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**Case Report**

# Myositis as a de-novo extraintestinal manifestation of ulcerative colitis in a pediatric patient developing post-colectomy: a case report

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## ABSTRACT

This report presents the case of a 13-year-old female with a history of ulcerative colitis who underwent ileostomy closure after total colectomy. Postoperatively, she developed right leg paresis and extensive swelling of the lower extremities. Imaging investigations revealed myositis with avascular necrosis in the distal femoral condyles. These findings were consistent with myositis as an extraintestinal complication of ulcerative colitis.

**Keywords:** Ulcerative colitis, Extraintestinal manifestations, Myositis, Colectomy, Avascular necrosis

## INTRODUCTION

Ulcerative colitis (UC) is an inflammatory bowel disease (IBD) characterized by chronic mucosal inflammation of the rectum that can extend continuously to the proximal segments of the colon. IBD is a systemic disorder that can affect the entire body beyond the gastrointestinal (GI) tract. The involvement of organs and tissues outside the GI tract is referred to as extraintestinal manifestations (EIMs) of IBD.[1] Between 6% and 47% of individuals with IBD develop at least one EIM.[1-3] The most commonly affected organs and associated conditions are joints (peripheral arthritis, axial arthropathies), skin and mucous membranes (pyoderma gangrenosum, erythema nodosum, Sweet's syndrome, aphthous stomatitis), liver (primary sclerosing cholangitis), and the eye (uveitis, episcleritis).[1,4] Myositis has rarely been reported as an EIM of UC. To our knowledge, three cases of myositis associated with UC have previously been reported in the pediatric population. We report the case of a 13-year-old female with UC diagnosed one year prior to developing myositis.

## CASE REPORT

In August 2023, a 13-year-old female was admitted for elective surgery for the closure of an ileostomy after a total colectomy. In April 2022, the patient was diagnosed with left sided UC. Later that year, the general condition of the patient gradually worsened, and she developed hemocolitis, hypoalbuminemia, and severe iron deficiency anemia. Treatment with prednisolone was initiated, but no effect was observed, leading to a therapy escalation with infliximab. In March 2023, the course of the disease had progressed with uncontrollable blood loss and an electrolyte imbalance. Therefore, it was decided to perform a total colectomy and create an ileostomy. Following surgical treatment, the general condition of the patient improved. On August 30th, 2023, transanal residual rectum proctectomy was performed, and an ileoanal anastomosis was created. On the first post operative day, the patient experienced paresis of the right leg. She was unable to lift the leg, bend the knee, or flex the foot. In the left leg, movements and strength were preserved. On the 3rd post operative day, the pain

management epidural therapy was discontinued, and movement in the right leg began to recover. Prophylactic antithrombotic therapy was also initiated.

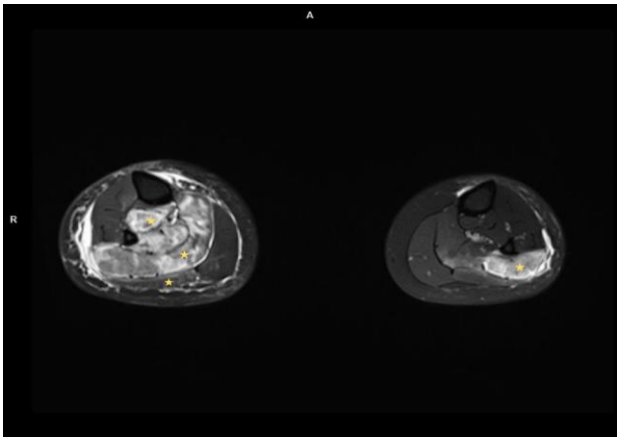


**Figure 1: MRI T2-weighted image (Bilateral edema within the right tibialis posterior muscle, right soleus muscle, and the right gastrocnemius muscle, along with edema in the left soleus muscle: yellow stars).**

