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Medi Quest BRS Hospital

A monthly News letter from BRS Hospital

TWO INTERESTING CASE REPORTS OF SLE

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Case Report - 1

Gastrointestinal symptoms are common in SLE but are usually attributable to infection, medication side effects or other underlying conditions. In rare instances, they are caused by the auto-immune process itself.

Here we report a case of SLE which presented with loose stools , vomiting, pain abdomen and decreased urine output , eventually diagnosed to be having lupus enteritis and lupus nephritis.

INTRODUCTION

SLE is a chronic autoimmune disease which has a fluctuating clinical course, characterised by intermittent flares involving virtually any organ or organ system of the body. In a female of child bearing age presenting with fever, rash, joint pains an entity of SLE should be always considered . Nevertheless, GI involvement due to SLE activity is uncommon, which is difficult to diagnose and is also life threatening, if diagnosis is delayed it can lead to ischemia, infarction and also perforation requiring emergent surgical intervention.

The term lupus enteritis refers to inflammation of the bowel wall due to SLE activity. Pathogenesis may include immune complex deposition in the bowel wall or vessel vasculitis. It is reported in 0.2 to 5.8 % of SLE patients, with a high mortality rates of ~ 50%, if complicated or if treatment is delayed.The classical manifestation of this disorder is severe acute abdominal pain. Abdominal pain in SLE can be secondary to peritonitis, pancreatitis, acalculous cholecystitis, gastroenteritis, mesenteric ischemia, intestinal pseudoobstruction , and peptic ulcer disease , thus making diagnosis of this rare debilitating condition even more difficult. Therefore deeper understanding of this disease entity is necessary.

CASE REPORT

24 year old female, with the background of SLE, under treatment for the same since 10 years of age in BRS Hospital, was admitted to our hospital through emergency department with severe abdominal pain, vomiting, loose stools of 10 days duration. Her symptoms worsened three days before admission along with decreased urine output. On clinical examination she was mildly febrile, dehydrated, BP was 130/90mm of Hg, pulse was 114/min. Her abdominal examination was significant with diffuse tenderness, distension and absent bowel sounds. Rest of the systemic examination was normal.

Post admission work up showed elevated creatinine,CRP. Urine albumin was 3+ with nephrotic range of proteinuria of 5.84gm/day. Motion routine was normal.

She was kept nil by oral in view of abdominal distension and absent bowel sounds. She was suspected to have infectious diarrhoea with AKI and also a possibility of lupus enetritis and lupus nephritis flare. She was commenced on IV fluids, IV ceftriaxone and IV metronidazole, IV pantoprazole infusion and IV ondansetron as needed.

Ultrasound abdomen was performed which showed fluid distended stomach,bilateral medical renal disease. CT Abdomen and Renal biopsy were not done as patient/ attenders were unwilling. C3, C4 levels were done which were low. Her SLEDAI score is about 14 indicating high disease activity. Note : The GI symptoms aren't included in SLEDAI (Systemic Lupus Erythematosus Disease Activity Index - ref Table 3).

In view of diagnosis of lupus nephritis flare and high clinical suspicion of lupus enteritis patient was commenced on pulse methylprednisolone 500mg IV OD after concurrence with nephrologist, following which her GI symptoms dramatically improved and oral intake was commenced which she tolerated well. Her urine albumin became trace and urine sediment also decreased.

With the advice of Rheumatologist she was discharged on Tab prednisolone, hydroxychloroquine and mycophenolate sodium.

DISCUSSION

Lupus enteritis is defined either as vasculitis / inflammation of small bowel wall that is supported with imaging or biopsy findings. Mechanism is believed to be due to immune complex deposition and thrombosis of the intestinal vessels which presents with very non-specific signs and symptoms such as abdominal pain(97%), ascites(78%), nausea (49%), vomiting(42%), diarrhoea (32%),fever 20(%).

Most common lab findings of lupus enteritis are haematological derangements(leukopenia, lymphopenia,anemia) ; positive ANA, Anti dsDNA , low complement. No specific auto antibody related to lupus enteritis has been identified.

