Amyloidosis is characterized by tissue deposition of misfolded proteins that can lead to a renal failure. There are multiple types of amyloidosis. One rare type is fibrinogen alpha chain amyloidosis (AFib). We report a unique case of de novo AFib amyloidosis in a kidney transplant recipient.

This is a 71 year old white male patient with history of ESRD treated with hemodialysis from 2011 to 2018. His ESRD was due to diabetes and HTN. He had deceased donor kidney transplantation on April 2018. Family history was remarkable for sister with ESRD due to diabetes.

From January to April 2021, his serum creatinine had fluctuated from 1.28 to 1.45 mg/dL. His work-up showed negative donor specific antibodies, bland urinalysis, and negative polyomavirus and cytomegalovirus serologies. A TruGraf Blood Gene test in June 2021 showed changes suggestive of rejection.

On August 2021, his creatinine increased to 1.71 mg/dL. He also developed lower extremity swelling and neuropathy. A transplant kidney biopsy showed amyloidosis on Congo-Red Stain with a positive IgM and C4d staining, but a negative for kappa and lambda chains. Mass spectrometry study showed fibrinogen alpha chain amyloidosis. Further genetic test showed point mutation with p.E545V variant.

AFib amyloidosis is caused by point mutation of fibrinogen A gene on chromosome 4 and the most common form is from the p.E545V mutation. This mutation causes abnormal fibrinogen A molecule deposition in tissues. This disease can leads to ESRD and can recur in transplanted kidney as well. However, *de novo* A Fibrinogen amyloidosis has never been reported. Our case is the first report of this condition.

Amyloidosis is usually seen as a renal disease in the native kidney. Our case showed how this disease can occur *de novo* after transplantation. This case shows how hematological disorders affect kidney transplants. These diseases should be suspected in a kidney transplant patient with unexplained acute kidney injury and systemic symptoms. Prompt recognition of this disease can help in management and preservation of allograft function. Treatment of AFib Amyloidosis usually consists of supportive care. Patient with AFib Amyloidosis with progression ESRD usually require liver or liver-kidney transplant evaluation.

40

RENAL RECOVERY AFTER TIPS IN A PATIENT WITH HRS ON HEMODIALYSIS FOR 8 MONTHS:

Sreekari Kedarisetti[1], Adel El-Hennawy[1], Divya Rajmohan[1], Daniel Contractor[2], Winston Lee[1], Elena Frolova[1]. [1]NYC Health+Hospitals Coney Island Hospital; [2]NYC Health+Hospitals Queens Hospital Center

Hepatorenal syndrome (HRS) is a functional renal impairment in advanced cirrhosis due to severe vasoconstriction of renal circulation. We present a rare case of renal improvement and discontinuation of hemodialysis after 8 months of treatment in a cirrhotic patient after transjugular intrahepatic portosystemic shunt (TIPS) placement.

A 60-year-old man with hypertension & alcohol abuse of 40 years presented with complaints of abdominal distension that started 2 months prior to admission. He was diagnosed with biopsy proven alcoholic liver cirrhosis. Initially, his renal function was normal (Creatinine: 0.79mg/dl). At the next admission, he developed AKI and all causes, including infections, obstruction, and hypovolemia were ruled out. He was diagnosed with HRS (BUN: 130mg/dl, Creatinine: 6.46mg/dl) and treated with midodrine, octreotide & intravenous albumin. He did not respond to medical management requiring hemodialysis (HD). During the next 4 months, the patient had multiple hospitalizations with similar complaints and recurrent tense ascites necessitating paracentesis. After getting a second opinion, the patient underwent TIPS placement due to the need for weekly ascitic taps for refractory ascites following which ascitic taps markedly decreased, kidney function improved and HD was held. Ultimately, patient tolerated discontinuation of HD with stabilization of BUN (25 mg/dl) & creatinine (1.68 mg/dl).

HRS is known to improve after TIPS due to reduction in portal hypertension. Our patient had partial renal recovery and did not require dialysis after 8 months of receiving thrice weekly sessions. His kidney function stabilized 4 months post TIPS. To our knowledge, this is the first such case report of discontinuation of long-term HD after TIPS in a patient with HRS. Of the several small case series of TIPS in patients with renal disease, there was improvement in underlying renal function after the procedure in some instances.

In ESLD patients on HD, clinicians should consider TIPS as it not only helps management of ascites but also can significantly improve renal function with an opportunity to discontinue HD even after many months of dialysis treatment.