On the 5th post operative day, the patient experienced significant swelling in the lower right leg and foot with a decrease in mobility. An ultrasound (US) examination was performed in suspicion of deep vein thrombosis based on symptoms and elevated D dimer levels (4.87 mg/l), but no signs of an intraluminal thrombus were found during the US examination of the right femoral, popliteal, and posterior tibial veins. Other laboratory tests showed mildly elevated levels of C reactive protein (31.03 mg/l). Liver function tests showed elevated alkaline phosphatase at 83.75 U/l and aspartate transferase at 243 U/l. Creatine phosphokinase and lactate dehydrogenase were also elevated at 6200.05 U/l and 495.95 U/l, respectively. Later that day, computed tomography (CT) angiography and CT venography were performed using a 120 ml bolus of Visipaque (320 mg/ml) at a rate of 3.5 ml/s. The CT revealed a lobulated fluid collection measuring 4.3×6.2×16.1 cm (LL×AP×CC) in the right lower leg within the posterior muscles (m. soleus, m. tibialis posterior, and m. gastrocnemius). In the left lower leg, within the posterior muscles, primarily m. soleus, a collection with identical characteristics measuring 4.3×2.2×7.2 cm was observed. The CT scan did not reveal arterial stenosis or venous thrombosis.

On the 7th post operative day, a repeated US examination was performed to determine the precise location for the drainage of the fluid collections. However, conclusive fluid collections were not identified, and it was also impossible to exclude muscle damage. Therefore, further evaluation with contrast enhanced ultrasound (CEUS) was required.

During the CEUS, uniformly perfused calf muscles were identified. Neither intramuscular fluid collection nor muscle necrosis were observed. However, there was a radiological appearance of myositis in both calf muscles. Magnetic resonance imaging (MRI) was indicated for further investigation. On the 14th post operative day, the patient underwent an MRI scan with intravenous contrast enhancement (Clariscan 14 ml, GE Health).

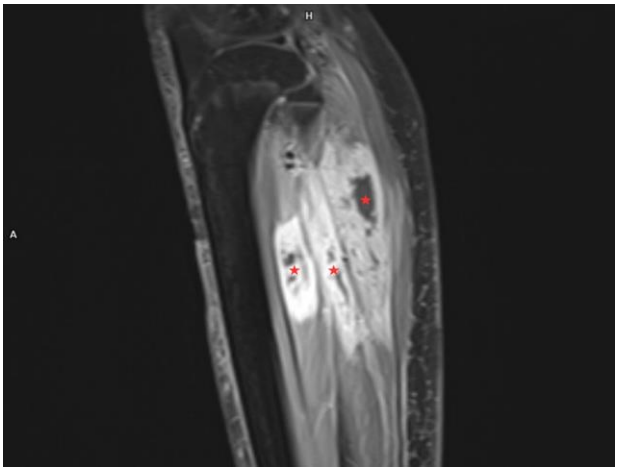


**Figure 2: T1 fat-saturation post-gadolinium sagittal image (Well defined contrast enhancement areas in the right tibialis posterior muscle, right soleus muscle, and right gastrocnemius muscle, along with irregularly shaped non-enhancing areas, indicative of necrotic zones; red stars).**

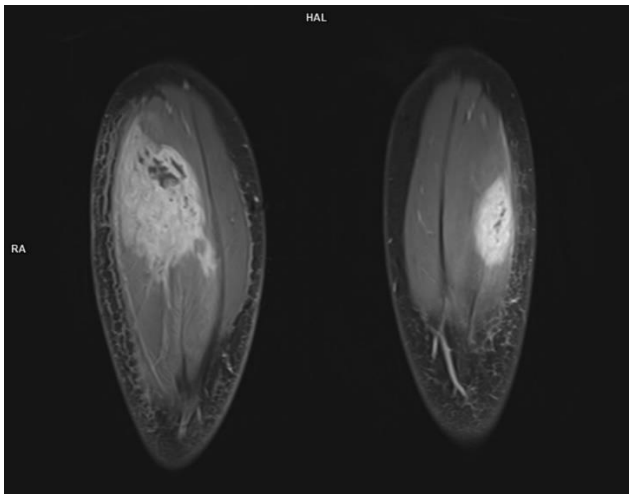


**Figure 3: T1 fat-saturation post-gadolinium coronal image (Well defined contrast enhancement areas along with irregularly shaped non-enhancing areas, indicative of necrotic zones in the right tibialis posterior muscle, right soleus muscle right gastrocnemius muscle, and the lateral head of left gastrocnemius muscle).**