Diagnosis is usually a combination of high clinical suspicion, haematological and imaging findings. Abdominal CT is the first line modality with three classical findings of bowel wall edema and enhancement (target sign), engorgement / increased number of mesenteric vessels (comb sign), increased attenuation of mesenteric fat. Ascites is also a common finding.However these are non specific and are also seen with other conditions like intestinal obstruction, pancreatitis, IBD.

USG Abdomen usually shows bowel wall thickening, dilation of intestinal segments and mild ascites.

Endoscopy may help to exclude the etiologies but doesn't contribute to the diagnosis.

First line treatment includes systemic corticosteroids; Methyl Prednisolone 250mg to 1gm IV followed by oral prednisolone 0.5 to 1 mg/kg/day; Bowel rest, Hydration, electrolyte replacement.

Cyclophosphamide and azathioprine, are added in cases and continued as maintenance therapy along with hydroxychloroquine and prednisolone. Refractory cases can be treated with rituximab.

Our patient also had similar clinical manifestations which were not responding to antimicrobial medications and other symptomatic treatment, and with laboratory evidence of lupus nephritis, a clinical diagnosis of lupus enteritis and lupus nephritis was made and she improved with pulse methylprednisolone therapy. Currently she is in remission with prednisolone, hydroxychloroquine, MMF.

CONCLUSION

In a young female with SLE and intractable abdominal pain, the possibility of lupus enteritis should be kept in mind. Early diagnosis and management with high index of clinical suspicion along with steroid pulse therapy may help avoid mortality due to life threatening complications such as bowel ischemia and perforation.

Case Report - 2

Although lupus erythematosus and scleroderma are regarded as two distinct entities, there have been multiple cases described in the literature showing an overlap between these two disease processes. Here we report a case of 25year old female with a diagnosis of SLE/ scleroderma overlap.

INTRODUCTION

Overlap syndromes are inflammatory rheumatic conditions in which patients have clinical manifestations suggestive of multiple distinct immune diseases. The diseases most commonly involved in overlap syndromes include SLE, Rheumatoid Arthritis, Systemic Sclerosis, Polydermatomyositis, Sjögrens syndrome. Any combination of co-existing rheumatic diseases are reported. We can approach these conditions either by detecting a specific antibody combined with particular clinical findings or by identifying a certain pattern of clinical features with out a specific serological marker.

The most well-characterised overlap syndrome, MCTD (Mixed connective tissue disease), was originally defined in 1972 as a connective tissue disorder characterized by the presence of high titers of a distinctive autoantibody, now called anti-U1 ribonucleoprotein (anti RNP antibodies) (previously termed antibody to extractable nuclear antigens [anti-ENA]) and signs and symptoms of a combination of disorders —primarily lupus erythematosus , scleroderma, and polymyositis. Many people with this uncommon disease also have Sjogren's syndrome..

Overlap syndromes are generally less common than the conditions they encompass.The accurate identification of overlap syndrome is

useful in order to clarify the prognosis and apply the proper therapeutic measures.

CASE REPORT

A 25 year old female came with a history of appearing and disappearing rash, photosensitivity, myalgias, arthralgias, fever, puffiness of fingers and toes, asthenia and weight loss. There was alopecia, loss of interphalangeal skin creases, hyper-pigmented thickened scaly lesions over lower extremities, flexion contractures of fingers . Her blood investigations showed raised ESR, positive ANA screening, raised anti dsDNA, and anti centromere antibodies. With the above clinical and laboratory evidence using ACR/EULAR criteria a working diagnosis of SLE/ scleroderma overlap was made which was confirmed by the consultant rheumatologist and she was commenced on steroid therapy, HCQ, mycophenolate and other supportive measures, discharged and advised periodic review.

