

Hydronephrosis and painless ascites: rare features of late-onset SLE

Sirs,

We present here a rare case which illustrates the difficulty in diagnosing and treating late onset SLE hydronephrosis and ascites. More research must be done to exactly understand the pathophysiology, the most effective treatment, and the long term prognosis of these unusual SLE manifestations

A 64-year-old white Hispanic female with systemic lupus erythematosus (SLE), hypertension and hypothyroidism who presented to our Emergency Room (ER) complaining of abdominal swelling. The patient's SLE had presented 4 years prior with malar rash, generalized arthritis, alopecia, serositis, and a positive fluorescent antinuclear antibody test (ANA). Following initial presentation, she had remained largely asymptomatic until 3 months prior to the ER visit, when there was increasing abdominal girth with pain, anorexia, nausea, emesis, 30 lb. weight loss, constipation and urinary retention. The patient denied fevers, chills, sweats, rashes, oral ulcers, arthritis, morning stiffness, dry eyes or mouth, hematuria or dysuria, shortness of breath, and pleuritic chest pain.

On physical examination, significant findings were generalized alopecia, bibasilar crackles, and massive ascites. Additionally, the computerized axial tomography scan image in Figure 1 displays the patient's hydronephrosis without stones, clots, or masses. After etiologies of hydronephrosis and ascites other than SLE were excluded, treatment was initiated with intravenous corticosteroids. Following symptomatic improvement, the patient was discharged on oral prednisolone at 1 mg per kg with outpatient follow up.

Hydronephrosis is an uncommon presenting feature of SLE; however, it tends to be associated with emesis, paralytic ileus, malabsorption, ascites, intestinal pseudo-obstruction, diarrhea, ITP renal failure, glomerulonephritis, and cystitis (1,2). The pathogenesis appears to be an immune complex mediated vasculitis causing inflammation and smooth muscle dysmotility (1,3). This can result in reduced bladder capacity, detrusor muscle spasm, vesiculoureteric reflux, bladder wall thickening and fibrosis with ensuing symptoms such as frequency, urgency, dysuria, nocturia, and incontinence (1,4). Late onset SLE patients, unlike younger patients, are thought to have a poor prognosis despite aggressive steroid and steroid sparing immunosuppression. 5 In previous case reports, high dose oral corticosteroids were required to treat SLE

Fig. 1. Contrast in ureters.



induced hydronephrosis (3, 5-7). If there is significant gastrointestinal vasculitis, however, intravenous corticosteroids and, possibly, nephrostomy stents have been necessary (2). Subjective improvement has been reported and correlated with radiological improvement in bladder volume and ureter and collecting system anatomy (2). Yet, more studies are needed to assess long term bladder, ureter, and kidney function (7).

While chronic peritonitis is found in over 63% of SLE patients on autopsy, frank ascites is only prevalent in approximately 8-11% of patients (6, 8). Cancer markers, such as CA 125, can also be elevated (in this case, level was 282 U/mL). While neoplasm is possible, chronic inflammation of the mesothelial cell lining may actually elevate levels of CA 125 which will return back to baseline upon treatment with immunosuppressive medication (9).

Ascites may be due to antibody-complement mediated immune complexes, antiphospholipid antibodies, or vasculitis-associated impaired circulation (8). Typical ascites laboratory findings include mostly lymphocytic white blood cells with a mean of 393/mm³, total protein mean of 3.9 g/dL, and ANA, anti-DNA antibodies, and LE cells; the complement levels tend to be low.

Intravenous corticosteroids, possibly in combination with cyclophosphamide, azathioprine, or intra-abdominal steroids, are also necessary for the treatment of ascites because of the chronic inflammation, bowel wall edema, vasculitis, and impaired vascular circulation (6, 8, 10). Unfortunately, marked ascites in late onset SLE patients tends to have a poor prognosis not only because of the above, but also secondary to diagnostic delays and complications like septic shock. Remission or suppression of ascites and chronic peritonitis is unlikely (10).

Hydronephrosis and painless ascites occur in a small subset of patients with SLE and prognosis in older patients is thought to be poor. The current treatment approach is the

use of high dose corticosteroids possibly in combination with other immunosuppressive agents. The optimal long term management regimen is not known. Physician awareness and early diagnosis may positively affect outcomes by allowing early intervention.

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