41

LOW OXALATE-DEGRADING ACTIVITY IN FECAL MICROBIOTA IS ASSOCIATED WITH HIGH SERUM INDOXYL SULFATE AND PLASMA OXALIC ACID CONCENTRATIONS IN DIALYSIS PATIENTS:

Natalia Stepanova[1], Lesya Korol[1], Ganna Tolstanova[2], Iryna Akulenko[2]. [1]State Institution "Institute of Nephrology of the National Academy of Medical Sciences of Ukraine"; [2]Taras Shevchenko National University of Kyiv

The gut microbiota has been suggested to play a crucial role in the accumulation of uremic toxins and impaired oxalate metabolism in end-stage kidney disease (ESKD) patients. [1] However, the association between gut microbiome-derived indoxyl sulfate (IS) and oxalate-degrading activity (ODA) in fecal microbiota in ESKD patients has never been investigated. The present study aimed to determine the association between total fecal ODA, serum IS and plasma oxalic acid (POx) concentrations in ESKD patients.

A total of 78 ESKD patients and 18 healthy volunteers were enrolled in this cross-sectional pilot study. Among the patients, were 33 (42.3%) patients treated with peritoneal dialysis and 45 (57.7%) hemodialysis patients. Total fecal ODA was evaluated using the method of redoximetric titration with $KMnO_4$ and expressed in % oxalate degradation per 0.01 g of feces. Serum IS and POx concentrations were measured using the spectrophotometry method.

For statistical analysis, we used the Kruskal-Wallis test and the Spearmen correlation test.

Total fecal ODA ranged from -23 to 24 %/0.01 g of feces and was statistically lower in the ESKD patients compared with the healthy volunteers [3.0 (-4.5-8) vs 4.5 (2-15.5) %/0.01 g of feces, $p = 0.03$]. The ESKD patients with positive total fecal ODA status had significantly lower serum IS [28.1 (15.3-41.2) vs 55.2 (35.1-82.7) μg/mL, $p < 0.001$] and POx [26.8 (24.1-35.7) vs 50.0 (42.2-78.2) μmol/L, $p < 0.001$] levels compared with the patients with negative ODA status. Total fecal ODA was inversely associated with serum IS ($r = -0.41$, $p < 0.001$) and POx ($r = -0.67$, $p < 0.001$) concentrations, while IS and POx levels had a strong direct association accordingly ($r = 0.75$, $p < 0.001$).

The results of our study have provided preliminary evidence that a decrease in total ODA in fecal microbiota is associated with serum IS and POx elevation in ESKD patients.

42

TREATMENT RESISTANT PRIMARY MEMBRANOUS NEPHROPATHY DUE TO CONCOMITANT OCCURRENCE OF SECONDARY MEMBRANOUS NEPHROPATHY FROM MERKLE CELL CARCINOMA.:

Tariku T. Gudura[1], Leal Herlitz[1], Hernan Rincon-Choles [1]. [1]Cleveland Clinic

Phospholipase A2 receptor antibody (PLA2RAb) causes primary membranous nephropathy (PMN). PMN may occur concomitantly with secondary membranous nephropathy (SMN). We present a case of nephrotic syndrome (NS) due to PMN occuring with SMN from Merkle cell carcinoma (MCC) with paraneoplastic syndrome (PS).

A 78 year-old male with history of hypertension and chronic kidney disease stage 3b with baseline serum creatinine (SCr) of 1.6 mg/dL (0.6 - 0.9) was referred for evaluation of NS with persitent proteinuria of urine protein to creatinine ratio (UPCR) of 4.6 mg/g despite treatment with lisinopril. Lab showed SCr 1.7 mg/dL, serum albumin 2.9 g/dL and UPCR of 6.2 mg/g, with negative autoimmune and virology testing. Kidney biopsy showed type 1 membranous nephropathy (MN); PLA2RAb was positive at 1:620 and cancer workup was negative for secondary causes of MN. He was treated with lisinopril, diuretics, and immunosuppression (IS) with tacrolimus, prednisone, and rituximab 1g IV given on days 0, 14, and 180. Two months after starting IS he developed left renal vein thrombosis. Anticoagulation led to hemorrhage. Three months after starting IS he developed ataxia, progressive dementia, and right parotid Merkell cell carcinoma (MCC), treated with radiotherapy. Eight months after starting IS he developed