CASE REPORT

Relapsing retroperitoneal abscess secondary to juvenile dermatomyositis: Complexity in management

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SUMMARY
Juvenile dermatomyositis (JDM) is a systemic autoimmune condition with myopathy. Gastrointestinal and pulmonary manifestations are rare presentation of JDM. Gastrointestinal perforation incidence in JDM is associated with vasculopathy and ischaemia. There are only few reported case of management of JDM with gastrointestinal complication. Management of such condition is challenging. We present a 21-year-old man with spontaneous descending colon perforation undergoing Hartmann’s procedure. He subsequently presented with recurrent retroperitoneal abscess at five and 30 months following the initial presentation which was treated with percutaneous drainage. A high index of suspicion is necessary in JDM patients presenting with acute abdomen.

CASE PRESENTATION
A 21-year-old man diagnosed with Juvenile dermatomyositis (JDM) since 9-year-old presented with left iliac fossa pain for three days, high grade fever, right wrist stiffness and facial rashes. He was receiving regular treatment of methotrexate, prednisolone and hydroxychloroquine from rheumatologist. He has also been on interim intravenous immunoglobulin three days, high grade fever, right wrist stiffness and facial rashes. He was receiving regular treatment of methotrexate, prednisolone and hydroxychloroquine from rheumatologist.

On examination, his abdomen was found to be tender at the left iliac fossa region, however, it was not distended and no mass was felt. Initial diagnosis of left pyelonephritis was made following abdominal ultrasonography. As his condition was not improving, contrast enhanced computed tomography (CT) of the abdomen (image not shown) was performed three days later revealing loculated air containing collection at left iliac fossa, posterior to distal descending colon and retroperitoneum. A laparotomy was performed, it revealed a 2cm perforation posterior of proximal descending colon with a localized feculent peritonitis seen at left paracolic gutter and retroperitoneum inferior to the left kidney. Mannheim Peritonitis Index score was 16 for which Hartmann’s procedure and peritoneal lavage was performed. Patient had steady recovery and was discharged seven days later.

He was able to tolerate orally well until he presented again five months later with fever, lower abdominal pain and left inguinal erythematous swelling associated with pus discharge. Contrast enhanced CT abdomen (Figure 1a, Figure 1b) shows loculated rim enhancing collection measuring 7.8X4.0X8.0cm at left retroperitoneal region extending to left iliopsoas and left inguinal region. Ultrasound guided percutaneous drainage of the collection was done using 10Fr pigtail catheter.

Patient was managed by a multidisciplinary teams which included the surgical, rheumatology, intervention radiologist, nutritional support team and physiotherapist. He was given multiple courses of intravenous immunoglobulin and subcutaneous methotrexate. A repeated ultrasound two weeks after the drainage revealed 2.3x3.6x8.0cm collection inferior to left kidney and 2.1x3.6x4cm at left inguinal. Two days upon removing the drain, he developed septic shock requiring inotropic support with persistent pus discharge from left inguinal swelling. Repeated unenhanced CT abdomen with oral contrast (figure 2a, 2b) was performed due to suspicion of enterocutaneous fistula. CT showed residual left retroperitoneal, iliopsoas abscess with left inguinal extension and a fistula demonstrated between jejunum and left iliopsoas collection. He was treated based on principles of enterocutaneous fistula with sepsis control with image guided drainage, antimicrobial and total parenteral nutrition. Interval imaging done after four months displayed spontaneous healing of the fistula. He was allowed orally and observed for two weeks before discharge.

He has a short stature with height of 1.5m and his weight on follow up improved significantly from 1st admission 32kg (BMI 14.2) to 36.5kg (BMI 16.2) at six months following discharge. He is now 42.2kg (BMI 18.7) three years following his surgery. He is still treated with low dose prednisolone and methotrexate.

He remained asymptomatic for 15 months, thereafter, presented again with left inguinal subcutaneous collection with retroperitoneal extension along left iliopsoas and iliopsoas muscle. Image guided drainage done, and he was discharged home well.

DISCUSSION
JDM is rare idiopathic autoimmune myopathy with systemic vasculopathy in childhood. The incidence is reported at 2-4 cases per million with female predominance. They commonly present at 5-10 years of age. Classical initial presentation of JDM are muscle weakness, Gottron’s rash,
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heliotrope rash, nail fold capillary changes, calcinosis and joint pain. Systemic presentation such as interstitial lung disease and gastrointestinal vasculopathy are less common and life threatening. The treatment goals are to control inflammatory myositis and prevent complication arising from disease. The treatment varies with evaluation of disease process and severity of disease. The main course of treatment is combination corticosteroids and methotrexate.

Gastrointestinal vasculopathy may present with abdominal colic, ulceration, bleeding and perforation. Presentation with spontaneous bowel perforation is rare and occurs in severe JDM. The pathophysiology is secondary to vasculopathy and ischemia of bowel wall. Common site of perforation is at duodenum 25%, right colon 17.1%. Management of JDM patient is challenging due to chronic consumption of corticosteroids and malnourished state. The presentation may be masked and delayed due to immunosuppressed state, as in our case. Acute abdomen in JDM patients should not be taken lightly and high index of suspicion needed. Abdominal x-ray may not be conclusive as perforation may be retroperitoneal as in our case, small or sealed. Pneumatosis intestinalis should be highly suspected with close monitoring of clinical condition and serial abdominal x-ray. There have been few successful survivors with medical treatment and bowel rest.

Intervention approach for gastrointestinal perforation associated to JDM should be carefully chosen. Exploratory laparotomy is an ideal option in most gross peritonitis condition, however there would be remarkable post-operative stress and recovery on labile JDM patients. Minimal access surgery may prove to be a good modality of treatment with minimal disruption to anatomy and patient recovery even in emergency condition. As this disease is lifelong, potential future intra-abdominal complication can be dealt with ease if minimal access surgery was offered. In our case, poor understanding about the nature of disease due to rarity affected our choice of intervention at first presentation.
Image guided drainage can be performed in selective acute setting as definitive or as a bridge to definitive surgical procedure. Considering the nature of the disease, image guided drainage is an ideal option and should be carefully considered depending on location and extent of collection. In our case, fistulous jejunal opening was seen tracking to ilio-psoas due to exposed retroperitoneum space following dissection during Hartmann’s procedure appears to be peculiar. We opt for percutaneous drainage of retroperitoneal collection and principle of treatment of enterocutaneous fistula was applied in our management.

CONCLUSION
Gastrointestinal vasculopathy in JDM is uncommon complication and management is challenging. A high index of suspicion is needed in acute abdomen scenario. Multidisciplinary approach is vital for best outcome. Due to nature of disease and potential new gastrointestinal perforation, minimal access surgery offers immediate and long-term benefit over laparotomy.

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ETHICAL APPROVAL
There is no ethical approval is required for this case report.

CONSENT
Written informed consent was obtained for this case report publication.

CONFLICT OF INTEREST
There is no conflict of interest.

REFERENCES