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The emergence of bladder dysfunction secondary to lupus cystitis in a patient with established lupus nephritis: the first case from Pakistan

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ABSTRACT

Lupus cystitis is an uncommon manifestation of systemic lupus erythematosus (SLE). We describe the case of a 23-year-old woman with lupus that complained of abdominal pain, vomiting, and diarrhea for one week. Two years back, she was brought with an affirmative history that satisfied the clinical criteria of SLE which was renal biopsy-proven (class V membranous lupus nephritis). On radiologic workup, she was found to have bilateral hydronephrosis and a thick-walled bladder with diverticula. Immunosuppressive therapy was commenced however augmentation cystoplasty had to be opted for as the symptoms did not subside with medical therapy alone.

Keywords: Lupus cystitis systemic lupus erythematosus lupus nephritis

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Introduction

Systemic lupus erythematosus [SLE] is a chronic, autoimmune, inflammatory disorder that affects a multitude of organs. This includes skin, joints, kidney, heart, lung, gastrointestinal tract, vascular system, and rarely the bladder. Lupus cystitis [LC] is a rare manifestation of SLE, with most of the cases reported from East Asia. LC usually presents with lower urinary tract symptoms such as suprapubic pain, frequency, dysuria, nocturia, and polyuria however, the condition often mimics enteritis with prime complaints being vomiting, diarrhea, and abdominal pain [1,2]. Imaging and urodynamic studies are integral to determine the extent of bladder wall thickening and decreased bladder capacity in LC [3]. We report an unusual case of a young South Asian female with lupus cystitis who presented with a one-week history of symptoms mimicking gastroenteritis. Reviewing the literature, we identified that recurrent bowel symptoms can be the initial presentation in the patients of LC.

Case presentation

A 23-year-old female was admitted to our hospital in February 2017 with a 1-month history of joint pain, primarily involving the wrists. Physical examination revealed a prominent facial malar rash and discoid rash involving the limbs. She complained that her skin condition deteriorated with exposure to sunlight. Her laboratory studies were remarkable for anemia [Hb= 9.0 g/dl], leucopenia [3000/μL], an antinuclear antibody titer of 1:320 dilution [homogenous speckled pattern], and an anti-ds-DNA antibody at 103 IU/mL. Her urinary tests showed proteinuria with a urinary protein/creatinine ratio [UPCR] of 400mg/nmol. This presentation led to her SLE diagnosis. Further on, a renal biopsy showed a class V membranous lupus nephritis [WHO classification]. Steroids and hydroxychloroquine were initiated, and the disease was kept under control for two years.

During her second year of disease, she had a serious episode of gastrointestinal symptoms requiring hospitalization. Her chief complaints were abdominal pain, vomiting, and diarrhea for one week. On examination, her vital signs were within the normal limits. Labs investigations showed hemoglobin 11.6g/dl, leukocyte count 6010/dl and renal function tests showed normal urea and creatinine levels. Liver function tests and serum electrolytes were within normal limits. Serum albumin was 3.25g/dl. Urine analysis showed numerous pus cells and proteinuria of 3+. Renal ultrasound was done as part of the routine examination that revealed bilateral dilatation of the pelvicalyceal systems showing bilateral mild to moderate hydronephrosis [Fig. 1]. Ultrasound bladder showed increased bladder wall thickness. CT Pyelogram revealed thick-walled urinary bladder with diverticula due to reflux along with bilateral moderate hydronephrosis with hydroureter down to urinary bladder without an obstructing calculus [Fig. 2].

Video urodynamic was done because the patient had an ongoing history of persistent frequency nocturia and urgency for 5 months. It showed a small capacity bladder, high filling pressure with detrusor overactivity, normal voiding pressure with low flow rate, impaired compliance, no vesicoureteral reflux, and no post-void residual urine. Cystoscopy was done that showed small capacity bladder approximately 250 ml with normal urethra; no growth was seen and biopsy was taken and sent for histopathology department that showed moderate acute on chronic inflammation with no evidence of tuberculosis or malignancy [Fig. 3]. Cystogram revealed multiple, irregular outlined small outpouchings in the bladder. Based on these investigations and findings we diagnosed the condition as lupus cystitis. Treatment was started with intravenous methylprednisolone and then switched to oral methylprednisolone. Later, the patient underwent augmentation cystoplasty Mitrofanoff and VQZ plasty because of the small capacity bladder and high filling pressures. The

surgery was uneventful, and the patient was discharged on oral steroids. On oral prednisolone 5mg/day and continuous intermittent self-catheterization, she has been free of urinary symptoms for 2 years.

