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Case report

Mesenteric vasculitis in a juvenile systemic lupus erythematosus patient

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ABSTRACT

Lupus mesenteric vasculitis (LMV) is a rare cause of acute abdominal pain. Few cases of LMV have been reported in adults, children and adolescents. However, to our knowledge, the prevalence of that severe vasculitis in a pediatric population with lupus is yet to be studied. This study reviewed data from 28 consecutive years and included 5,508 patients being followed up at the hospital of the Faculdade de Medicina of the Univesidade de São Paulo (FMUSP). We identified 279 (5.1%) patients meeting the American College of Rheumatology classification criteria for the diagnosis of systemic lupus erythematosus (SLE), one of whom (0.4%) had LMV. That male patient was diagnosed with SLE at the age of 11 years. At the age of 13 years, he was hospitalized with diffuse and acute abdominal pain, nausea, bilious vomiting, abdominal distension, rebound tenderness, and abdominal muscle guarding. The patient underwent laparotomy immediately, and segmentary intestinal ischemia with intestinal wall edema and adhesions were identified. Partial small bowel resection with lysis of the adhesions was performed, as were pulses of intravenous methylprednisolone. The histopathologic analysis evidenced mesenteric arteritis. After 13 days, the diffuse and intense abdominal pain recurred, and the patient underwent a new laparotomy, during which adhesive small bowel obstruction with intestinal gangrene was identified. New intestinal resection was performed, and the patient received pulses of intravenous methylprednisolone and infusion of immunoglobulin. Thus, LMV is a rare and severe abdominal manifestation of the pediatric population with lupus, and can be the only manifestation of disease activity. In addition, this study stresses the importance of the early diagnosis and immediate treatment.

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Vasculite mesentérica em paciente com lúpus eritematoso sistêmico juvenil

Palavras-chave:
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RESUMO

A vasculite mesentérica lúpica (VML) é uma rara causa de dor abdominal aguda. Há poucos relatos de caso demonstrando VML em adultos e, particularmente, em crianças e adolescentes. No entanto, para o nosso conhecimento, a prevalência dessa grave vasculite em uma população pediátrica com lúpus ainda não foi estudada. Portanto, dados de 28 anos consecutivos foram revisados e incluídos 5.508 pacientes em seguimento no Hospital da Faculdade de Medicina da Univesidade de São Paulo (FMUSP). Identificamos 279 (5,1%) casos que preencheram critérios de classificação diagnóstica do American College of Rheumatology para lúpus eritematoso sistêmico (LES) e um (0,4%) desses apresentou VML. Este paciente recebeu diagnóstico de LES aos 11 anos de idade. Aos 13 anos foi hospitalizado com dor abdominal difusa e aguda, náuseas, vômitos biliosos, distensão e rigidez abdominal, com descompressão brusca positiva. O paciente foi prontamente submetido à laparotomia exploradora, identificando isquemia intestinal segmentar, com edema de parede intestinal e aderências. Foi realizada ressecção parcial de intestino delgado, com lise das aderências e pulsoterapia com metilprednisolona. A análise histopatológica identificou arterite de vasos mesentéricos. Após 13 dias, apresentou recorrência de dor abdominal difusa intensa, sendo novamente submetido à laparotomia exploradora, identificando obstrução em intestino delgado por aderências, com gangrena intestinal. Nova ressecção intestinal foi realizada, além de pulsoterapia com metilprednisolona e infusão de imunoglobulina. Portanto, VML é uma rara e grave manifestação abdominal na população com lúpus pediátrico, e pode ser a única manifestação de atividade da doença. Além disso, este estudo reforça a importância do diagnóstico precoce e do tratamento imediato.

