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Succsessful Treatment of aSLE Patient with Acute Renal Cortical Necrosis

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1. Abstract

Acute Renal Cortical Necrosis (ARCN) is characterized by the destruction of the renal cortex with poor prognosis. Here, we reported a Chinese female patient presenting abrupt anuria, acute kidney injury and heart failure. Examination revealed proteinuria, hematuria, anemia, thrombocytopenia, acute renal injury and acute left heart failure. Serologic tests showed positive antinuclear antibody, positive anti-double-stranded DNA antibody, positive antiphospholipid antibody, and low serum complement C3 level. Magnetic resonance imaging confirmed the diagnosis of ARCN. Transthoracic echocardiography images showed mitral valve thickening with calcificated vegetations and severe regurgitation. Renal and heart function improved after renal replacement therapy and mitral valve replacement surgery. Six months later, her urine volume returned to normal and she discontinued dialysis one year later. This is a case showing severe complication of Systemic Lupus Erythematosus (SLE)-Associated with Antiphospholipid Syndrome (APS), and the renal function improved obviously after immunosuppressive therapy.

2. Keywords: Lupus;Antiphospholipid antibody; Mitral regurgitation;Cardio-renal syndrome

3. Introduction

Acute Renal Cortical Necrosis (ARCN) is rare to cause acute kidney injury by destructing the renal cortex severely. Non-obstetric causes of ARCN have been steadily increased since the 1970s[1,2]. Libman and Sacks originally reported Systemic Lupus Erythematosus (SLE) cases with verrucose vegetative endocarditis and since then, Libman-Sacks Endocarditis (LSE) has become a typical cardiac manifestation of SLE. Here, we report a case of ARCN with LSE and SLE associated Antiphospholipid Syndrome (APS).

4. Case Report

A 20-year-old girl was admitted to our Kidney Intensive Care Unit department on the 7th of May 2015, with proteinuria, hematuria, anemia, thrombocytopenia and serum creatinine elevation.

From the second of May 2015, her serum creatinine increased from 82μmol/l to 241μmol/l and she was administered with Methyl Prednisolone (MP) 40mg/day and Ceftazidime, 2g twice daily for fever and pulmonary inflammation. On admission, blood pressure was 113/77mmHg, pulse was 72 bates/min, temperature was 36.8°C, and oxygen saturation was 100% on room air. Urine output in 24 hours was 1200ml. The rest of the physical examination was unremarkable. Electrocardiogram was normal. Biological tests showed the following values: serum creatinine 384 μmol/l, potassium 4.57 mmol/l, hemoglobin 79 g/l, white blood cells 5.8×10^9/L, platelets 78×10^9/L, reticulocyte 2.35 (0.5-1.5)%, lactate dehydrogenase (LDH) 523 (60-240) U/L, Brain Natriuretic Peptide (BNP) 1180pmol/L, aspartate aminotransferase 11 U/L, alanine aminotransferase 18 U/L, prolonged activated thrombin time 30.8s (ratio 0.8), C-Reactive Protein (CRP) 41 mg/L, Procalcitonin (PCT) 1.21μg/L, and no evidence of schistocytes on blood smear. ANA was present at a titer of 1:256; anti-double-stranded DNA antibody (A-dsDNA) was positive; anti-β2GPI antibody, lupus anticoagulant, anti-C1q antibody, MPO-ANCA and PR3-ANCA were all negative. Anticardiolipin Antibodies (ACL)-Immunoglobulin (Ig) G, ACL-IgA and ACL-IgM titer were 33.92 (<12) IU/ml, 15.24 (<12) IU/ml and 13.94 (<12) IU/

ml respectively. ACL was determined by using standardized enzyme-linked immunosorbent assay. Serum C3 and C4 were low at 0.5g/l and 0.1g/l respectively. CD4 lymphocyte cell, CD 8 lymphocyte cell and CD20 lymphocyte cell were 181/μl, 369/μl and 240/μl respectively. Coomb's test was positive. Antibody to ADAMTS 13 was negative and the activity of ADAMTS 13 was normal. Blood culture was negative. Computed Tomography (CT) of chest showed mild pneumonia with pleural and pericardial cavity effusion. Kidney ultrasound showed two normal-sized kidneys. On the second day after admission, her temperature elevated to 38.4°C, and urine output decreased to 510ml/24 hours. On the third day after admission, her urine out decreased further to 90ml/24 hours; serum creatinine, potassium, LDH, PCT, CRP and BNP elevated to 667μmol/L, 5.23mmol/L, 897U/L, 1.39μg/L, 66.5mg/L and 4138 pmol/L (Figure 1) respectively. The patient urgently underwent haemodialysis and received impulse MP 500mg/d for 3 days and Biapenem, 300mg/8 hours, which rapidly improved fever. Then she administered with corticosteroids, Cyspin and hydroxychloroquine for SLE.