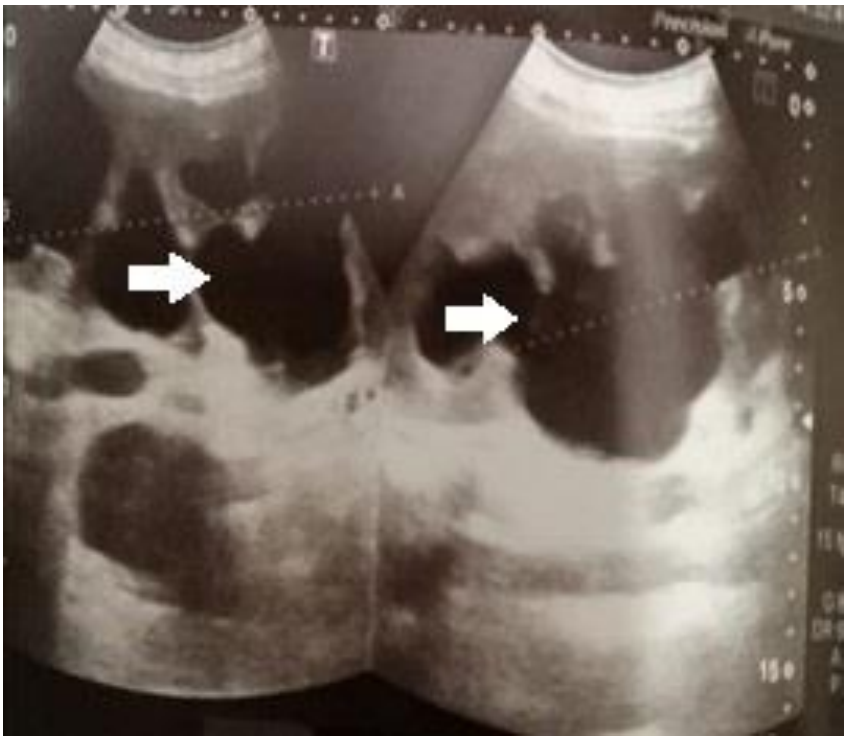

Figure 1-Ultrasound of the patient, arrows showing: bilateral dilated renal collecting system with hydronephrosis.

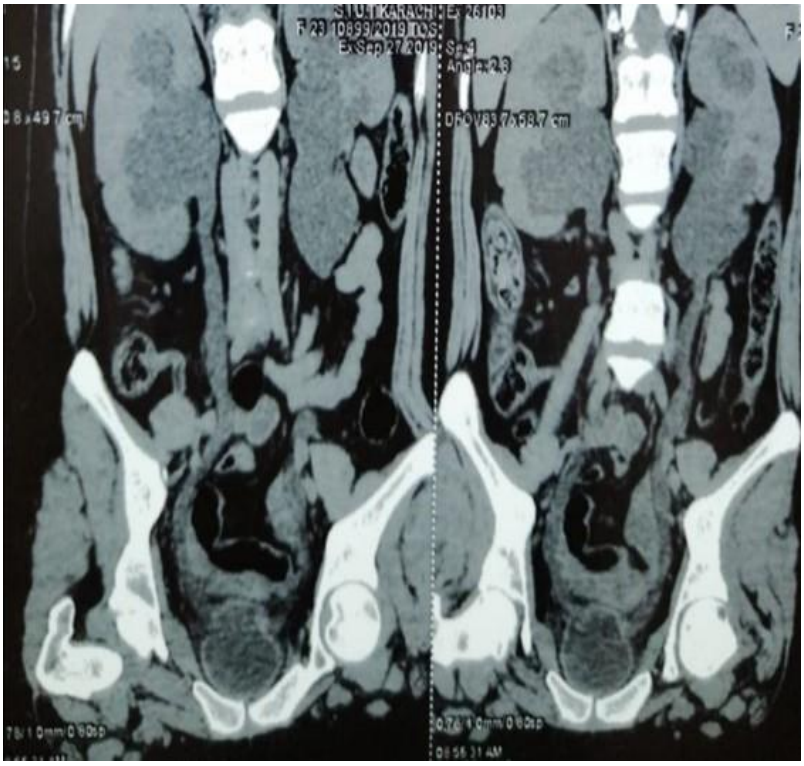

Figure 2- CT pyelogram showing thick walled urinary bladder and bilateral moderate hydronephrosis.

