract We present the first case of cat-scratch disease described to trigger juvenile polymyositis. A 15-year-old male presented with a prolonged febrile illness. He had been diagnosed with cat-scratch disease 4 weeks earlier based on his exposure to kittens and serologic testing. Treatment with antimicrobials provided partial relief. He continued to have unremitting fever and developed headache, jaw pain, blurry vision, myalgias and impaired ambulation. Imaging studies were most consistent with myositis of the lower extremities and muscles of mastication. A muscle biopsy of anterior tibialis muscle confirmed a diagnosis of polymyositis. Patient had an excellent response upon initiation of immunosuppressive treatment. Juvenile polymyositis is a rare disease and should be part of the differential diagnosis of patients presenting with prolonged fever and severe myalgias with normal muscle enzymes, particularly following a *bartonella henselae* infection.

Keywords: *juvenile polymyositis, idiopathic inflammatory myopathy, masticatory myositis, Bartonella henselae, cat-scratch disease*

Cite This Article: Maria Pereira, Catherine Mary Healy, Carrie A. Mohila, and Monica Marcus, "An Unusual Case of Juvenile Polymyositis Triggered by *Bartonella henselae* Infection." *American Journal of Medical Case Reports*, vol. 6, no. 4 (2018): 72-74. doi: 10.12691/ajmcr-6-4-5.

1. Introduction

Juvenile polymyositis (JPM) is a form of idiopathic inflammatory myopathy. The exact cause of JPM remains unknown. Seasonal clustering in the months of April and May suggests a role of environmental and infectious triggers in the onset of the disease in a genetically predisposed individual [1]. Case reports of polymyositis have been described following human immunodeficiency virus (HIV-1) infection in adults [2,3], and human T-cell lymphotropic virus type 1 (HTLV-1) infection in children [4]. Several mechanisms by which an infection can lead to autoimmunity in idiopathic inflammatory myopathies have been proposed, including molecular mimicry, induction of anti-idiotypic antibodies, and modification of self-antigen through microbial proteins [5,6]. To our knowledge, this is the first case of reported JPM induced by *Bartonella henselae* infection.

2. Case Presentation

A previously healthy 15-year-old Caucasian male presented with fevers for over a month. A diagnosis of Cat-scratch disease (CSD) have been made previously based on the history of playing with kittens and having elevated serum titers of Immunoglobulin G antibody to *B. henselae*. He received treatment with doxycycline and rifampin and was given a short course of corticosteroids for elevated inflammatory markers and leg pain. Despite treatment, he continued to have fevers with worsening muscle pain that progressed to difficulty walking. He also developed headache, jaw pain, and blurry vision. He was brought to the hospital for evaluation.

His physical exam revealed a distressed teenager with blood pressure of 163/91 mmHg, heart rate of 92, temperature of 99.4^{0}F, and respiratory rate of 20. He had exquisite tenderness, increased warmth, and soft tissue swelling of bilateral calves. There was pain with movement of the legs, yet he had normal strength in all extremities. He had an antalgic gait. His cardiac, pulmonary and gastrointestinal examination findings were normal. He had no obvious lymphadenopathy, and skin exam was normal. He was admitted to the hospital for pain control and further investigation.

Laboratory evaluation revealed a CBC with an elevated white count 27x10^3/UL with 78% neutrophil predominance, sedimentation rate 106 mm/h (normal 0-20 mm/h), and C-reactive protein 17.7 mg/dl (normal <1.0mg/dl). He had a normal basic metabolic panel and urinalysis. Muscle enzymes were normal with a creatinine kinase 197 IU/L, lactate dehydrogenase 358 IU/L, aspartate aminotransferase 30 U/L, alanine aminotransferase 28 U/L and aldolase 4.9 U/L. He had a negative myositis-specific autoantibody panel. Magnetic resonance imaging of the bilateral lower extremities revealed increased signal

intensity of nearly all muscles of the legs below the knees that involved deep and superficial fascial planes most consistent with an inflammatory process (Figure 1).

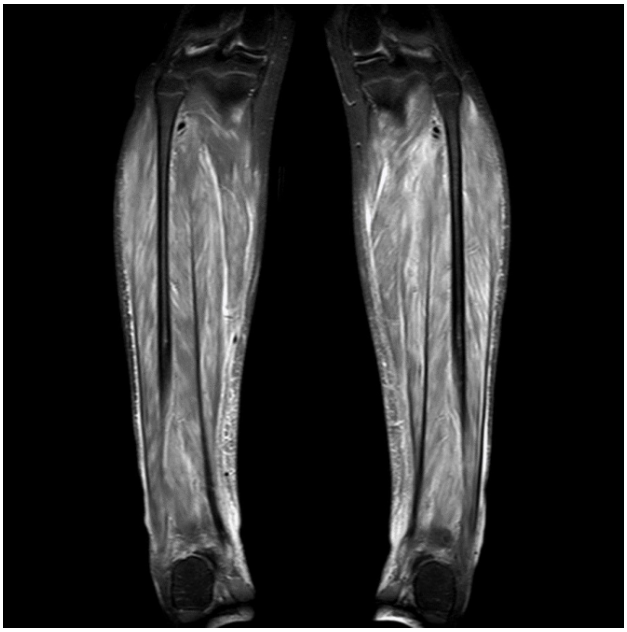

Figure 1. T2-weighted MRI lower extremities coronal view

Imaging of the brain and orbits was performed to evaluate headaches and blurry vision after a neurological and eye exam were normal. Unexpectedly, marked enhancement of all muscles of mastication (temporalis, masseter, and pterygoid muscles) was seen and concerning for an inflammatory process (Figure 2).

