台灣腎臟醫學會
中部地方會

【第 246 次腎臟學學術討論會】

節目表及摘要

第 246 次腎臟學學術討論會

時　　間：111 年 6 月 26 日(星期日)下午 2:00 – 4:15

地　　點：線上會議

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♣ 第 246 次腎臟學學術討論會 ♣

【病例討論】　主持人：林柏松 陳呈旭

14:00 － 14:15　1. Tacrolimus toxicity due to drug interaction with intravenous Nicardipine after renal transplantation
蕭榮隆、林柏松
童綜合醫療社團法人童綜合醫院腎臟科

14:15 － 14:30　2. A rare case of lipoprotein glomerulopathy
黃新迪[1] 許永禎[2] 吳致瑩[2] 游棟閔[1] 陳呈旭[1] 吳明儒[1]
[1]台中榮民總醫院內科部腎臟科、[2]台中榮民總醫院病理部

14:30 － 14:45　3. Systemic sarcoidosis with acute renal failure
施佳瑜[1] 吳軍毅[2]
台中榮民總醫院腎臟科

14:45 － 15:00　4. Hypokalemia as an initial presentation of congenital adrenal deficiency (CAH) due to 17-alpha hydroxylase deficiency in a 4-year-old girl
方華美、蔡政道、林炫沛、丁瑋信
馬偕兒童醫院小兒科

15:00 － 15:15　5. Atypical hemolytic uremic syndrome induced by acute gastroenteritis in a 3-year-old boy
鄭裕橙[1]、蔡政道[1]、徐世達[1]、趙彥鈞[1]
[1]馬偕兒童醫院小兒科

15:15 － 15:30　6. Acute kidney injury induced by secondary oxalate nephropathy due to exocrine pancreatic insufficiency by pancreatic cancer
劉冠宏[1], 田亞中[1], 鄭雅蓮[1], 廖述寬[1], 陳泰迪[2], 劉守璿[1]
[1]林口長庚醫院腎臟科, [2]林口長庚醫院病理科

15:30 － 15:45　7. Thymoma-associated minimal change disease
馬立宜[1], 塗昆樺[1], 陳泰迪[2], 葉琦如[2], 田亞中[1].
[1]林口長庚醫院腎臟科, [2]林口長庚醫院病理科

15:45 － 16:00　8. Simultaneous IgG4-related interstitial nephritis and membranous nephropathy in a patient with nephrotic syndrome
吳逸然[1,3], 塗昆樺[1], 陳泰迪[2],田亞中[1], 洪振傑[1]
[1]林口長庚醫院腎臟科, [2]林口長庚醫院病理科

16:00 － 16:15　9. A case of IgG4-related disease
*林益民[1]、邱炳芳[1]
[1]彰化基督教醫院內科部腎臟科

【1】

腎移植後使用針劑 Nicardipine 降血壓藥造成的 Tacrolimus 腎毒性

Tacrolimus toxicity due to drug interaction with intravenous Nicardipine after renal transplantation

蕭榮隆、林柏松
童綜合醫療社團法人童綜合醫院腎臟科

We present a case of insidious reversible nephrotoxicity because of a drug interaction between Tacrolimus and intravenous Nicardipine.
Tacrolimus is a calcineurin inhibitor (CNI) commonly used for immunosuppression following renal transplantation. The therapeutic benefits in the prevention of rejection are well established, but high Tacrolimus levels are associated with acute nephrotoxicity. Both acute reversible and irreversible CNI nephrotoxicity have been found. CNI-induced nephrotoxicity is due to arteriolar vasoconstriction, which is mediated by overexpression of endothelin leading to interstitial fibrosis and is carried significant risk for renal allograft loss. Calcium Channel Blockers (CCBs) such as Nicardipine are commonly used to manage hypertension both before and after renal transplantation. Nicardipine inhibits the cytochrome P450 enzyme CYP3A4, which along with CYP3A5 participates in the metabolic pathway for Tacrolimus in the liver. Use of Tacrolimus together with Nicardipine may increase the risk of supra-therapeutic levels of Tacrolimus.
Not all abnormal post-transplant renal allograft losses are due to surgical technique or embolic events. This case highlights the potential for complications because of a drug toxicity and interactions between CNIs and CCBs. Such drug combinations are to be expected in the population requiring renal transplantation as patients with end-stage renal disease (ESRD). They are often hypertensive and persistent hypertension post-operatively can shorten renal allograft survival.
A careful review of the drug history and drug drug interaction is important for post-operative care and evaluation of the renal allograft.