The MRI revealed a well-defined contrast enhancement area measuring 2.0×2.5×5.2 cm in the upper third of the right tibialis posterior muscle, with increased signal intensity in the STIR sequence. There were also irregularly shaped, non-enhancing areas, indicative of

necrotic zones. Areas of similar appearance were also visible within the structure of the right soleus muscle, measuring 1.3×1.6×7.4 cm, and within the right gastrocnemius muscle, measuring 2.6×6.2×8.3 cm. These muscle structures exhibited diffusely increased signal intensity in the STIR sequence, resembling edema. In the lateral head of the left gastrocnemius muscle, a well-defined contrast enhancing area measuring 1.3×2.1×5.3 cm was observed, along with small non enhancing focal lesions (Figure 1-3).

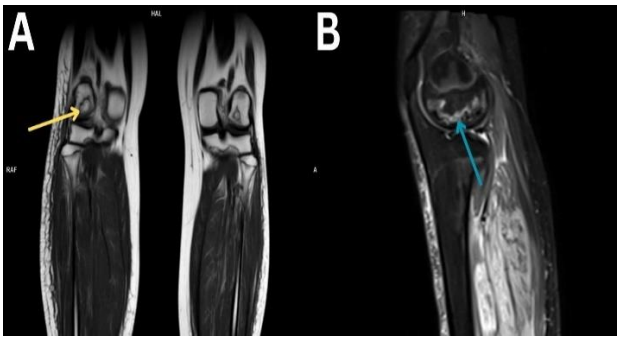


**Figure 4: A) T1 turbo spin-echo; Image B) T2 Dixon Water image (Bilateral avascular necrosis of lateral femoral condyles. Image A shows low signal intensity peripheral contour representing edema (yellow arrow), which appears as a hyperintense line in image B; blue arrow).**

Perifocal muscle tissue involvement and mild subcutaneous soft tissue edema in the lower leg were also observed. Changes in the bones were also observed. In the distal epiphysis of the right femur, there was an irregularly shaped, sharply contoured lesion measuring 4.7×1.6 cm. Also, in the distal epiphysis of the left femur, there was a lesion with identical characteristics measuring 5.3×1.7 cm. The MRI findings suggested bilateral necrotizing myositis with avascular necrosis of the lateral condyles of the distal epiphyses of the femurs.

## DISCUSSION

In this case, the patient complained of swelling and weakness in the lower extremities, and numerous radiological examinations indicated signs of necrotizing myositis and avascular osteonecrosis of the distal femoral condyles. A review of the literature revealed three other cases that describe the correlation between ulcerative colitis and myositis in pediatric patients. The data gathered from published case reports has been summarized in (Table 1). All three patients developed myositis simultaneously with the diagnosis of UC, and only in our case did the patient develop myositis 1 year after the initial UC diagnosis.[5-7]

The diagnosis of myositis should be considered in all patients with IBD who complain of myalgia, muscle weakness, and edema, as these were the most commonly encountered symptoms in the described patients. A muscle biopsy is the most important step in the diagnostic process of myositis, and it is essential to distinguish between the various subtypes of this disease.[8-9] Unfortunately, in our case, the diagnosis of myositis was made based on a combination of the patient's symptoms, the findings of the neurological examinations, and the MRI results. A muscle biopsy was not performed, which could have further supported the diagnosis.