Introduction

Systemic lupus erythematosus (SLE) may affect multiple organs and systems, such as gastrointestinal involvement.[1,2] Digestive tract manifestations in SLE patients are generally caused by treatment related to adverse events, infections,[3] and disease activity.[2,4]

Of note, lupus mesenteric vasculitis (LMV) is a cause of acute abdominal pain, associated with nausea, vomiting, diarrhea in SLE patients usually with disease activity.[4] The diagnosis of LMV requires the evaluation of bowel wall and the abdominal vasculature by image examination,[4] such as abdominal ultrasound,[3,5,6] computer tomography scan,[3,7,8] magnetic resonance image,[6] digital arteriography,[8] and/or particularly histopathological findings.[4]

A few case reports have demonstrated LMV in adult SLE[7,8,10] and particularly in juvenile SLE (JSLE) patients.[5,6,9,10] However, to our knowledge, the prevalence of this severe vasculitis in paediatric lupus population has not been studied.

Therefore, from January 1983 to December 2010, 5,508 patients were followed-up at the Paediatric Rheumatology Unit of the Instituto da Criança, Hospital das Clínicas, Faculdade de Medicina, Universidade de São Paulo (ICr-HC-FMUSP) and 279 (5%) of them were acquainted to the American College of Rheumatology (ACR)[11] classification criteria for SLE. Only one (0.4%) of our JSLE patients had LMV, confirmed by the description of histopathological findings and required intestinal resections. This study was approved by the Local Ethics Committee of FMUSP.

Case report

An 11-year old boy was diagnosed with JSLE based on malar rash, arthritis, pericarditis, psychosis, lymphopenia, and thrombocytopenia, and positivity of the following autoantibodies: antinuclear antibodies (ANA) 1:1280 (speckled pattern), anti-Sm, and anti-double stranded DNA (anti-dsDNA) antibodies. At that moment, the SLE Disease Activity Index 2000 (SLEDAI-2K)[12] was 19 and he received three pulses of intravenous methylprednisolone, intravenous cyclophosphamide (500–1,000 mg/m^2/month for 24 months) and prednisone (2.0 mg/kg/day), with progressive dose decrease to 7.5 mg/day. At the age of 13, he was hospitalized due to an acute diffuse abdominal pain, nausea and bilious vomiting. On physical examination, he had abdominal distension, rebound tenderness and abdominal muscle guarding, compatible with acute surgical abdomen. At that moment, he was under 7.5 mg/day of prednisone and abdominal ultra-sound examination showed mild ascites, diffuse distension and bowel-wall thickening. Laboratory tests showed hemoglobin 13.5 g/L, hematocrit 40%, white blood cell count 5,400/mm^3 (72% neutrophils, 21% lymphocytes, 4% eosinophils and 3% monocyte), platelets 265,000/mm^3, proteinuria (0.02 g/24h), urinalysis – leukocytes 500 high-power field, and erythrocytes 250 high-power field, urea 19 mg/dL (normal range 10–42), creatinine 0.5 mg/dL (normal range 0.5–0.9), C3 0.86 mg/dL (normal range 0.5–1.8), C4 0.13 mg/dL (normal range 0.1–0.4), and amylases 46 U/L (normal < 106 U/L). The erythrocyte sedimentation rate was 42 mm in the first hour. Immunological tests revealed negative for anti-dsDNA, lupus anticoagu-

lant, and IgG and IgM anticardiolipin antibodies. The patient immediately underwent a laparotomy and segmental edema with adhesions and small bowel infarctions were identified. Partial small bowel resection (4 cm vs. 2.5 cm) was performed with lysis of adhesions. The histopathology showed submucosa edema with a diffuse inflammatory infiltrate of mononuclear cells, and mesenteric vessels arteritis. At that moment, his SLEDAI-2K[12] was 8 and he received three pulses of intravenous methylprednisolone. After four days, he was transferred to out-patient clinic of FMUSP receiving 10 mg/day of prednisone. Thirteen days later, he had recurrence of severe and diffuse abdominal pain and distension, with bilious vomiting and acute diarrhea. A plain radiography of the abdomen showed diffuse bowel distension with air-fluid level compatible to partial intestinal occlusion. The patient was promptly submitted to a new laparotomy which identified adhesive small bowel obstruction with intestinal gangrene. Remarkably, resections of 12 cm vs. 6.5 cm of small bowel and 12 cm vs. 7 cm of caecum, lysis of adhesions and terminal ileostomy were also performed. At that moment, he received three pulses of intravenous methylprednisolone and a single infusion of intravenous immunoglobulin (2 g/kg/day), followed by 10 mg/day of prednisone, with rapid recovery and improvement of his gastrointestinal symptoms. At 13.5 years, loop ileostomy closure was carried out and the SLEDAI-2K[12] was 0. The Systemic Lupus International Collaborating Clinics of the American College of Rheumatology – Damage Index (SLICC/ACR-DI)[13] was 2 and he was receiving prednisone 5 mg/day. No complications after intestinal resections were observed.