Keywords: Tacrolimus, Nicardipine, Renal transplantation

【2】

異常高血脂肪蛋白質性腎絲球病變

A rare case of lipoprotein glomerulopathy

黃新迪[1] 許永禎[2] 吳致瑩[2] 游棟閔[1] 陳呈旭[1] 吳明儒[1]
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Lipoprotein glomerulopathy(LPG) is a very rare disorder of **renal lipidosis** that was characterized by **lipoprotein thrombi in glomeruli.** Serum lipid profile discloses an abnormal plasma lipoprotein profile that resembles type III hyperlipoproteinemia, and a remarkably increase in serum apolipoprotein E(apo E) concentrations. The first case was reported in a Japanese patient in 1989.

A case of nephrotic syndrome with daily urine protein 8413 mg/day visited our nephrology outpatient destination on 2020.

According to patient's statement, he suffered from persistent onset of severe lower legs edema two months ago prior to this admission. He is a worker of plastic modeling . His ADL was totally independent. Neither travel history nor insect-bite was mentioned simultaneously. Biochemistry profile reported that total cholesterol: 291 mg/dl, LDL: 179 mg/dl, triglyceride: 223 mg/dl , HDL: 51 mg/dl was noted. Kidney biopsy was performed. Renal pathological findings showed: lipoprotein glomerulopathy with membranous glomerulonephritis. Based on the findings, Cyclosporine(CsA) , Valsartan , Methylprednisolone were administered for the patient and that his daily urine protein decreased to the level of 2195 mg/day.

Lipoprotein glomerulopathy is a very uncommon disease in our daily practice. High index of suspicion is warranted in cases with membranous glomerulonephritis. In the present case, it is responsive to current immunosuppressant agents.

【3】

以急性腎衰竭表現的 sarcoidosis

Systemic sarcoidosis with acute renal failure

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Chia-Yu Shih[1], Chun-yi Wu[2]
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This 64 year-old patient presented with polyuria and body weight loss for one month. Laboratory examination reported deteriorating renal function(creatinine 1.79 mg/dl->8.75mg/dl) and hypercalcemia(13.7mg/dl). Urine analysis revealed hematuria(RBC-5-10 /HPF) and pyuria (WBC 10-20 /HPF). Spot urine protein was 677mg/g. Tracing back her medical history, she had regular followed up at OPH OPD for uveitis. Steroid and cyclosporine were gradually tapered off within this year. We did a serial survey for AKI. Autoimmue profile only disclosed positive ANA titer. Serum/urine IFE both showed no monoclonal band. High levels of angiotensin converting enzyme (71.2 IU/l/37°C) was also noticed. Renal ultrasound revealed relatively normal size but increased echogenicity of bilateral kidneys. Ultrasound-guided percutaneous renal biopsy was performed smoothly on 2022/3/30. Pathology reported non-caseating granulomatous tubulointerstitial nephritis with giant cells. Non-contrast chest CT indicated multiple enlarged lymph nodes (length<4.8cm) at visible mediastinum and bilateral hilum. Combined with the clinical presentation and histomorphology, systemic sarcoidosis with renal involvement was diagnosed. Due to persistent hypercalcemia and uremia symptom, we added steroid and started hemodialysis. Due to much improvement of clinical condition, we tapered hemodialysis frequency to once a week.

Key words：
Sarcoidosis，acute kidney injury，hypercalcemia

【4】

4 歲大女童因先天性腎上腺增生症 (17-alpha 羥化酵素缺乏症)造成的低血鉀

Hypokalemia as an initial presentation of congenital adrenal deficiency (CAH) due to 17-alpha hydroxylase deficiency in a 4-year-old girl

方華美、蔡政道、林炫沛、丁瑋信
馬偕兒童醫院小兒科
Hua-Mei Fang, Jeng-Daw Tsai, Shuan-Pei Lin , Wei-Hsin Ting
Department of Pediatrics, MacKay Children's Hospital

