

# Juvenile Dermatomyositis in Remission Complicates Appendicitis: A Case of Intraabdominal Appendiceal Abscess Tracking to the Foot

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<b>Background</b>	A child with a history of Juvenile dermatomyositis (JDM), quiescent for seven years on no medications, presented with severe right lower extremity and pelvic pain.
<b>Summary</b>	A 16-year-old female child was admitted to the children's hospital with intractable right lower quadrant pain. Blood serum laboratory values indicated myositis. Imaging demonstrated diffuse muscle and soft tissue inflammation with multi-loculated abscesses tracking down the ileo-femoral artery distribution in the pelvis, right thigh and calf requiring extensive debridement and drainage to control sepsis. This is the first reported case of a ruptured appendix with multiple pelvic and right lower extremity abscesses tracking down the femoral canal to the foot.
<b>Conclusion</b>	Complications from JDM remain life threatening years after clinical remission is achieved. Urgent imaging and aggressive surgical drainage should be undertaken to resolve sepsis.
<b>Keywords</b>	juvenile dermatomyositis JDM, appendicitis, abscess, leg, inflammatory myopathy

**DISCLOSURE STATEMENT:**

The authors have no conflicts of interest or financial disclosures.

**To Cite:** Bazan VM, Savage C, Mercado-Deane M, Zwischenberger JB. Juvenile Dermatomyositis in Remission Complicates Appendicitis: A Case of Intraabdominal Appendiceal Abscess Tracking to the Foot. *ACS Case Reviews in Surgery*. 2020;2(6):38-42.

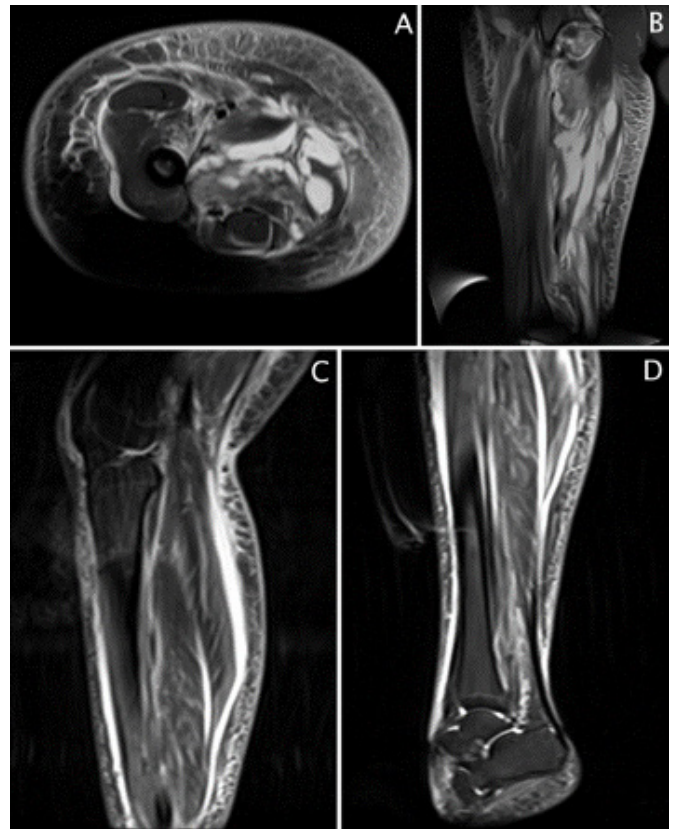
## Case Description

Juvenile dermatomyositis (JDM) is an autoimmune disease manifested by skin lesions and proximal muscle weakness. Diagnosis is made by clinical presentation, muscle biopsy demonstrating inflammatory infiltrate, perifascicular atrophy and connective tissue fibrosis, and blood studies showing elevated muscle enzymes and inflammatory markers. Initial therapy is systemic steroids, with additional options including intravenous gamma globulin and methotrexate to achieve remission.<sup>1,2,3</sup>

Once remission is achieved, sequelae from the muscle and soft tissue inflammation may result in residual fatty replacement and atrophy of the muscle and connective tissue fibrosis.<sup>4</sup>