Discussion

We described a case of LMV followed at the tertiary Paediatric University Hospital that evidenced the rare occurrence of this severe vasculitis as a cause of permanent disease damage. Moreover, this complication required multiple intestinal resections without late bowel complications.

LMV in adult patients has been reported from 0.2% to 9.7%,[3,4,8] with very few cases described in paediatric lupus population.[5,6,10,14] This complication is a life-threatening abdominal manifestation characterized by bowel ischemia, with high mortality rates up to 50%.[3,4,8] Usually, the abdominal pain triggered by LMV shows a diffuse pattern and is generally associated with nausea, vomiting, diarrhea, abdominal distension, rebound tenderness and abdominal muscle guarding,[3,4,6–8] as observed in this case . In addition, the occurrence of acute abdomen that required intestinal resections was rarely reported in lupus adolescent population,[14] as also evidenced herein.

The signs and symptoms of LMV are non-specific, and radiological investigations[3,4,6,8] or pathological findings[3,4,6] are necessary to confirm the diagnosis. Abdominal ultrasonography can demonstrate small intestinal edema and thickening of the bowel-wall.[3] In fact, the patient's ultrasound revealed mild ascites, diffuse distension and bowel-wall thickening. Remarkably, the LMV histopathology evidences sub-mucosal edema, diffuse inflammatory infiltrate of mononuclear cells, and both small vessel arteritis and venulitis.[4]

Generally lupus patients with mesenteric vasculitis have active disease with high SLEDAI scores,[4,7,8] associated with nephritis,[6,7,8] neuropsychiatric involvement,[6] and/or hematological abnormalities.[7] The presence of LMV with low disease activity was rarely described.[6] The pathogenesis of LMV is unknown, but it is suggested that it involves a vasculitis with immune-complex deposition and/or intestinal vessels thrombosis[3] leading to mesenteric ischemia.[3,4] Autoantibodies, such as antiphospholipid and anti-endothelial cell antibodies, may also contribute in the immunopathogenesis of this complication.[3,4]

Of note, LMV requires prompt diagnose and appropriate treatment with corticosteroids,[15] particularly intravenous infusion of methylprednisolone, and immunosuppressive therapy in non-responsive cases.[3,4,6–8] The patient was previously treated with intravenous cyclophosphamide, therefore, we used intravenous immunoglobulin associated with corticosteroid.

Furthermore, as utilized in this present case, early surgical treatment should be indicated in JSLE patients with extensive bowel ischemia and/or perforation, reducing the risk of morbidity and mortality.[3,4,6] The patient had an additional complication with adhesive small bowel obstruction and intestinal infarction, requiring extensive bowel resections without any further complications, possible related to low prednisone dose after the first surgery. Moreover, corticosteroids have already been tested for the prevention of abdominal adhesions caused by surgical procedures, with limited results.[16]

In conclusion, LMV should be evaluated in all children and adolescents with acute abdominal pain and may be an isolated manifestation of lupus activity. This study reinforces the importance of early diagnosis and prompt treatment for this severe gastrointestinal vasculitis.

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Conflicts of interest

The authors declare no conflicts of interest.

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