A 4-year-old girl was referred to our outpatient department due to persistent hypokalemia diagnosed after acute viral enterocolitis. She was admitted at the previous hospital due to fever, cough, vomiting and diarrhea for a few days. Initial laboratory tests resulted in non-specific findings except hypokalemia (2.8 mEq/L) and positive rotavirus antigen test. Alkalosis (pH 7.495, pCO2 26, HCO3 20.5) and normal Mg level (2.2 mEq/L) were noted. During hospitalization, lowest potassium level to 1.9 mEq/L and urine loss of K and chloride were noted. When discharged, her potassium level was 2.6 mEq/L under potassium supplement. To rule out Bartter syndrome, she was referred to our hospital. His initial BP was elevated (119/88 mm/Hg). Recheck laboratory data after holding medications for one week showed hypokalemia (2.6 mEq/L), metabolic alkalosis (pH 7.46, HCO_3^- 26 mmol/L) and high rate of potassium excretion (fractional excretion of potassium 17.19%, potassium to creatinine ratio 1.6 mEq/mg, TTKG 15.57), hypercalciuria (urine calcium to creatinine ratio 0.29). Renal wasting of sodium and chloride were also noted. Renal echo showed right mild hydronephrosis, no nephrocalcinosis, normal adrenal glands and rudimentary female reproductive organs. Bartter syndrome was initial suspected. However, there was no polyuria, nephrocalcinosis, recurrent dehydration, or failure to thrive. Apart from that, hypertension and elevated ACTH levels with low renin and cortisol are not compatible with Bartter syndrome. Chromosome studies and whole exome sequencing was performed. The karyotype of the patient was 46, XX, and the analysis of CYP17A1 gene by Sanger sequencing revealed a novel homozygous deletion cc985_987delTACinsAA. ACTH stimulation test resulted in poor response of 17-OH progesterone which could be seen in congenital adrenal hyperplasia. The karyotype of the patient was 46, XX, and the analysis of CYP17A1 gene by Sanger sequencing revealed a novel homozygous deletion cc985_987delTACinsAA which led to isolated 17-alpha hydroxylase deficiency. Same mutation is found in both parents. CAH due to 17-alpha hydroxylase deficiency can explain hypokalemia, metabolic alkalosis, high renal wasting of electrolytes, hypertension and low cortisol level. It is a rare autosomal recessive disease and accounts for only 1% of all CAH cases. Lack of genital ambiguity in girls could cause a delay in diagnosis even until puberty if further evaluation did not perform. Normal K level and blood pressure was followed without potassium supplement or antihypertensive drugs after low dose cortisone acetate was prescribed. Meticulous assessment of clinical and laboratory presentations and genetic testing is necessary for accurate diagnosis.

Key word: 低血鉀、代謝性鹼血症、先天性腎上腺增生症、17-alpha 羥化酵素缺乏症, hypokalemia, metabolic alkalosis, congenital adrenal hyperplasia, 17-alpha hydroxylase deficiency

【5】

一位三歲男童因急性腸胃炎引發之非典型溶血尿毒症候群

Atypical hemolytic uremic syndrome induced by acute gastroenteritis in a 3-year-old boy

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A 3-year-old boy was brought to our emergency department with fever, blood-tinged diarrhea, vomiting, poor activity and appetite for 2 days. Physical examination showed tympanic sound on abdominal percussion with mild non-localized tenderness and hypertension (133/61 mmHg). Laboratory examinations revealed leukocytosis (12,200/uL) with elevated CRP level (6 mg/dL). There were no electrolyte imbalance and hypoglycemia. Empiric antibiotics with ceftriaxone and metronidazole were prescribed for suspected severe intra-abdominal infection and sepsis. After 2-day course of antibiotic treatment, her fever and diarrhea persisted, and she developed severe abdominal pain with decreased urine output. Follow-up laboratory examination found increased BUN (62 mg/dL) and creatinine (5.3 mg/dL), anemia (Hb: 11 g/dL), thrombocytopenia (Plt: 63,000/uL), and decreased CRP (3.11 mg/dL). Nephrologist was consulted due to anuria, the patient appeared tachypneic with periorbital edema and increased body weight (+3.4 kg). Atypical hemolytic uremic syndrome (aHUS) was highly suspected. Blood smear study revealed schistocytes, LDH was 2574 IU/L, suggestive of microangiopathic hemolytic anemia. Emergent renal replacement therapy with CVVH was arranged and plasma exchange (PE) started in 8 hours according to the inter-national consensus (Pediatr Nephrol. 2009 Apr;24(4):687-96.) after diagnosis of aHUS and blood samples were collected for gene and other studies. Stool was negative for detection of *E. coli* O157:H7. Coombs test, viral studies (includes CMV, EBV, HAV, HBV, HCV, SARS-CoV-2), FilmArray respiratory panel, vasculitis associated antibodies were negative. Blood and stool cultures were negative. There was no cobalamin C defect and ADAMTS13 level was normal. Urine appeared after 5-day course of PE and CVVH, then furosemide was given, CVVH was weaned off after 10 days of treatment with optimal urine output (2.3 ml/kg/hr) and other clinical parameters improved. After 4 weeks of standard PE course, catheter-related infection with *Serratia marcescens* occurred, it was well controlled under adequate antibiotic use after catheter removal. The 5th week course of PE was changed to fresh frozen plasma transfusion, and creatinine level decreased to 0.5 mg/dL at discharge. The genetic studies (WES) for aHUS are still undergoing. Here, we report a 3-year-old boy with severe aHUS which is successfully managed by prompt diagnosis and aggressive plasmapheresis.