Because of sudden anuria and rapidly progressive failure in the absence of evidence of urinary tract obstruction, ARCN suspected, Magnetic Resonance Imaging (MRI) of bilateral renal was performed immediately. Imaging study showed bilateral and total cortical necrosis. The patient remained anuria, underwent hemodialysis 3-4 times a week.

In July 2015, the girl presented with cough and hemoptysis. The transthoracic echocardiography showed mitral thickening and severe regurgitation and calcificated vegetations on the mitral valve with left heart cavity expanded. The Ventricular Ejection Fraction (LVEF) decreased from 60% to just 34%. A valve replacement surgery was performed uneventfully. The patient presented no symptom of left heart failure after surgery. The culture of the excised vegetation was negative. Histopathological examination of vegetation revealed inflammation with neutrophil infiltration and fibrin-platelet thrombi formation.

Characteristics of the calcificated vegetations and mitral regurgitation on cardiac imaging, combined with the clinical evidence of active SLE, provide strong support for the diagnosis of Libman-Sacks endocarditis in SLE-associated APS. In September 2016, the patient discontinued dialysis. Her serum creatinine decreased to 1.81mg/dl in March 2019, after the therapy of cyclophosphamide 0.4g per month for 13 months. (**Figure 1**).

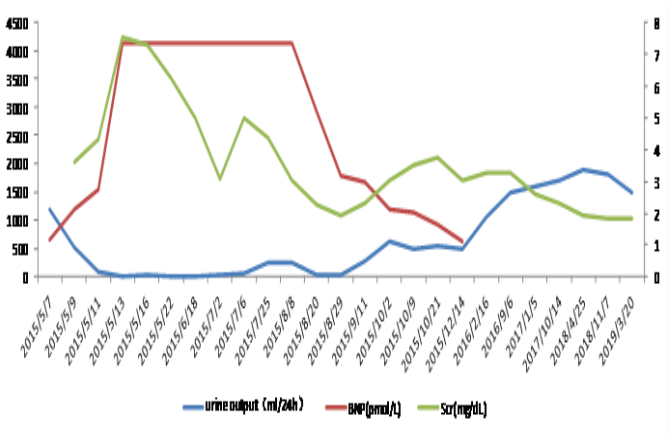

Figure 1: BNP, urine output and Scr changes.
BNP: Brain Natriuretic Peptide; Scr: Serum creatinine; MP: Methylprednisolone; IVIG: Intravenous Immunoglobin; MVR: Mitral Valve Replacement; CTX: Cyclophosphamide

5. Discussion

Antiphospholipid Syndrome (APS) was first introduced in 1983. Since almost every organ in the body has been reported to be involved in APS, it has evolved into a systemic and multidisciplinary disease. Renal thrombotic histological manifestations contain thrombotic microangiopathy, glomerular ischemic, interstitial fibrosis, tubular atrophy and arterial and venous thrombus and sclerosis [3,4].Heart valve abnormalities are the most common cardiac manifestation of APS. The lesions such as vegetations and valve thickening mostly involved the mitral and aortic valve [5, 6].

Rapid anuric acute kidney injury in a SLE-associated APS patient with endocarditis is reported here. For her anemia, thrombocytopenia and acute kidney injury, MRI of bilateral renal has been performed instead of renal biopsy or contrast enhanced CT. The MRI images documented severe bilateral renal cortical necrosis. Causes of ARCN include abruption placenta, postpartum hemorrhage or toxemia in the last century and hemolytic uremic syndrome, pancreatitis, snake bite, sepsis, drugs, trauma, allograft rejection, aortic dissection, polyarteritis nodosa and Antiphospholipid Syndrome (APS) nowadays [1,2,7]. Venous thrombosis is the usual sites of thrombosis in patients with APS, nonetheless, the kidney is major and severe target for APS with thrombosis in arteries, capillaries and veins with consequences of proteinuria, hypertension and cortical necrosis [8,9]. ARCN is characterized by necrosis of the renal cortex, sparing of the medulla and a thin layer of subcapsular cortex. Most patients develop irreversible renal failure and only very few cases with partial recovery have been reported [10]. Recovery depends on the amount of injured cortical tissue.

Libman-Sacks Endocarditis (LSE) is seen as a cardiac manifestation of SLE. Mitral and aortic valve is mostly involved. Transthoracic echocardiography is the commonly diagnostic

technique, however, blood culture even valve vegetation analysis is necessary to distinguish LSE from infectious endocarditis. Anticoagulation treatment is required for the risk of thrombo-embolic events. Guidelines also suggest long-term anticoagulation in APS[11]. For patients in stable hemodynamic condition, drug treatment is recommended; for those with severe intractable symptomatic valvular dysfunction, surgical intervention is required [12,13]suggested that repair rather than replacement was considered as a good surgical opinion for only localized abnormalities. In this case, the patient presented symptoms of acute left heart failure, surgical intervention was taken into consideration on the basis of adequate steroid and anticoagulation treatment. The heart function of this patient improved obviously after surgery, which indicated that it might be type I cardiorenal syndrome[14]. Her outcome of getting off dialysis also supported this diagnosis, especially for the effectiveness of ultrafiltration in cardiorenal syndrome[15].

