Case Report:

A rare case of Focal Segmental glomerulosclerosis in Mixed connective tissue disease

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Abstract:

In this case report, a 25 year old female resented to OPD with complaints of progressive pain over bilateral knee joint since 1 year which was dull aching type, intermittent in nature, aggravates on prolonged walking and relieves on medication and rest. Patient also had pain in proximal interphalyngeal joints and metatarsophalyngeal joint. No significant medical history in the past. On examination, she was conscious, oriented, moderately built and nourished with PR- 88bPm regular, BP-100/80 mmHg. No pallor or lymphadenopathy. Bilateral Metacarpophalynheal joints were hyperpigmented and nodules were present. On knee joint examination, swelling was present over bilateral knee joints, para patellar fullness was present and tenderness was present over medial joint line. Terminal flexion was painful. From this case report , we may conclude that renal involvement can be present in patients with MCTD and it can manifest in the form of subnephrotic range proteinuria and normal creatinine values. Though membranous nephropathy is the most common lesion seen in MCTD, FSGS can also be seen.

Keywords: lymphadenopathy , glomerulonephritis

Introduction:

Mixed connective tissue disease(MCTD) was first described by Gordon Sharp in 1972 as a distinct connective tissue disorder characterized by overlapping features of systemic lupus erythemetosis, progressive systemic sclerosis and polymyositis [(1)].Presence of high titres of distinctive autoantibody called anti-U1 Ribonucleoprotein is diagnostic of mixed connective tissue disease. Mixed connective tissue disease is more common in females than males. Usually presents in second or third decade of life. Common symptoms of MCTD are low grade fever, joint pain and joint swelling, puffy fingers, myalgia, easy fatigability, Raynaud phenomenon. Major organs involved in MCTD are lungs, heart, gastrointestinal tract, kidneys and central nervous system. There are 2 best criteria useful in diagnosing Mixed connective tissue disease. They are Alarcon-Segovia criteria and Kahn criteria[(2)]. Renal involvement is usually mild and uncommon in MCTD. Previous studies have suggested that membranous nephropathy is the most common finding among patients with MCTD. In this case report, we are describing a case of biopsy proven Focal segmental glomerulosclerosis in a patient with MCTD

Case report:

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were hyperpigmented and nodules were present. On knee joint examination, swelling was present over bilateral knee joints, para patellar fullness was present and tenderness was present over medial joint line. Terminal flexion was painful. She was investigated for above complaints.

Table :1 List of investigations performed

Haemoglobin	12.0g/dl
Total count	7600cells/mm^3
Differential count	Neutophils-66% Lymphocytes-31% Eosinophils-3%
ESR	50mm 1^{st} Hr
Platelet count	294000cells/mm^3
Total protein	10.7g/dl
Albumin	4.3g/dl
Globulin	6.40g/dl
SGOT	21U/I
SGPT	8U/I
Blood urea	19.5mg/dL
Creatinine	0.98mg/dL
Uric acid	5.7mg/Dl
TSH	389UIu/mL
T3	0.862ng/mL
T4	3.41ug/dL
Sodium	134mmol/L
Potassium	4.20mmol/L
Calcium	9mg/dl
Urine Protein Creatinine Ratio	1.63:1
Urine Micro Albumin	1211.02mg Albumin/gCreatinine
ASO Titer	53.81IU/ml
R.A. Test	75.30IU/ml
CRP	0.74mg/l
DRVV screen (Test)	48.1sec
DRVV screen control	42.0sec
DRVV Screen ratio	1.14
Lupus Anticoagulant	Absent
Anti CCP	2.03
SM/RNP antibody	Positive
Anti Ro52 antibody	Positive
ACLA	IgM Positive
B2GP1	Positive
HIV,HBSAg,HCV spot	Negative

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Ultrasonography showed normal sized kidneys with no parenchymal changes. Urine routine showed 1+ proteinuria without microscopic haematuria. She underwent renal biopsy. Histopathology revealed presence of segmental sclerosis in two glomeruli with obliteration of capillary membrane There is no interstitial sclerosis /tubular atrophy of the visualized cortex. No interstitial inflammation, granuloma or eosinophils are noted. The arteries and arterioles did not reveal any diagnostic pathology. No fibrinoid necrosis or vasculitis noted. Overall features were of focal segmental glomeruloslerosis. (Figure 1) Patient was started on corticosteroids, ACE inhibitors and was advised regular follow up.

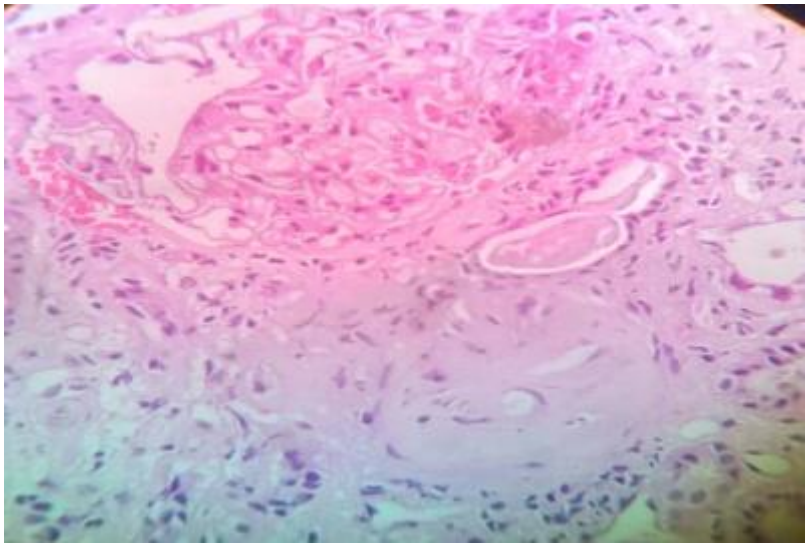

Figure:1 –Focal segmental glomerulosclerosis

The definitive diagnosis of MCTD is often complicated by the fact that the overlapping features tend to occur sequentially. Yoshida et al[(3)]did histopathological study on 17 MCTD patients out of which 4 had membranous nephropathy,2 had mesangial proliferative glomerulonephritis, and 11 had minor glomerular abnormality. Kitridou RC et al[(4)] did a follow up study among 30 patients who had MCTD.10 patients among them developed immune complex nephropathy (6 membranous nephropathy, 2 mesangial, 1 mixed, and 1 sclerosing. Sawai T et al [(5)] studied autopsy specimens of 25 MCTD patients. Membranous type of glomerular lesion was most frequently found followed by membranoproliferative and mesangioproliferative type. Kobayashi et al [(6)] studied renal biopsy samples of 5patients with MCTD out of which 2 patients had membranous nephritis and 3 patients had mesangialproliferative glomerulonephritis. Hence the case presented by us which had focal segmental glomerulosclerosis is a very rare finding possibly the first case reported in India till date. Prognosis of the patients with MCTD varies from benign course to severe progressive disease. Persistent morbidity is due to recurrent musculoskeletal pain, fatiguability, dyspnea on exertion[(7)] Major causes of death include Pulmonary arterial hypertension, ILD, myocarditis, renovascular hypertension and cerebral haemorrhage. MCTD is believed to be incurable. Treatment is individualized based on the major organs involved. Most of the patients respond to corticosteroids.

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Conclusion:

Renal involvement can be present in patients with MCTD and it can manifest in the form of subnephrotic range proteinuria and normal creatinine values. Though membranous nephropathy is the most common lesion seen in MCTD, FSGS can also be seen.

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