Key words: Atypical hemolytic uremic syndrome, acute kidney injury, therapeutic plasma exchange, continuous venovenous hemofiltration

【6】

胰臟癌導致的胰外分泌不全引起的繼發性草酸腎病變之急性腎損傷

Acute kidney injury induced by secondary oxalate nephropathy due to exocrine pancreatic insufficiency by pancreatic cancer

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Kuan-Hung Liu[1], Ya-Chung Tian[1], Ya-Lien Cheng[1], Shuh-Kuan, Liau[1], Tai-Di Chen[2], Shou-Hsuan Liu[1].
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Acute oxalate nephropathy can be due to primary or secondary hyperoxaluria. The potential etiologies of secondary hyperoxaluria may be due to enteric hyperoxaluria, ethylene glycol ingestion, increased intake of oxalate containing foods, or excessive vitamin C intake. Here we presented a case with underlying pancreatic head well differentiated neuroendocrine tumor, who developed acute kidney injury with biopsy proven oxalate nephropathy.

This is a 67-year-old female with underlying disease of pancreatic head well differentiated neuroendocrine tumor with portal vein compression and superior mesenteric artery invasion, complicated with obstructive jaundice, status post palliative chemotherapy with Dacarbazine during 2020/07/14-2021/10/21.

Acute kidney injury was found accidently during routine Outpatient Clinic visit then she was admitted to our hospital. The patient only had vomit 2 times with food content in two days. She denied fever, loss of appetite, abdominal pain, diarrhea, urinary symptoms or decreased urine amount. She also denied NSAID use and iodinated contrast media exposure recently.

Laboratory data revealed elevated creatinine level (Cr 1.19->2.90->5.79 mg/dl). Blood culture yielded Klebsiella variicola and Enterococcus faecium, so Ceftriaxone then Augmentin according to sensitivity test was applied. Coffee ground vomitus was also noted during admission. However, renal function still deteriorated although after blood transfusion and antibiotic use. Kidney biopsy was arranged and revealed oxalate nephropathy. Finally, renal function improved gradually after pancreatic enzyme, calcium supplementation and cholestyramine used.

Overall, secondary oxalate nephropathy is a rare but severe condition, and up to 40% of patients progressed to *End stage kidney disease*. Physicians should be alert with hyperoxaluria-enabling conditions because prompt recognition and management may improve renal outcomes.

Keyword: oxalate nephropathy, acute kidney injury, Pancreatic cancer

關鍵字：草酸腎病變，急性腎衰竭，胰臟癌

【7】

胸腺瘤相關之微小變化腎病變

Thymoma-associated minimal change disease

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Thymoma is one kind of rare malignancy which originates from epithelial cell in the thymus. Considering the original lymphoid cell papulation, thymoma frequently brings paraneoplastic syndrome with autoimmune features, such as myasthenia gravis and bullous pemphigoid. We report a case who came to clinics with initial presentation of nephrotic syndrome. Thereafter, she suffered from oliguric renal failure, recurrent opportunistic infections, progressive muscle weakness. Finally, thymoma involving anterior mediastinum was diagnosed.

A 56-year-old woman has underlying disease of bullous pemphigoid. She presented to local hospital with acute legs edema for one month, and nephrotic syndrome was diagnosed. Kidney biopsy on June 10th, 2021, revealed podocytopathy, favored minimal change disease (MCD) or focal segmental glomerulosclerosis (FSGS). Prednisolone 60mg per day was given. However, she was admitted in July for herpes zoster, followed by acute hypoxic respiratory failure status post intubation and ventilation support during August 8th to 13th. Pneumocystis jirovecii pneumonia (PJP) and cytomegalovirus (CMV) pneumonitis were diagnosed. After completed treatment, she was discharged and came to our hospital for further management of kidney disease. Right pneumothorax owing to former PJP infection was found. Besides, oliguric renal failure happened. Hemodialysis was initiated since October 14th. Bilateral lower legs progressive weakness and ptosis were also noted. Myasthenia gravis was diagnosed based on positive anti-cholinergic receptor antibody and repetitive stimulation test. After reviewing chest CT and correlating to history, thymoma involving anterior mediastinum with multiple paraneoplastic syndromes was impressed. Operation for tumor resection was performed on November 17th. Pathology report proved the diagnosis of thymoma WHO type A, Masaoka-Koga stage IIa, 8th AJCC pT1N0M0 stage 1. She was kept in end-stage renal disease with dialysis dependence even after surgical resection of thymoma.