A 16-year-old female was admitted to our children's hospital for intractable right lower extremity pain, edema, fever, elevated white blood count, and elevated serum creatinine kinase. The patient reported a 2-week history of pain that began in the mid abdomen and migrated to the lower extremity. Bedside ultrasound was remarkable for diffuse thigh cellulitis and fluid collections. Emergent magnetic resonance imaging (MRI) was performed demonstrating multi-loculated thigh abscesses, with profound muscle and soft tissue inflammatory changes extending from the femoral canal to the knee (Figure 1A, 1B). In the operating room, a 27-cm medial thigh incision drained 400 cc of foul-smelling purulent drainage from a collection extending from the anterior hip capsule to the knee. Gram stains and culture indicated *Escherichia coli* (*E. coli*). There was significant tissue and fascial inflammation with necrosis of the sartorius, pectineus, and adductor magnus, longus, and brevis muscles, necessitating extensive debridement. Following irrigation and debridement, a vacuum assisted wound closure device was placed over the medial thigh. Calf swelling noted during surgery, prompted MRI of the calf demonstrating inflammation and fluid. (Figure 1C, 1D). On hospital day 2, surgery for additional washout yielded additional foul-smelling necrotic tissue from the thigh, necessitating a 37-cm right lateral thigh incision with irrigation and debridement. Lower leg fasciotomy with a 17-cm right lateral incision and a 15-cm medial leg incision exposed all four compartments, with additional purulent drainage. Medial and lateral vacuum assisted wound closure devices were placed on the thigh and leg. Following washout, abdominal and pelvic computed tomography (CT) was obtained to evaluate the abdomen and pelvis. CT showed a dilated appendix with appendicoliths and multiple pelvic abscesses from ruptured appendi-

citis (Figure 2). Interventional radiology placed a 10 French right lower quadrant drain, obtaining 20 cc of purulent drainage, likewise growing *E. coli* on culture. Her sepsis progressed, requiring inotropic support with epinephrine and norepinephrine. On day 3, patient was evaluated for appendectomy. She was considered too unstable for laparotomy, therefore, using an extraperitoneal approach, a 10-cm flank incision surgically drained a right iliopsoas abscess. Three separate extraperitoneal Penrose drains (1-inch) were then placed in the retrorenal area, through the femoral triangle, and in the subcutaneous tissue.



**Figure 1.** MRI imaging. Right thigh (A) axial and (B) sagittal STIR weighted images demonstrate multiloculated abscesses in the posterior thigh, inflammation of the adductor magnus, adductor longus, sartorius, gracilis, semimembranosus and vastus medialis. There is diffuse soft tissue edema. MRI of the right calf sagittal (C) upper calf and (D) lower calf STIR-weighted images demonstrate extensive subcutaneous and muscular edema. There is fluid within the crural fascia.



**Figure 2.** CT abdomen and pelvis with contrast. (A) Axial image demonstrates multiple abscesses (\*), free fluid(x), inflammatory changes of the subcutaneous and intra abdominal fat and enlarged pelvic appendix (arrow). (B) Coronal image shows dilated appendix with appendicolith (arrow). Postoperative intramuscular air seen (>).

On hospital day 7, as sepsis resolved, the wounds were clean and exhibited granulation tissue. Washout and wound closure of the medial and lateral leg and partial closure of the lateral thigh was uncomplicated. Likewise, over the following 4 days, the remaining open wounds were sutured closed. No further surgical interventions were performed. On hospital day 28, she was transferred to the rehabilitation floor, then discharged on hospital day 47. The patient’s hospital course is summarized in Table 1. An uncomplicated interval appendectomy was performed three months later. Findings at surgery included a long appendix with the midpoint adherent to the retroperitoneum.

Hospital Day	Procedure	New Findings
1	Hospital admission MRI of the right thigh Incision and drainage of medial thigh	Multiloculated thigh abscesses, profound muscle and soft tissue inflammatory changes extending to the knee  Tissue necrosis, significant fascial inflammation  Cultures positive for <i>Escherichia Coli</i>
2	MRI of right calf Incision and drainage of: right lateral thigh, right lateral leg, medial leg and drainage of medial thigh CT of abdomen/pelvis Interventional Radiology placed 10 French drain for iliopsoas abscess Intubation/ventilation initiated	Inflammation extending to distal leg, fluid in the crural fascia  Ruptured appendicitis with intra-abdominal abscess  Septic shock
3	Surgical drainage of iliopsoas abscess with insertion of 3 Penrose drains	
4	Washout and drainage of thigh and leg incisions	
5	Weaned off of sedation and pressor support	
7	Closure of medial and lateral leg incisions	
9	Closure of lateral thigh incision	
11	Closure of medial thigh incision with placement of 2 right lower quadrant Penrose drains	
28	Transfer to rehabilitation floor following wound care	
47	Discharged with plan for interval appendectomy	