**Table 1: Literature on pediatric case reports collected through PubMed using the keywords “myositis” and “ulcerative colitis” from 1986 to 2023.**

| Author | Age | Sex | Myositis | Sx of myositis | Myositis Sx onset (from Dx of UC) | Dx modality | Involved muscles |
|---|---|---|---|---|---|---|---|
| **Hernández et al[5]** | 11 | M | PM | Pain, weakness | Simultaneous | Muscle biopsy, EMG | Proximal lower/upper extremities, neck flexors |
| **Ishiwada et al[6]** | 14 | F | Undifferentiated myositis | Pain, swelling, fever | Simultaneous | CT, MRI (biopsy refused) | Masseter muscles |
| **Kim et al[7]** | 14 | F | PM | Pain and swelling | Simultaneous | Muscle biopsy, MRI | Lt. masseter, deltoid muscle, teres minor, infraspinatus muscle |

M, male; F, female; Sx, symptoms; Dx, diagnosis; PM, polymyositis; EMG, electromyography; CT, computed tomography; Lt, left; Rt, right; CEUS, contrast-enhanced ultrasound;

Our patient developed myositis six months after colectomy as a de novo EIM. One of the aims of a retrospective study conducted by Roth et al was to investigate whether colectomy reduces the incidence of de novo EIMs. The study revealed that 13.5% of IBD patients develop a de-novo EIM after colectomy; therefore, in patients who never experienced EIMs prior to colectomy, de novo manifestations should be expected in up to one in seven patients.[10]

An additional finding in this patient's MRI scan indicated osteonecrosis (aseptic necrosis) of the lateral condyles of the distal epiphyses of both femurs. We believe that this patient developed osteonecrosis (aseptic necrosis) due to the use of steroids as we excluded other risk factors such as congenital defects (Legg Perthes Calve disease, slipped capital femoral epiphysis, and congenital hip dislocation), iatrogenic risk factors (cigarette smoking, alcohol consumption), infections, metabolic conditions, coagulopathies, rheumatologic diseases, and malignancies.[11] Osteonecrosis is a known side effect of systemic steroids, which are used to treat a variety of inflammatory conditions, including UC. Osteonecrosis in patients with UC most frequently affects the femoral head and is usually bilateral, although it can also affect the proximal humerus, proximal tibia, talus, scaphoid, patella, metatarsal heads, and femoral condyles.[12-14] This patient developed osteonecrosis in the lateral condyles of the distal epiphyses of both femurs after taking oral prednisolone for a total of 69 days at a daily dose of 60 mg.

The cumulative lifetime dose of prednisolone was calculated to be 4140 mg. There is no clear relationship between the dose of steroids a person takes and their risk of developing osteonecrosis.[13] Koo et al reported that until the diagnosis of osteonecrosis on MRI, the total dose of steroids received ranged from 1800 to 15,505 mg of prednisolone or its equivalent, with a mean dose of 5928 mg.[15]

As per treatment, our patient only received subcutaneous enoxaparin administered twice daily (80 mg/day), and no other therapy was administered due to the observed rapid improvement of symptoms. Within two days, the previously noted right leg paresis had resolved. Subsequently, swelling of the right leg developed, yet no neurological deficit or pain were observed. The patient received physical therapy throughout the entire hospitalization period to improve mobility in the right ankle joint as well as maintain active movement in the knee and hip joints. The patient was discharged from the hospital on the 14th post operative day in satisfactory condition, with a recommendation to continue physical therapy on an outpatient basis.

The authors would like to emphasize that significant differential diagnosis in this case is bilateral compression neuropathy of the tibial and peroneal nerves due to post operative compartment syndrome after surgery performed in supine position. A study by Warner et al., shows that compartment syndrome is more common in patients undergoing surgery in lithotomy and lateral decubitus positions, however it also develops in patients after surgeries in supine position and the reported frequency is 1 per 92,441 surgeries.[16] Although compartment syndrome after surgery in supine position is rare, this differential diagnosis cannot be excluded in this case. Therefore, a follow up MRI scan is scheduled to support or exclude this differential diagnosis.

## CONCLUSION

This case highlights the importance of considering myositis as a differential diagnosis in patients with UC who present with muscle weakness, pain, and edema, especially in the setting of elevated creatine kinase levels. Muscle biopsy remains the gold standard for diagnosis, but MRI findings may be useful in conjunction with the clinical presentation. Osteonecrosis can be an additional complication associated with UC, potentially due to steroid use.

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