Minimal change diseases were occasionally associated with thymoma. Clinical presentations and treatment responses varied in the reported cases. Thymoma should be taken into consideration for older patients or patients with poor response to conventional treatment.

Keyword: Thymoma, nephrotic syndrome, minimal change disease (MCD)
關鍵字：胸腺瘤，腎病症候群，微小變化腎病變

【8】

在一腎病症候群患者同時表現膜性腎病變及 IgG4 相關間質性腎炎

Simultaneous IgG4-related interstitial nephritis and membranous nephropathy in a patient with nephrotic syndrome

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Immunoglobulin G4-related disease ((IgG4-RD) is a systemic immune-mediated disease that typically manifest as fibroinflammatory masses affecting multiple organs, including pancreas, salivary glands, biliary tract, liver, lung, and kidney. IgG4-RD is well recognized in the form of tubulointerstitial nephritis (IgG4-related TIN), which may present either as a mass-like lesion or renal failure. IgG4-related membranous nephropathy is much less common than IgG4-related TIN, but these two renal complications sometimes occur together. Here we report a case of nephrotic syndrome with simultaneous IgG4-related TIN and membranous nephropathy.

A 74-year-old male with underlying coronary artery disease, type 2 diabetes mellitus, hypertension, and gout, came to our clinic with complaint of acute-onset foamy urine in 1 month. After initial work-up, acute nephrotic syndrome was impressed based on presentations of severe hypoalbuminemia (serum albumin: 2.04 g/dL), heavy proteinuria (9.3g/day) and hypertriglyceridemia. Serologic studies also revealed elevated IgG, IgG4 and hypocomplementemia. Renal biopsy was performed which revealed membranous nephropathy and tubulointerstitial nephritis. IgG4/IgG ratio in the interstitial area is over 40%. Immunohistochemistry (IHC) stain for PLA2R1 was negative. Whole body tumor scan showed right supraventricular and mediastinal lymphadenopathies; no bulky mass was found in favorite location of IgG4-RD. IgG4-related interstitial nephritis with secondary membranous nephropathy was diagnosed. Glucocorticoid were given. Complete remission was achieved rapidly.

According to literature review, patients with IgG-related TIN frequently have association with IgG4-related condition in other organs, especially the pancreas. In addition, most of these patient are likely to be profoundly hypocomplementemic. Glomerular lesions could be found in some of this patients, mostly membranous nephropathy. IHC stain for PLA2R1 in these patients are exclusively negative Patients of IgG4-related disease with kidney involvement mostly response to glucocorticoid well.

Keyword: IgG4-related disease, tubulointerstitial nephritis, membranous nephropathy.
關鍵字：IgG4 相關性疾病，腎小管間質性腎炎，膜性腎病變。

【9】

IgG4 相關疾病-個案報告

A case of IgG4-related disease

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IgG4-RD is a recently recognized clinical entity that often involves multiple organs and is characterized by high levels of serum immunoglobulins G4, dense infiltration of $IgG4^+$ cells and storiform fibrosis. Cellular immunity, particularly T-cell mediated immunity, has been implicated in the pathogenesis of IgG4-RDs. The most frequent renal manifestations of IgG4-RD are IgG4-related tubulointerstitial nephritis, membranous glomerulopathy and obstructive nephropathy secondary to urinary tract obstruction due to IgG4-related retroperitoneal fibrosis.

This 64-year-old male has medical history of Acute kidney injury on chronic kidney disease suspect acute interstitial nephritis s/p renal biopsy on 2014-10-30、Right hydronephrosis from bilateral retroperitoneal fibrosis、Hypertension、Hyperuricemia、Bilateral retroperitoneal fibrosis with hydronephrosis s/p D-J insertion since 2012-11

Due to bilateral retroperitoneal fibrosis with hydronephrosis, the patient received bilateral D-J insertion, and change tube every three months.

This time, persist elevated renal function and right hydronephrosis were noted. Thus, renal biopsy was suggested.

During admission, the renal biopsy was performed. The biopsy reports: 1. tubulointerstitial nephritis and acute tubular injury; 2. hypertensive glomerular nephrosclerosis. The IgG subclass revealed IgG4: 380mg/dL. Thus, IgG4 related disease was impressed.