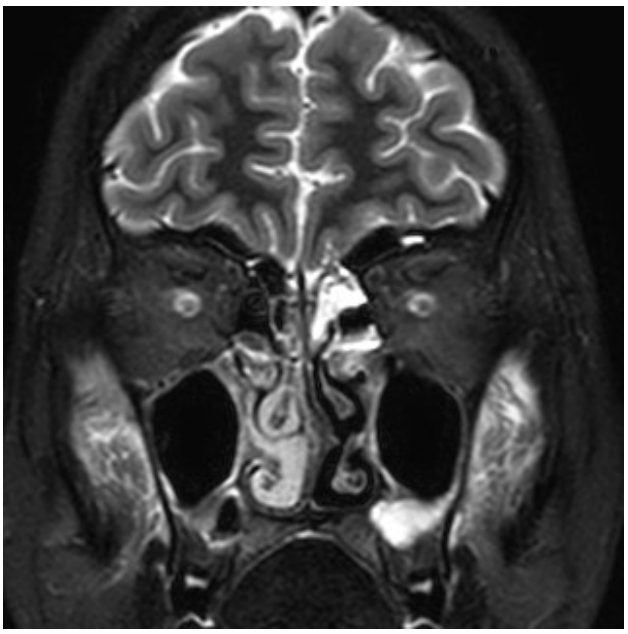

Figure 2. MRI brain and orbits coronal view

A muscle biopsy of the anterior tibialis muscle was then performed and revealed and inflammatory myopathy consistent with polymyositis (Figure 3).

Remarkably, this patient experienced resolution of symptoms after 3 consecutive days of high dose intravenous corticosteroids. He was discharged home on oral prednisone 60 mg daily and methotrexate 25 mg weekly subcutaneous injections as a steroid-sparing agent. A year later, he continues to do very well with no recurrence of his symptoms.

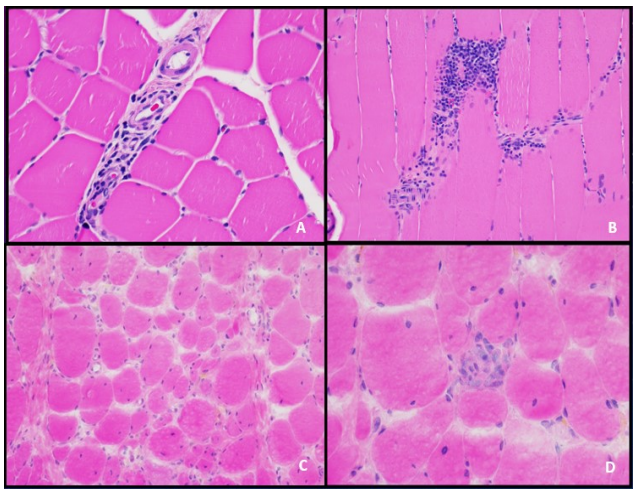

Figure 3. Muscle biopsy of the anterior tibialis muscle shows an inflammatory myopathy with a lymphohistocytic inflammatory infiltrate in endomysium (A) and perimysium (B). There is variation in myofiber size and shape with a mild increase in internalized nuclei (C) with rare fiber necrosis with phagocytosis (D)

3. Discussion

This case is unique in that herein we report the first case of JPM induced by CSD in a pediatric patient. CSD is a common self-limited infection characterized by tender lymphadenopathy at the inoculation site that can last up to several months. Systemic symptoms such as fever, malaise, anorexia and abdominal pain can be present in about half of the cases. Approximately 10% of patients with CSD develop atypical or extra-nodal manifestations [7]. Musculoskeletal manifestations are rare and have not been well defined, with only a few case series of patients with reactive arthritis, myalgias, and osteomyelitis [8,9]. In a large cohort of 913 patients with CSD in Israel, 10.5% had musculoskeletal manifestations, including myalgias (5%), and arthropathies of medium and large joints (5%). Myalgias were often severe with a median duration of 4 weeks, and it was more prominent in patients between ages 20-59 years [9]. None of these patients had a muscle biopsy. Generalized myalgias have been described in patients with systemic CSD, with persistent muscle pain even after fever defervesce [10].

