Case Report

Lupus Peritonitis with Nephritis Post-Surgery : A Rare Cause of Ascites

Vishal V Ramteke[1], Afzal S Shaikh[2], Prakash Khetan[3], Tanzilur Rehman[4]

ABSTRACT

Background : Serositis is common in patients with systemic lupus erythematosus (SLE) presenting as pleural or pericardial inflammation. Peritoneal serositis and massive ascites also called as lupus peritonitis is extremely rare. It can present as the first manifestation of SLE or can be noted in diagnosed case of lupus.

Case Presentation : We report a case of female patient with SLE presenting with massive ascites following laparoscopic hysterectomy. On evaluation it was exudative with lymphocytic predominance and serology were suggestive of lupus activity in the form of decreased complements and Anti-Nuclear Antibody (ANA) and Double stranded DNA (DsDNA) positivity. She had lupus nephritis which as evident from new onset proteinuria and hematuria. Intravenous methylprednisolone followed by oral prednisolone, cyclophosphamide and hydroxychloroquine led to significant improvement in features of peritonitis and glomerulonephritis.

Conclusions : Lupus peritonitis is severe manifestation of peritoneal inflammation in patients with SLE. It is a diagnosis of exclusion and treatment steroids with cyclophosphamide and hydroxychloroquine lead to good recovery.

Key-words : Systemic lupus erythematosus, Lupus peritonitis, Lupus nephritis, Laparoscopic hysterectomy, Diagnostic laparoscopy.

Introduction :

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease characterized by autoantibodies against nuclear antigens and deposition of immune complexes in several tissues.[1,2] Serositis is a common finding noted in 16% of SLE patients mostly with pleuropericardial involvement.[3] Though peritoneal inflammation is observed in two thirds of the patients on necropsy, clinical manifestation of massive ascites, abdominal pain and ileus is extremely rare and seen in only 4 % of patients.[4,5]. Lupus peritonitis as a presenting feature of SLE is an even rarer occurrence. We describe here a case of a female patient with lupus peritonitis following surgical interventions who eventually responded to steroids and cyclophosphamide. We intent to report the management of this rare manifestation of SLE.

[1]Consultant Nephrologist, [2]Consultant Critical Care Medicine,
[3]Consultant Nephrologist, Kingsway Hospital, Nagpur
[4]Consultant Urologist, Seven Star Hospital, Nagpur

Address for Correspondence -
Dr. Vishal V. Ramteke
E-mail : vvramteke@gmail.com

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Case Report :

A 38 years old female patient reported to the gynecology outpatient department with history of heavy menstrual bleeding and abdominal pain for 6-8 months. She was evaluated and found to have multiple uterine fibroids of average size 10 cm × 4 cm with anemia. In the past she was diagnosed as a case of lupus nephritis ISN RPS class IIIa 4 years back for which she had received tab Mycophenolate mofetil with tab Prednisolone for one year followed by tab Azathioprine with tab Prednisolone for next 2 years. She was in remission and was on Tab Prednisolone 5 mg once daily and Tab Hydroxychloroquine 200 mg once daily. She had normal renal function with no proteinuria and RBCs in urine and no clinical features of lupus activity. Patient was offered option of medical management (Selective progesterone receptor modulator Uripristal Acetate) versus surgical management (Laparoscopic hysterectomy). Since she had completedfamily, she opted for total laparoscopic hysterectomy. After thorough evaluation preoperative she was subjected to the surgery. Her oral prednisolone was switched to injectable hydrocortisone till she was kept nil by mouth. Her perioperative course was uneventful and was

discharged in health on post operative day 2.

Patient reported on post-operative day 9 with complaints of lower abdominal pain, pervaginal discharge and high fever grade. There were associated complaints of severe headache and polyarthralgia. Clinical exam revealed tachycardia with severe tenderness in the umbilical and hypogastric region. Hemogram showed hemoglobin - 8.4 gm/dl, WBC - 12,800 /mm^3, platelets - 3.24 lacs/mm3.Her kidney and liver function tests were normal. Urinalysis showed 2+ albumin, plenty of pus cells, RBC 12-15/hpf and 2-3 RBC cast. On Ultrasound examination a collection of 40 ml in pelvis superior to vaginal vault posterior to bladder was noted. After sending vaginal swab and urine sample for culture sensitivity, she was started on injection Piperacillin Tazobactam with pervaginal metronidazole and clindamycin pessary. She was continued on oral prednisolone was continued at a dose of 10 mg/day. Her complement levels were low C3 - 53mg/dL (88201mg/dL) and C4-4 mg/dL (16-47 mg/dL) and urine protein creatinine ratio of 2.2. Hence diagnosis of lupus nephritis with postoperative abdominal sepsis was arrived on. In/v/o abdominal sepsis, steroids were kept at a low dose of 10 mg/day of prednisolone with tab Hydroxychloroquine 200 mg/day. Urine culture was sterile while vaginal swab pus culture showed significant growth of E. Coli (> 1 lac colony count) sensitive to inj Meropenem and hence antibiotics were changed as per sensitivity. On post-operative day 13, she had leaking PV and blood spotting with severe lower abdominal pain. Urgent contrast enhanced CT abdomen pelvis was done which showed a loculated peripherally enhancing collection of 80 cc in pelvis closely abutting distal right ureter duplex moiety with fat stranding and air pockets ***(Fig. 1).*** There was new onset mild to moderate ascites which was not present on initial imaging on admission. The radiocontrast excretion was abruptly cut off in right lower ureter in the vicinity of the phlegmon without dye extravasation with normal excretion from the left kidney ***(Fig. 2).*** A cystoscopy with right retrograde pyelography was done which was suggestive of narrowing of lower 4 cm right ureter with delayed dye excretion corresponding to location of the phlegmon. A Double J stent was inserted, and evacuation of the pelvic collection was done, and pus cultures were sent. Post procedure there was worsening of abdominal distention and pain with ileus and incessant vomiting. She was maintained on intravenous fluids and kept nil per orally with nasogastric decompression. Oral steroids were switched to inj Hydrocortisone 50 mg tds.