**Table 1.** Timeline of Hospital Course

## Discussion

Ruptured appendicitis with contamination of the ipsilateral lower extremity is recognized in cases of retrocecal appendix, primarily affecting older or immunocompromised patients (Table 2). The retrocecal appendix is in close proximity to the psoas, allowing infection from a ruptured appendix to track along tissue planes following the psoas muscle into the thigh as it inserts on the lesser trochanter of the femur.<sup>5</sup> A thigh abscess resulting from rupture of a non-retrocecal appendix positioned in the right pelvis (Figure 2), as in this case, is unreported in the literature.<sup>6</sup> Furthermore, ruptured appendiceal abscess tracking along the femoral vessel all the way to the foot is unreported.

The only predisposing factor in the patient's medical history was dermatomyositis at age 5, treated with steroids and methotrexate. Of the three clinical courses described for JDM (chronic, polycyclic and monocyclic) this patient was categorized as monocyclic due to the successful treatment of JDM with no further recurrences.<sup>7</sup> A prospective cohort study found the most common clinical course for JDM is chronic (60%) followed by monocyclic (37%) and least commonly polycyclic (3%).<sup>7</sup> She had been in remission and off steroids for seven years. We suspect residual changes in muscle and fascia from the original JDM episode predisposed the patient to infection spread down the soft tissue planes to the foot. Connective tissue fibrosis also may have compromised the normal separation of compart-

	Age	Abscess Location	Appendix Location	Complications	Treatment*	Survived
Edwards 198610	76	Psoas to thigh	Retrocecal	Myositis, necrosis, sepsis	Surgical drainage	No
Dheer, 200111	57	Gluteal/abdominal wall to knee	Retrocecal	Myositis, subcutaneous emphysema, necrosis	Surgical drainage and debridement	Not reported
El-Masry 200212		Psoas to thigh	Retrocecal		Appendectomy Surgical drainage	Yes
Sharma 200513	6	Psoas to thigh	Retrocecal	Myositis, sepsis	Appendectomy Surgical drainage	Yes
Ushiyama 200514	83	Psoas to thigh	Retrocecal	Myositis, subcutaneous emphysema	Appendectomy Curettage	Yes
Hsieh 20065	56	Psoas to thigh	Retrocecal	Myositis, subcutaneous emphysema	Appendectomy Surgical drainage	Yes
Yildiz 200715	27	Inguinal ligament to groin	Retrocecal	Sepsis	Appendectomy Surgical drainage and debridement	Yes
Sookraj 200916	67	Psoas to thigh	Retrocecal	Necrosis	Appendectomy Curettage	Yes
Lal 201217	40	Gluteal to thigh	Retrocecal	Subcutaneous emphysema	Appendicectomy Surgical drainage Ileotransverse colectomy	Yes
English 201218	56	Iliacus to thigh	Retrocecal	Necrosis	Surgical drainage and debridement Interval appendectomy	Yes
Nanavati 201519	53	Gluteal/psoas to thigh	Retrocecal		Surgical drainage Ileocolostomy	Yes
Naidoo 201620	50	Psoas to knee	Retrocecal	Necrosis, sepsis	Appendectomy Surgical drainage and debridement	No
Van Hulsteijn 20176	73	Iliopsoas to thigh	Retrocecal	Myositis	Surgical drainage Ileoascendostomy	Yes
Case Report	16	Psoas to foot	Not retrocecal	Myositis, necrosis, sepsis	Surgical drainage and debridement Interval appendectomy	Yes

**Table 2.** Cases of appendicitis resulting in lower extremity abscess

ments.<sup>8</sup> In this case, MRI evidence of atrophy and fatty deposits may have been obscured by swelling and the fluid signal from edema. While the patient experienced unilateral leg weakness, muscle inflammation, and laboratory values consistent with active myositis, laboratory values did not confirm recurrence of juvenile dermatomyositis.

## Conclusions

Although the most common pediatric inflammatory myopathy, JDM still remains rare and clinical implications of remission poorly understood.<sup>9</sup> Clinicians must remain cognizant of potential life-threatening complications from JDM even when the disease has been clinically quiescent, and patients are not on steroid therapy.<sup>10-20</sup>

## Lessons Learned

Based on this case experience, we recommend comprehensive imaging to delineate the sites of infection to direct aggressive surgical drainage as necessary to resolve progressive sepsis. Earlier determination of the source of infection, including cultures positive for organisms directing an abdominal or rectal source, allows strategies for drainage to minimize progression.

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