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Secondary polycythaemia in nephrotic syndrome – an uncommon Association

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Introduction: Polycythaemia is defined as haemoglobin (Hb) of over16.5 g/dL in men and 16 g/dL in women or haematocrit of over 49% in men and 48% in women (WHO 2017). Primary polycythaemia is caused by erythropoietin (EPO) receptor mutations or Janus Kinase 2 (JAK2) mutations. Secondary polycythaemia is occasionally seen with renal diseases such as cystic kidney disease, tumours, renal artery stenosis and after renal transplantation. Polycythaemia associated with Nephrotic Syndrome (NS) is very rare.

This association was first described by Emmanuel DA and Wenzel FJ in 1962. Here, we describe four patients with NS who had polycythaemia at presentation. Heavy protienuria leading to renal interstitial oedema causing hypoxia and increased EPO production and high serum IL8 and increased stability of IL8 mRNA(in minimal change disease) are thought to cause secondary polycythaemia in NS. The resolution of polycythaemia following reduction in proteinuria further strengthens the causation. The exact pathogenesis remains to be elucidated.

Methods and material: This is a retrospective observational study conducted in NU Hospitals, Bengaluru, India. There were four patients with NS and polycythaemia, between February 2015 to May 2021. All patients with NS were subjected to biochemical investigations and ultrasonography (USG) of the abdomen after a detailed history and physical examination as per our hospital protocol. Coagulation profile and blood grouping are done prior to renal biopsy. Percutaneous renal biopsy was done under real-time USG guidance. The tissue obtained was sent for Light microscopic examination, Immunofluorescence microscopy and Electron microscopy. In view of the polycythaemia, all these patients underwent a Chest X Ray, arterial blood gas (ABG) analysis, serum EPO and serum JAK2 mutation studies.

Results: All four patients in our study were men. Mean age was 32.25 ± 11.70 years. Mean Hb was 18.75 ± 1.22 g/dL. Mean hematocrit was 54.67 ± 6.5 %. Mean proteinuria at onset was 7.62 ± 7.45 grams/day. Mean serum creatinine at the onset was 1.30 ± 0.58 mg/dL. None had splenomegaly. All had normal total white cell and platelet count. None were smokers. ABG was normal in all. All patients had normal serum EPO levels (mean 3.4+1.64 mU/ml) and negative JAK2 mutation analysis. The etiology of NS was minimal change disease (MCD), primary membranous nephropathy (MGN), primary focal segmental glomerulosclerosis (FSGS) and IgA nephropathy in each one of them. PLA2R antibodies were positive in

the serum and renal tissue of the patient with MGN.Immunosuppression was initiated in all patients except the patient with IgA nephropathy who had significant chronicity. Three out of four patients had resolution of polycythemia in parallel with remission of NS.The patient with MCD was detected to have polycythaemia during a relapse.

Conclusion: This highlights polycythaemia as an unusual complication of NS.Most cases of NS and polycythemia documented in the literature correspond to FSGS, mainly associated with polycythaemia vera. There are case reports of secondary polycythaemia associated with primary FSGS, MGNand MCD. We didn't go IL8 and IL11 assays in our patients. To the best of our knowledge ours is probably the largest case series of polycythaemia associated with NS.