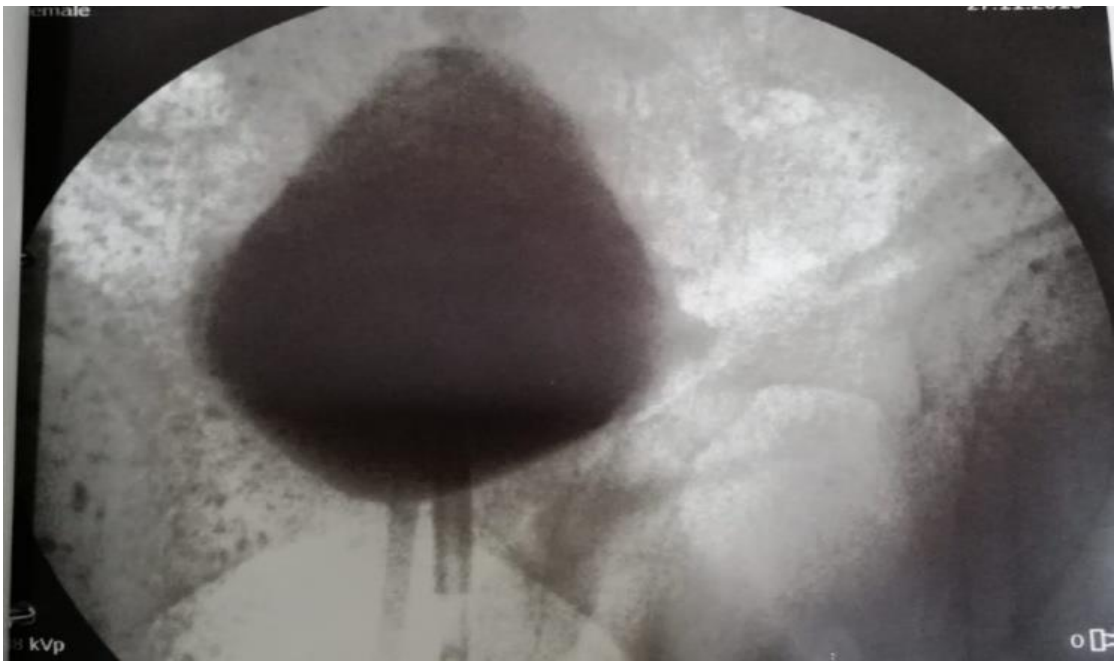

Figure 3- Cystogram showing multiple irregular outpouchings in the bladder.

Discussion

The presence of gastrointestinal complaints along with bladder irritation symptoms with a mildly deranged urinalysis should raise the suspicion of interstitial cystitis in the setting. The diagnosis however needs reinforcement via findings of hydronephrosis in ultrasonography or tomography along with reduced capacity bladder on urodynamics [4]. Our patient exhibited suprapubic pain, nocturia, frequency, and polyuria for one week, however, the superlative complaints remained persistent vomiting, diarrhea, and abdominal pain, suggesting lupus cystitis with ongoing enteritis.

Interstitial cystitis can manifest in connective tissue disorders like SLE, Sjogren's syndrome, and Rheumatoid arthritis [5]. The pathogenesis of the condition in SLE remains vague. The co-occurrence of urinary and gastrointestinal symptoms in LC has been authenticated by immunohistochemical studies which suggest IgG and C3 deposition in the vasculature of intestines and the bladder [6,7]. Only a few cases of LC have been reported and most of these are from Asian countries [8]. Our case is the first such presentation from Pakistan. Our patient was diagnosed with SLE two years before the onset of LC. One study evaluated 413 patients with SLE; 10 had bladder irritation symptoms and only 5 had established LC [9]. However quite remarkably all these patients presented with gastrointestinal complaints. This is suggestive of the fact that gastrointestinal manifestations should not be neglected in SLE patients; these can be crucial for the early diagnosis of LC.

