

# NephSAP 19-2 NIO onderwijs

1. (10) A 63-year-old man presents with a 6-week history of systemic illness including malaise, weight loss, low-grade fever, and upper respiratory symptoms. He is found to have AKI with creatinine 4.5 mg/dL [398  $\mu$ mol/l] and new-onset hematuria and proteinuria. ANCA is positive, with elevated anti-proteinase 3 (PR3) titer. Renal biopsy shows pauci-immune crescentic glomerulonephritis, with 20% glomerulosclerosis. He has been prescribed pulse corticosteroids and wishes to discuss his treatment options for additional therapy.

Which of the following statements is correct regarding induction therapy for AAV?

- (A) The MYCYC trial showed that induction therapy with mycophenolate and steroids was inferior to cyclophosphamide and steroids for remission induction at 6-month follow-up
- (B) The PEXIVAS trial showed that in cases of moderate to severe renal impairment, plasma exchange was associated with reduced rates of ESKD
- (C) In the PEXIVAS trial, reduced corticosteroid dosing was associated with no difference in risk of ESKD, but with reduced rates of infectious complications
- (D) The addition of a small molecule C5a inhibitor, avacopan, to induction therapy was associated with higher rates of infectious complications as compared with standard treatment with corticosteroids and rituximab/ cyclophosphamide

2. (11) A 37-year-old woman received a diagnosis of AAV 6 months ago and was treated with rituximab and prednisone for induction therapy. Her ANCA titer became negative within 3 months of initiating therapy, and her peripheral B cell count was zero at most recent check. She has been on tapering dose steroids but remains on prednisone 5 mg daily. She has been reading about the risks of immunosuppression and wants to discuss the benefits of azathioprine versus rituximab.

Regarding remission maintenance in AAV, which of the following is correct?

- (A) In a clinical trial comparing it with maintenance therapy with azathioprine, rituximab was superior at maintaining remission of AAV
- (B) A rituximab dosing strategy based on repopulation of peripheral B cells and/or changes in ANCA titer led to fewer relapses and longer relapse-free survival than regular scheduled 6-monthly dosing of rituximab
- (C) In a study examining the duration of treatment with oral immunosuppressants to maintain remission, there were no differences in rates of ESKD associated with the use of azathioprine and prednisone for 24 versus 48 months
- (D) Recent observational studies suggest that elderly patients tolerate corticosteroids well, but the use of cyclophosphamide for induction therapy is associated with higher rates of infectious complications

- 3.** (12) You have been asked to give a presentation to your colleagues in primary care on lupus nephritis, with emphasis on the presentation of lupus nephritis, interpretation of the serologic and histologic evaluation, and recommendations for follow-up testing.

In the evaluation of lupus nephritis, which of the following is correct?

- (A)** Observational studies suggest that lupus nephritis is being diagnosed at a younger age
- (B)** The time from diagnosis of SLE to the development of lupus nephritis is shorter in African Americans and other minority groups
- (C)** In patients with proliferative lupus nephritis who undergo repeated biopsy, histologic findings correlate closely with clinical response to treatment
- (D)** Anti-PLA2R antibodies are occasionally identified in patients with class V lupus nephritis

- 4.** (13) A 28-year-old woman presents to her primary care provider with joint pain, fatigue, and a malar rash. Her laboratory evaluation shows positive ANA and anti-double stranded DNA antibodies; urinalysis shows 1+ blood and 2+ protein; serum creatinine is 0.8 mg/dL [71  $\mu\text{mol/l}$ ].

Her biopsy specimen reveals class IV lupus nephritis, and you plan to start immunosuppressive therapy. You recommend starting mycophenolate mofetil and steroids, but she asks about alternative options and about recent developments in treatment.

Which of the following is correct regarding the treatment of lupus nephritis?

- (A)** The LUNAR trial showed improved outcomes from the addition of rituximab to mycophenolate mofetil and steroids
- (B)** Post hoc analyses suggest that patients who achieve full depletion of peripheral B cells may gain greater benefit from rituximab
- (C)** Calcineurin inhibitors have shown little benefit in the treatment of lupus nephritis
- (D)** The addition of voclosporin to treatment with mycophenolate and steroids was associated with rates of remission, and adverse events that were similar to those of mycophenolate and steroids alone

- 5.** (24). A 73-year-old man with no known history of kidney disease presents to the hospital with abdominal fullness, a new rash, and decreased urine output. On examination, he is found to have nonblanching palpable purpura on his upper and lower extremities and new lower extremity edema. He also exhibits splenomegaly and has palpable bilateral cervical lymphadenopathy on examination. He reports occasional fevers and night sweats. His serum creatinine on admission labs is 3.4 mg/dL (eGFR 17 mL/min/1.73 per m<sup>2</sup>). His urinalysis results are significant for 3+ protein and 3+ blood, with 20 to 50 red blood cells per high powered field on urine microscopy, including the presence of dysmorphic red blood cells. Antinuclear antibody, anti-myeloperoxidase and anti-serine proteinase 3 antibody test results are negative, as are antibodies for HIV, hepatitis B, and hepatitis C. He undergoes kidney biopsy.

What is the most likely diagnosis?

- A** Proliferative glomerulonephritis with monoclonal immunoglobulin deposits (PGNMID-Ig)
- B** Pauci-immune, necrotizing, and crescentic glomerulonephritis
- C** Type I cryoglobulinemic glomerulonephritis
- D** Type II cryoglobulinemic glomerulonephritis

- 6.** (28). A 34-year-old woman presents to the emergency room with necrotic skin ulcers on her nose and ears, arthralgias, and myalgias. Initial laboratory results are notable for neutropenia and hematuria. A serologic evaluation reveals she has positive titers for both PR3-ANCA and MPO-ANCA.

Double-positive MPO and PR3-ANCA is associated with which of the following?

- A** Progression to end-stage renal disease
- B** Development of pulmonary hemorrhage
- C** Increased risk of mortality
- D** Drug-induced AAV