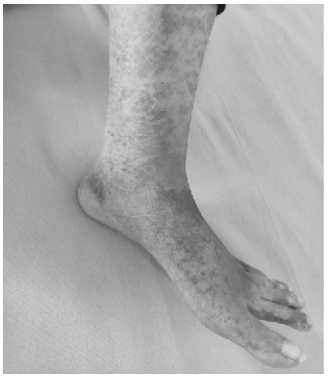

Fig.1 : Image of Post-inflammatory hyperpigmentation in shin and feet

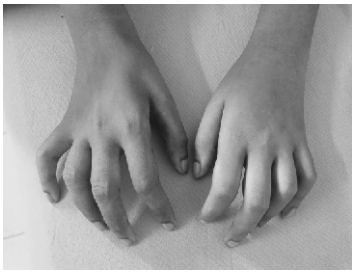

Fig.2 : Image of hands exhibiting sclerodactyly

DISCUSSION

Overlapping features of lupus can be associated with both limited and diffuse cutaneous subtypes of systemic sclerosis, however the former subgroup is less commonly observed. Raynaud's phenomenon, calcinosis cutis, telangiectasis, digital tip ulcers and diffuse skin involvement are some of the manifestations. Scleroderma renal crisis, interstitial lung disease, pulmonary arterial hypertension, polyserositis are the major systemic complications. The real challenge in this overlap is the patient presenting with hypertension and renal dysfunction with differential diagnosis being lupus nephritis and scleroderma crisis. The treatment is completely different for both and treatment of scleroderma renal crisis with high dose steroids which is used for the management of lupus nephritis may lead to detrimental outcome.

CONCLUSION

Overlap syndrome is a disease entity to consider in patients with multiple symptoms that cannot be classified into one connective tissues disease. Treatment of this disease should be individualised and based on the connective tissue diseases involved.

Table 1 : ACR/EULAR Criteria for Systemic Sclerosis

Items	Sub-items	Weight
Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints		9
Skin thickening of the fingers *(only count the highest score)*	Puffy fingers Whole Finger, distal to MCP	2 4
Finger tip lesions *(only count the highest score)*	Digital Tip Ulcers Pitting Scars	2 3
Telangiectasia		2
Abnormal nailfold capillaries		2
Pulmonary arterial hypertension and/or Interstitial lung Disease		2
Raynaud's phenomenon		3
Scleroderma related antibodies (any of anti-centromere, anti-topoisomerasel [anti-ScL 70], anti-RNA polymerase III)		3
	TOTAL SCORE:	
Patients having a total score of 9 or more are being classified as having definite systemic sclerosis.		

Table.2 : ACR / EULAR Criteria for SLE

Entry criterion
Antinuclear antibodies (ANA) at a titer of ≥1:80 on HEp-2 cells or an equivalent positive test (ever)

↓

If absent, do not classify as SLE
If present, apply additive criteria

↓

Additive criteria
Do not count a criterion if there is a more likely explanation than SLE.
Occurrence of a criterion on at least one occasion is sufficient.
SLE classification requires at least one clinical criterion and ≥10 points.
Criteria need not occur simultaneously.
Within each domain, only the highest weighted criterion is counted toward the total score§.

Clinical domains and criteria	Weight	Immunology domains and criteria	Weight
Constitutional		***Antiphospholipid antibodies***	
Fever	2	Anti-cardiolipin antibodies OR	
Hematologic		Anti-β2GP1 antibodies OR	
Leukopenia	3	Lupus anticoagulant	2
Thrombocytopenia	4	***Complement proteins***	
Autoimmune hemolysis	4	Low C3 OR low C4	3
Neuropsychiatric		Low C3 AND low C4	4
Delirium	2	***SLE-specific antibodies***	
Psychosis	3	Anti-dsDNA antibody* OR	
Seizure	5	Anti-Smith antibody	6
Mucocutaneous			
Non-scarring alopecia	2		
Oral ulcers	2		
Subacute cutaneous OR discoid lupus	4		
Acute cutaneous lupus	6		
Serosal			
Pleural or pericardial effusion	5		
Acute pericarditis	6		
Musculoskeletal			
Joint involvement	6		
Renal			
Proteinuria >0.5g/24h	4		
Renal biopsy Class II or V lupus nephritis	8		
Renal biopsy Class III or IV lupus nephritis	10		

Total score:

↓

Classify as Systemic Lupus Erythematosus with a score of 10 or more if entry criterion fulfilled.