This patient's clinical presentation was different from typical cases of polymyositis that present with muscle weakness in a symmetric and proximal distribution. In contrast, our index patient had distal muscle involvement, which has been described in patients with polymyositis that have a more extensive disease course [11]. Muscle tenderness may be present, but it is not commonly seen on initial presentation.

Involvement of muscles of mastication however, has not been described in JPM. Masticatory muscle myositis is an inflammatory myopathy that presents with jaw pain, swelling, trismus, or atrophy of the muscles of mastication. This condition has not been described in humans, but it is not uncommon in large-breed dogs. In a review of 200

canine cases, the predominant clinical signs were bilateral jaw involvement with ocular symptoms seen in 44% [12], similar to our index case that developed jaw pain and blurry vision. Treatment is based on immunosuppression, which is generally corticosteroids, although azathioprine and cyclosporine can also be considered [13]. In 2007, a review was published describing the similarities and differences between inflammatory myopathy in dogs and humans. The authors discussed the implications of using canine muscle-specific autoantibodies in clinical setting. The antibody against type 2M fiber type found in masticatory muscles may be of relevant use for human patients that present with jaw involvement [14].

The diagnosis of JPM is based on the Bohan and Peter criteria established in 1975. The criteria consist of the presence of symmetric weakness of the proximal muscles, serum elevation of muscle enzymes, electromyography demonstrating denervation and myopathy, and muscle biopsy displaying necrosis and inflammation. A muscle biopsy confirms the diagnosis of JPM when clinical criteria is not met, or diagnosis remains uncertain. A magnetic resonance imaging, specifically T2-weighted and fat-suppressed images, can show muscle inflammation as edema. This imaging modality is increasingly used in the diagnosis of childhood inflammatory myopathy to avoid the morbidity of the muscle biopsy and electromyography.

4. Conclusion

JPM is a rare disease and should be in the differential diagnosis of patients presenting with prolonged fever and pain in distal musculature and muscles of mastication. The findings in the case we describe highlight the musculoskeletal manifestations of CSD and raise the possibility that *Bartonella henselae* may induce JPM, although a causal relationship could not be demonstrated. New muscle-specific autoantibodies may be available in the future and aid in the diagnosis of atypical cases of JPM. An MRI of the lower extremities with T2-weighted and fat suppressed images can help establish a diagnosis in the absence of typical clinical features of JPM. However, a muscle biopsy is critical to confirm the diagnosis in atypical cases.

References

[1] Sarkar K, Weinberg CR, Oddis CV, et al. Seasonal influence on the onset of idiopathic inflammatory myopathies in serologically defined groups. Arthritis Rheum. 2005; 52(8): 2433-8.

[2] Johnson RW, Williams FM, Kazi S, Dimachkie MM, Reveille JD. Human immunodeficiency virus-associated polymyositis: a longitudinal study of outcome. Arthritis Rheum. 2003; 49(2): 172-8.

[3] Roedling S, Pearl D, Manji H, et al. Unusual muscle disease in HIV infected patients. Sex Transm Infect. 2004; 80(4): 315-7.

[4] Ferreira AS, Costa CM, Dantas IK, et al. Polymyositis in childhood as clinical manifestation associated with HTLV-1. Arq Neuropsiquiatr. 2010; 68(6): 962-4.

[5] Zampieri S, Ghirardello A, Iaccarino L, et al. Polymyositis-dermatomyositis and infections. Autoimmunity. 2006; 39(3): 191-6.

[6] Walker EJ, Jeffrey PD. Polymyositis and molecular mimicry, a mechanism of autoimmunity. Lancet. 1986; 2(8507): 605-7.

[7] Florin TA, Zaoutis TE, Zaoutis LB. Beyond cat scratch disease: widening spectrum of Bartonella henselae infection. Pediatrics. 2008; 121(5): e1413-25.

[8] Giladi M, Maman E, Paran D, et al. Cat-scratch disease-associated arthropathy. Arthritis Rheum. 2005; 52(11): 3611-7.

[9] Maman E, Bickels J, Ephros M, et al. Musculoskeletal manifestations of cat scratch disease. Clin Infect Dis. 2007; 45(12): 1535-40.

[10] Margileth AM, Wear DJ, English CK. Systemic cat scratch disease: report of 23 patients with prolonged or recurrent severe bacterial infection. J Infect Dis. 1987; 155(3): 390-402.

[11] Hilton-Jones D. Diagnosis and treatment of inflammatory muscle diseases. J Neurol Neurosurg Psychiatry. 2003; 74 Suppl 2: ii25-ii31.

[12] Evans J, Levesque D, Shelton GD. Canine inflammatory myopathies: a clinicopathologic review of 200 cases. J Vet Intern Med. 2004; 18(5):679-91.

[13] Anderson JG, Harvey CE. Masticatory muscle myositis. J Vet Dent. 1993; 10(1): 6-8.

[14] Shelton GD. From dog to man: the broad spectrum of inflammatory myopathies. Neuromuscul Disord. 2007; 17(9-10): 663-70.