We report a case of abrupt anuria and acute heart failure in SLE, whose renal function recovered successfully one year after the initiation of dialysis. Our patient showed even ACL with a low titer could cause catastrophic injury in organs such as renal and heart. For patients with severe acute kidney injury, underlying diseases call for more attention. SLE, APS, LSE and cardiorenal syndrome all played important roles in this case. Clinicians are suggested to consider ARCN when patients present with anuria suddenly and MRI is appropriate to diagnosis bilateral ARCN. Long term follow-ups after heart surgery should be continued and further immunosuppressive therapy of SLE is necessary.

6. Acknowledgement

We thank next of kin of the patient for publication of this case report and accompanying images.

7. Ethical approval

All procedures performed in studies involving human participant were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

8. Informed Consent

Informed consent was obtained from the patient reported here.

References

1. Kim HJ.Bilateral renal cortical necrosis with the changes in clinical features over the past 15 years (1980-1995). J Korean MedSci. 1995;10:132-41.

2. Prakash J, Vohra R, Wani IA,Murthy AS, Srivastva PK, Tripathi K et al. Decreasing incidence of renal cortical necrosis in patients with acute renal failure in developing countries: a single-centre experience of 22 years from Eastern India. Nephrol Dial Transplant. 2007;22:1213-7.

3. Cacoub P, Wechsler B, Piette JC, Beaufils H, Herreman G, Bletry O et al.Malignant Hypertension in antiphospholipid syndrome without overt lupus nephritis. ClinExpRhumatol. 1993;11: 479-85.

4. Griffiths MH, Papadaki L, Neild GH. The renal pathology of primary antiphospholipid syndrome: a distinctive form of endothelial injury. QJM. 2000; 93:457-67.

5. Hojnik M, George J, Ziporen L,Shoenfeld Y.Heart valve involvement (Libman-Sacks endocarditis) in the antiphospholipid syndrome. Circulation.1996; 93:1579-87.

6. Neshar G, Ilany J, Rosenmann D, Abraham AS. Valvular dysfunction in antipospholipid syndrome: prevalence, clinical features and treatment. Semin ArthritisRheum. 1997;27:27-35.

7. Wilson WA, Gharavi AE, Koike T, Lockshin MD, Branch DW, Piette JCet al.International consensus statement on preliminary classification criteria for definite APS. Arthritis Rheum.1999; 42:1309-11.

8. Nzerue CM, Hewan-Lowe K, Pierangelli S,Harris EN."Black swan in the kidney": Renal involvement in theantiphospholipid antibody syndrome. Kidney Int. 2002;62:733-44.

9. Nochy D, Daugas E, Droz D, Beaufils H, Grünfeld JP, Piette JCet al.The intrarenal vascular lesions associated with primary antiphospholipid syndrome. JAm Soc Nephrol.1999; 10:507-51.

10. Koo JR, Lee YK, Kim YS, Cho WY, Kim HK, Won NH.Acute renal cortical necrosis caused by an antifibrinolytic drug (tranexamic acid). Nephrol DialTransplant. 1999;14:750-2.

11. McMurray JJ, Adamopoulos S, Anker SD, Auricchio A, Böhm M, Dickstein Ket al.Esc guidelines for the diagnosis and treatment of acute and chronic heart failure 2012: the task force for the diagnosis and treatment of acute and chronic heart failure 2012 of the European Society of Cardiology. Developed in collaboration with the heart failure association (hfa) of the esc. Eur Heart J. 2012;33(14):1787-847.

12. Bouma W, Klinkenberg TJ, van der Horst IC, Wijdh-den Hamer IJ, Erasmus ME, Bijl Met al.Mitral valve surgery for mitral regurgitation caused by libman-sacks endocarditis: a report of four cases and a systematic review of the literature. J Cardiothorac Surg. 2010;5:13.

13. Ronco C,Cicoira M, McCullough PA.Cardiorenal syndrome type1: pathophysiological crosstalk leading to combined heart and kidney dysfunction in the setting of acutely decompensated heart failure. J Am CollCardiol. 2012;60(12):1031-42.

14. Bart BA, Goldsmith SR, Lee KL, Givertz MM, O'Connor CM, Bull DAet al. Ultrafiltration in decompensated heart failure with cardiorenal syndrome. N Engl J Med. 2012;367(24):2296.