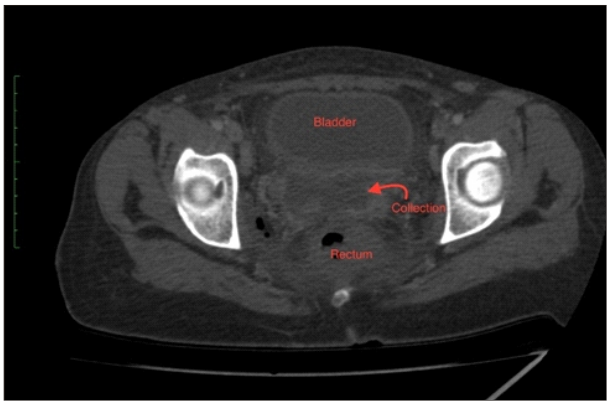

Figure 1 : **Axial view of CECT abdomen pelvis showing a loculated peripherally enhancing collection of 80 cc in pelvis**

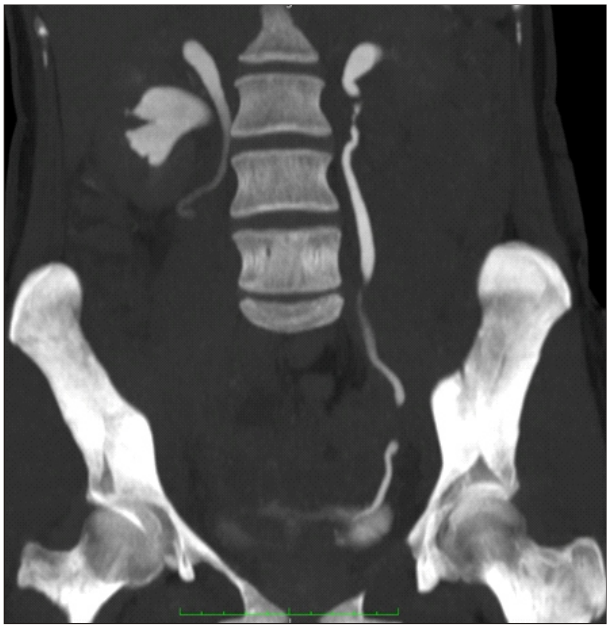

Figure 2 : **Coronal view of CECT abdomen pelvis showing non excretion of radiocontrast in right lower ureter and normal excretion from the left kidney**