Bilateral hydronephrosis in LC has been attributed to smooth muscle dysmotility [4]. Also, the pattern of concomitant enteritis, cystitis, and nephritis has been described in a few studies: our case is the continuation of a similar trend encountered in practice. In a study conducted by Yuji et al, 78 patients with lupus cystitis were analyzed out of which 81.7% had simultaneous or subsequent lupus enteritis and 61.3% had renal involvement [10]. Furthermore, female gender, ongoing enteritis, and positive anti-ds-DNA antibody were recognized as risk factors for LC.

LC responds well to steroid pulse therapy and immunosuppressive agents provided that the treatment is timely. In our case, intravenous methylprednisolone was commenced promptly, and the response was good. Her gastrointestinal symptoms completely subsided with the steroid

therapy however bladder symptoms persisted for which augmentation cystoplasty had to be performed. In a few studies, cetirizine hydrochloride was reported to be efficacious against LC refractory to steroids [10].

In summary, we have reported the very first case of LC with nephritis from Pakistan. LC is a rare yet treatable manifestation of SLE. The prognosis of the condition is dependent on quick diagnosis and prompt treatment with corticosteroids and cyclophosphamide. In case of delay, adverse outcomes of irreversible bladder dysfunction and renal failure can complicate the condition. We hope that this case report will draw the attention of the physicians towards the management of LC and will enlighten them with the diverse presentation of the condition.

Abbreviations: SLE: Systemic lupus erythematosus, LC: Lupus cystitis, CT: computed tomography

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References

[1]. Koh JH, Lee J, Jung SM, Ju JH, Park SH, Kim HY, Kwok SK. Lupus cystitis in Korean patients with systemic lupus erythematosus: risk factors and clinical outcomes. Lupus. 2015 Oct;24[12]:1300-7.

[2]. Yuan S, Ye Y, Chen D, Qiu Q, Zhan Z, Lian F, Li H, Liang L, Xu H, Yang X. Lupus mesenteric vasculitis: clinical features and associated factors for the recurrence and prognosis of disease. InSeminars in arthritis and rheumatism 2014 Jun 1 [Vol. 43, No. 6, pp. 759-766]. WB Saunders.

[3]. Tsushima Y, Uozumi Y, Yano S. Reversible thickening of the bowel and urinary bladder wall in systemic lupus erythematosus: a case report. Radiation medicine. 1996 Mar 1;14[2]:95-7.

[4]. Kinoshita K, Kishimoto K, Shimazu H, Nozaki Y, Sugiyama M, Ikoma S, Funauchi M. Two cases of lupus cystitis with no bladder irritation symptoms. Internal Medicine. 2008;47[16]:1477-9.

[5]. KOIKE T, TAKABAYASHI K. Lupus cystitis in the Japanese. Internal medicine. 1996;35[2]:87-8.

[6]. Weisman MH, McDanald EC, Wilson CB. Studies of the pathogenesis of interstitial cystitis, obstructive uropathy, and intestinal malabsorption in a patient with systemic lupus erythematosus. The American journal of medicine. 1981 Apr 1;70[4]:875-81.

[7]. Nakauchi Y, Suehiro T, Tahara K, Kumon Y, Yasuoka N, Ohashi Y, Kawada M, Hashimoto K. Systemic lupus erythematosus relapse with lupus cystitis. Clinical and experimental rheumatology. 1995 Sep 1;13[5]:645-8.

[8]. Boye E, Morse M, Huttner I, Erlanger BF, MacKinnon KJ, Klassen J. Immune complex-mediated interstitial cystitis as a major manifestation of systemic lupus erythematosus. Clinical immunology and immunopathology. 1979 May 1;13[1]:67-76.

[9]. Marques MR, Matos C, Oliveira S. Enteritis and cystitis-a cause of abdominal pain in lupus. Acta reumatologica portuguesa. 2009 Apr 1;34[2A]:241-5.

[10]. Nishizaki Y, Tamaki H, Yukawa S, Matsui Y, Okada M. Comparison between Japanese and non-Japanese features of lupus cystitis based on case reports including novel therapy and a literature review. Internal Medicine. 2011;50[9]:961-8.