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A Case of FSGS in Sickle Cell Disease

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Abstract: ***A 20-year-old male with known case of sickle cell Disease presented with complain of Breathlessness, pedal oedema and reduced urine output. On urine investigation showing nephrotic range proteinuria and Renal biopsy suggestive of Focal segmental glomerulosclerosis. Patient treated with Steroids, antibiotics and iv fluids.***

Keywords: Sickle cell disease, Nephrotic range proteinuria, Renalbiopsy, Focal segmental glomerulosclerosis, HPLC report, Steroids

1. Introduction

FSGS is most common type of nephrotic syndrome occurring in middle age group presenting with complain of facial puffiness, weight gain associated with reduced urine output. In FSGS at some focal spaces part of glomeruli are fibrosed resulting in alteration in glomerular filtration with protein loss in urine. It is primary genetically acquired or secondary to some other disorder. Sickle cell Disease is one of the causes of secondary FSGS which is caused by point mutation in B globin chain of haemoglobin due to substitution between Glutamine & valine leading to disordered Hb synthesis leading to Sickle shaped cells in Blood smear. This sickle cells involve multiple systems leading to wide range of manifestations. Sickle induced renal involvement is not uncommon.30% cases of Sickles nephropathy are of FSGS variety reason being ischemic and free radical induced microvascular injury leading to sclerosis of glomeruli at variety of places ultimately leading to renal failure. Routine blood investigations, urine analysis & renal biopsy aids in diagnosis. The disease should be treated aggressively with Steroids, antibiotics & Iv fluids. Rarely renal replacement therapy is needed.

2. Case Study

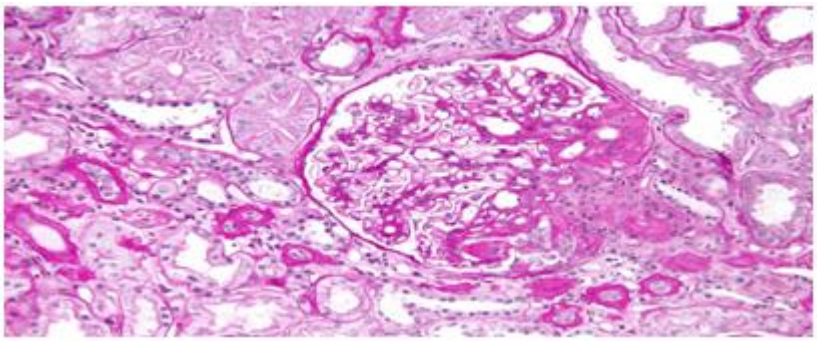
A 20/M known case of sickle cell Disease for 5 years on tab folic acid 5 mg 1 OD. Presented with complain of breathlessness NYHA grade IV for 3 days that is associated with facial puffines, reduced urine output, B/L symmetrical pitting type of pedal oedema & continuous backache. The patient had no complain of chest pain, cough, fever, nausea and vomiting, altered sensorium.

On general examination patient had pallor, mild icterus, facial puffiness, pedal edema. On admission vitals were, Blood pressure of 136/86 MMHG, Heart rate of 112 /min, Spo2 98% on 5 lit nasal O2, Respiratory rate of 24/min. On systemic examination, Respiratory examination showed bilateral infrascapular fine crepitations &other system examination were normal.

3. Investigation

UACR	30.56
URINE PROTEIN	3 +
UREA	112 mg /dl
CREATININE	4.61 mg/dl
HEMOGLOBINE	7 g /dl
C3	46
C4	32
PARVO B19	negative
USG KUB	Bilateral CMD lost
RIGHT KIDNEY	135*67 cm
LEFT KIDNEY	120 *57 cm

Renal Biopsy



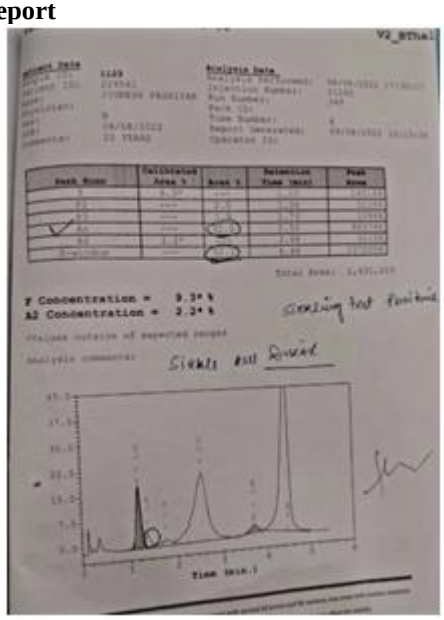
Biopsy impression:

1) Glomeruliappear enlarged with evidence of focal and segmental glomerular sclerosis involving one glomerulus. focal congestion of capillaries by sickled RBC's are noted.
2) DIF study do not show significant glomerular/extra glomerular immune deposit.

3) Acute tubular injury and focal chronic interstitial inflammation admixed with eosinophils and histiocytes are noted.
4) Focal deposition of golden-brown pigment granules in few tubular epithelial cell cytoplasm which are positive for iron/hemosiderin stain.

	07-Aug	15-Aug	22-Aug	27-Aug	04-Sep
Weight	60.5Kg	58 Kg	55 Kg	52 Kg	50 Kg
Pedal Edema	3+	2+	1+		Nil
Facial Puffiness	Present				Absent
Urea	112	80	63	38	100
Creatinine	4.61	3.23	2.58	1.53	0.59
UPCR	30.56				

HPLC Report



Management

Considering the diagnosis of FSGS patient was treated with Tablet Precin 1 mg/kg for 6 weeks followed by tapering dose given and Higher antibiotics, analgesic, and other supportive management given. On discharge tabhydroxyurea (15 mg/kg /day), tab sodamint given and Tab Folic acid added.

Outcome

Patient has clinically improved with decrease facial puffiness, pedal edema and adequate uop, breathing difficulty has been relieved. FSGS type of nephrotic syndrome has relatively good prognosis comparing nephritic syndromes. Steroid responsive FSGS has good prognosis over steroid resistant FSGS.

4. Discussion

We report a 20-year-old male patient known case of sickle cell disease who presented with complain of facial puffiness, pedaloedema, breathleness and decrease uop. Diagnosis of FSGS was confirmed by renal biopsy. One study suggestive of older sickle cell disease patient with acute parvo B 19 infection may be more susceptible to development of nephrotic syndrome. Sickle cell Disease is one of the causes of secondary FSGS which is caused by point mutation in B globin chain of haemoglobin due to substitution between Glutamine & valine leading to disordered Hb synthesis leading to Sickle shaped cells in Blood smear. This sickle cells involve multiple systems leading to wide range of manifestations. In FSGS at some focal spaces part of glomeruli are fibrosed resulting in alteration in glomerular filtration with protein loss in urine. It is primary genetically acquired or secondary to some other disorder. The disease should be treated aggressively with Steroids, antibiotics & Iv fluids.

5. Conclusion

The presenting complains of bilateral pitting pedal oedema with breathlessness and decreased urine output in a sickle cell disease patient point towards renal system involvement which on further evaluation confirmed FSGS. It is aggressively treated with immunosuppressant and rarely renal transplant needed.

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