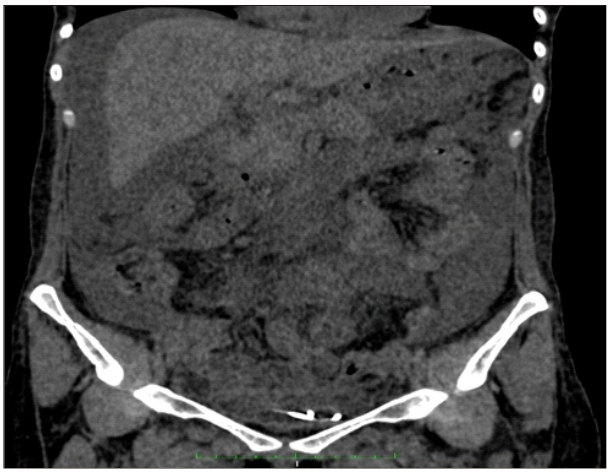

Figure 3 : **Coronal view of CECT abdomen pelvis showing moderate ascites**

In view of worsening complaints, CT Abdomen pelvis was done after 4 days which was suggestive of moderate to severe ascites with omental thickening ***(Fig. 3).*** Her serum total proteins and albumin were 6.4 mg/dl and 3.3 mg/dl respectively. Ascitic fluid analysis showed exudative fluid with 600 cells (all lymphocytes) with negative gram stain and acid-fast bacilli. SAAG (Serum Ascites Albumin Gradient) was 0.6 and ADA was 20,with normal ascitic fluid lipases and creatinine values. Ascitic fluid culture was negative. Diagnosis of lupus serositisin addition to nephritis was confirmed. Fresh urine cultures and culture from abdominal pus drained earlier did not show any growth. The procalcitonin was normal - 0.3 ng/ml (normal < 0.5 ng/ml) range and ferritin was high - 1210 ng/ml (normal 5-204 ng/mL). To control the ongoing serositis, patient was administered inj. Methyl prednisolone pulses - 500 mg / day for 3 days followed by oral prednisolone 1 mg/kg/day and tab Hydroxychloroquine 200 mg OD. With this management, there was remarkable improvement in ileus, abdominal pain, distention and vomiting. By day 5 of initiation of pulse methylprednisolone injections, she had no vomiting and was tolerating oral feeds. After stabilization, she was subjected to ultrasound guided kidney biopsy which was suggestive of Lupus Nephritis ISN RPS Class III A. Right ureteric DJ stent was removed after 4 weeks of insertion and retrograde pyelography did not show any narrowing or delayed excretion of dye from the ureter. She was started on tab Cyclophosphamide 1.5 mg/kg/day with tapering oral steroids for 4 months with which she achieved remission from lupus nephritis with no abdomi-nal complaints. Maintenance of remission was done with mycophenolate sodium alongwith steroids and hydroxychloroquine. On follow up, she is in remission from lupus nephritis and peritonitis since past 16 months.

Discussion :

Systemic lupus erythematosus is an autoimmune disorder characterized by small vessel vasculitis and inflammation of the serousmembranes like pericardium, pleura, and peritoneum[1,2]. It affects gastrointestinal tract causing mesenteric vasculitis, acute pancreatitis, acute diarrhea, inflammatory bowel diseases, protein-losingenteropathy, intestinal pseudo obstruction and peritoneal serositis[3,4]. Peritoneal serositis is a frequent finding noted in 60-70 % cases of SLE on autopsy[6]. But peritoneal serositis with ascites (known as lupus peritonitis) is in frequently seen with only few cases reported.

B lymphocytes produce autoantibodies that bind circulating antigens, forming immune complexes that deposit on the peritoneum. This leads to upregulation of expression by proinflammatory cytokines such as interleukin- 1b, interferon- , vascular endothelial growth factor (VEGF), and fibroblast growth factor causing local inflammatory response and exudate formation[4,5]. Vasculitis of peritoneal vessels contributes to inflammation further leading to insidious onsetexudative ascites[6]. Ascitic fluid examination typically has a low Serum Ascites Albumin Gradient (SAAG) < 1.1. Mixed cellularity is observed with lymphocytic predominance and sterile culturesas observed in our case. Histopathological examination of the peritoneal membrane consists of infiltrations of mononuclearor polymorphonuclear leukocytes and immune complexes around the peritoneal vessels.[5,6] Concomitant vasculitis is also noted which contribute to the pathogenesis of lupus peritonitis.

Lupus peritonitis is usually observed in diagnosed

lupus patients with other organ involvement but can be a presenting manifestation in newly diagnosed case of lupus. Low complements levels with positive antinuclear antibody (ANA), double stranded DNA (ds DNA) and high SLEDAI score as noted with lupus activity correlate with the onset of the abdominal symptoms[2]. No specific autoantibodies are associated with lupus peritonitis as with another organ involvement. High ferritin levels > 1000 ng/ml as observed in the current case has been a finding in the reported cases of lupus peritonitis. It is a diagnosis of exclusion and conditions like spontaneous bacterial peritonitis, tuberculous and fungal peritonitis should always be ruled out which can commonly occur in lupus patients on immunosuppressants.

The prognosis of lupus peritonitis is good if there is prompt diagnosis and institution of immunosuppressive therapy. If untreated high mortality rates (19%) are observed.[3,6] Pulse dose of steroids with gradual taper and hydroxychloroquine is the mainstay of treatment. Refractory cases benefit from cyclophosphamide, mycophenolate mofetil, azathioprine and rituximab[5,6]. Due to abdominal symptoms and poor oral intake there is high risk of malnutrition, hypoalbuminemia and recurrent refill of ascites which must be addressed during managing such patients. In the current case since there was associated lupus nephritis, she was treated with steroids in combination with cyclophosphamide and hydroxychloroquine to which both nephritis and peritonitis responded well.

Conclusion :

Lupus peritonitisis rare presentation of the serositis which is commonly seen in SLE patients. It should be considered a diagnosis of exclusion, after extensive clinical and laboratory evaluation for alternative causes of exudative ascites. Prognosisis usually good with available treatment for SLE like steroids and hydroxychloroquine. Cyclophosphamide, mycophenolate mofetiland rituximab are indicated in refractory cases.

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List of abbreviation : Systemic Lupus Erythematosus (SLE), Anti-Nuclear Antibody (ANA), Double stranded DNA (DsDNA), International Society of Nephrology Renal Pathology Society (ISN RPS), Serum Ascites Albumin Gradient (SAAG)

Conflict of interest : None

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