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Table 3 : SYSTEMIC LUPUS ERYTHEMATOSUS DISEASE ACTIVITY INDEX

SELENA MODIFICATION

Physicians Global Assessment

0 None | 1 Mild | 2 Med | 3 Severe

SLEDAI SCORE

Check box: If descriptor is present at the time of visit or in the proceeding 10 days

Wt	Present	Descriptor	Definition
8	☐	Seizure	Recent onset. Exclude metabolic, infectious or drug cause
8	☐	Psychosis	Altered ability to function in normal activity due to severe disturbance in the perception of reality. Include hallucinations, incoherence, marked loose associations, impoverished thought content, marked illogical thinking, bizarre, disorganized, or catatonic behavior. Excluded uremia and drug causes.
8	☐	Organic Brain Syndrome	Altered mental function with impaired orientation, memory or other intelligent function, with rapid onset fluctuating clinical features. Include clouding of consciousness with reduced capacity to focus, and inability to sustain attention to environment, plus at least two of the following: perceptual disturbance, incoherent speech, insomnia or daytime drowsiness, or increased or decreased psychomotor activity. Exclude metabolic, infectious or drug causes.
8	☐	Visual Disturbance	Retinal changes of SLE. Include cytoid bodies, retinal hemorrhages, serious exodate or hemorrhages in the choroids, or optic neuritis. Exclude hypertension, infection, or drug causes.
8	☐	Cranial Nerve Disorder	New onset of sensory or motor neuropathy involving cranial nerves.
8	☐	Lupus Headache	Severe persistent headache: may be migrainous, but must be non-responsive to narcotic analgesia.
8	☐	CVA	New onset of cerebrovascular accident(s). Exclude arteriosclerosis
8	☐	Vasculitis	Ulceration, gangrene, tender finger nodules, periungual, infarction, splinter hemorrhages, or biopsy or angiogram proof of vasculitis
4	☐	Arthritis	More than 2 joints with pain and signs of inflammation (i.e. tenderness, swelling, or effusion).
4	☐	Myositis	Proximal muscle aching/weakness, associated with elevated creatine phosphokinase/adolase or electromyogram changes or a biopsy showing myositis.
4	☐	Urinary Casts	Heme-granular or red blood cell casts
4	☐	Hematuria	>5 red blood cells/high power field. Exclude stone, infection or other cause.
4	☐	Proteinuria	>0.5 gm/24 hours. New onset or recent increase of more than 0.5 gm/24 hours.
4	☐	Pyuria	>5 white blood cells/high power field. Exclude infection.
2	☐	New Rash	New onset or recurrence of inflammatory type rash.
2	☐	Alopecia	New onset or recurrence of abnormal, patchy or diffuse loss of hair.
2	☐	Mucosal Ulcers	New onset or recurrence of oral or nasal ulcerations

Wt	Present	Descriptor	Definition
2	☐	Pleurisy	Pleuritic chest pain with pleural rub or effusion, or pleural thickening.
2	☐	Pericarditis	Pericardial pain with at least 1 of the following: rub, effusion, or electrocardiogram confirmation.
2	☐	Low Complement	Decrease in CH50, C3, or C4 below the lower limit of normal for testing laboratory.
2	☐	Increased DNA binding	>25% binding by Farr assay or above normal range for testing laboratory .
1	☐	Fever	>38° C. Exclude infectious cause
1	☐	Thrombocytopenia	<100,000 platelets/mm3
1	☐	Leukopenia	<3,000 White blood cell/mm3. Exclude drug causes.

_____ TOTAL SCORE (Sum of weights next to descriptors marked present)

Mild or Moderate Flare ☐	Severe Flare ☐
☐ Change in SLEDAI > 3 points	☐ Change in SLEDAI > 12
☐ New/worse discoid, photosensitive, profundus, cutaneous vasculitis, bullous lupus Nasopharyngeal ulcers Pleuritis Pericarditis Arthritis Fever (SLE)	☐ New/worse CNS-SLE Vasculius Nephritis Myositis Pk < 60.000 Home anemia: Hb <7% or decrease in Hb > 3% **Requiring:** double prednisone Prednisone>0.5 mg/kg/day hospitalization
☐ Increase in Prednisone, but not to >0.5 mg/kg/day	☐ Prednisone >0.5 mg/kg/day
☐ Added NSAID or Plaquenil	☐ New Cytoxan, Azathioprine, Methotrexate, Hospitalization (SLE)
☐ 1.0 Increase in PGA, but not to more than 2.5	☐ Increase in PGA to > 2.5

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