

2023

A Case of Maintenance Prednisone in ANCA Glomerulonephritis

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Recommended Citation

Sekar A, Gandhi P, Sheldon V, Reddy A, Campbell R. A Case of Maintenance Prednisone in ANCA Glomerulonephritis. *Advances in Clinical Medical Research and Healthcare Delivery*. 2023; 3(1). doi: 10.53785/2769-2779.1140.

ISSN: 2769-2779

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A Case of Maintenance Prednisone in ANCA Glomerulonephritis

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Abstract

Anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV) is a rare condition that can cause rapid renal failure. Treatment involves steroids and other immunosuppressive agents. Agents for induction include rituximab, cyclophosphamide, pulse dose steroids and avacopan.

Maintenance regimens include tapered doses of steroids, azathioprine and rituximab

We present a case of severe AAV that maintained remission with a protracted course of low dose prednisone without maintenance rituximab or azathioprine.

A 70-year-old woman was admitted for acute kidney injury (AKI), with a serum creatinine (sCr) of 6.93 mg/dL (baseline sCr of 0.9 mg/dL, nil proteinuria.) Serologic work-up was positive for P-ANCA. She required one session of hemodialysis and solumedrol was started. Biopsy showed rapidly progressive glomerulonephritis with necrotizing granulomas and severe interstitial fibrosis and tubulointerstitial atrophy (IFTA). Rituximab 375mg/m² 4 doses weekly was the induction. She maintained off dialysis and her creatinine stabilized, improving to 3.13 mg/dL over three months. Patient declined maintenance cytotoxic therapy due to concern for lowered immunity during the COVID-19 pandemic. Whenever prednisone was tapered below 10 mg, creatinine would worsen prompting a prolonged course of steroids(12 months).

AAV is a rare condition that can cause rapid renal failure. Treatment includes steroids and immunosuppressive agents, given as induction and maintenance therapies. Glucocorticoids have many side effects, and recent trials evaluate reducing cumulative steroid dose. Our report describes a patient with severe disease that required a longer than usual course of steroids to maintain remission. Her regimen presents some treatment challenges, given the current recommendations to taper steroids off sooner. However, her case is unique, as she declined traditional maintenance immunosuppression, but remained in remission with steroids alone.

Keywords

glomerulonephritis, ANCA, prednisone

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Conflict of Interest Statement

All authors have no conflicts of interest

Cover Page Footnote

All authors have contributed to the manuscript and are in agreement with submission

CASE REPORT

A Case of Maintenance Prednisone in ANCA Glomerulonephritis

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Abstract

Anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV) is a rare condition that can cause rapid renal failure. Treatment involves steroids and other immunosuppressive agents. Agents for induction include rituximab, cyclophosphamide, pulse dose steroids and avacopan.

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AAV is a rare condition that can cause rapid renal failure. Treatment includes steroids and immunosuppressive agents, given as induction and maintenance therapies. Glucocorticoids have many side effects, and recent trials evaluate reducing cumulative steroid dose. Our report describes a patient with severe disease that required a longer than usual course of steroids to maintain remission. Her regimen presents some treatment challenges, given the current recommendations to taper steroids off sooner. However, her case is unique, as she declined traditional maintenance immunosuppression, but remained in remission with steroids alone.

Keywords: Glomerulonephritis, ANCA, Prednisone

1. Introduction

Anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV) is a rare condition that can lead to rapid onset renal failure. Treatment options are typically centered on steroids and immunosuppressive agents, such as cyclophosphamide or rituximab. More recently, rapid steroid tapers are recommended with newer medications like avacopan showing promise in that regard. We present a case of severe AAV that responded to

induction with rituximab and glucocorticoids but required a protracted period of low dose steroids only (without the traditional maintenance regimen) to maintain remission.

2. Case report

A 70-year-old woman with a past medical history of diabetes mellitus and hypertension presented to the hospital for weakness and nausea. On arrival, she was hemodynamically stable, but found to have an acute

Accepted 20 February 2023.
Available online ■■■

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<https://doi.org/10.53785/2769-2779.1140>
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kidney injury, with a serum creatinine (sCr) of 6.93 mg/dL with 1+ proteinuria, up from baseline sCr of 0.9 mg/dL and nil proteinuria on dipstick. She was otherwise asymptomatic. Serologic work-up was positive for P-ANCA; otherwise negative. She required one session of hemodialysis, underwent kidney biopsy, and was started on solumedrol following return of serologic results. Her biopsy showed rapid progressive glomerulonephritis with necrotizing granulomas and severe interstitial fibrosis and tubulointerstitial atrophy (IFTA), with no underlying diabetic kidney disease. Pulse dose of solumedrol was given for 3 days and rituximab was given for induction as four doses of 375 mg/m². She was non-oliguric and stable off dialysis, and following induction, creatinine stabilized to mid-5 range, slowly improving to 3.13 mg/dL over three months.

Maintenance therapy with rituximab was recommended at 6 months post induction, but the patient declined this out of concerns for risk of lowered immunity during the COVID-19 pandemic. 6 months later, prednisone was tapered to 5 mg daily, but creatinine increased to 4.06 mg/dL. At that time, UA showed microscopic hematuria with acanthocytes suggesting an ANCA flare. Prednisone was increased back to 10 mg daily and creatinine came down to 3 mg/dL. A second time, 3 months later when prednisone was decreased to 5 mg, creatinine again went up to 3.5 mg/dL. Therefore, she was kept on 10 mg prednisone. 18 months post induction and following this increase, her most recent creatinine was 2.9 mg/dL.

3. Discussion

AAV is a rare autoimmune condition secondary to autoantibodies (ANCA) against cytoplasmic antigens (notably, MPO and PR3) which are expressed in neutrophils.¹ AAV causes inflammation and necrosis of small and medium blood vessels, leading to end organ damage, and can be renal limited or can cause systemic disease.¹ Typical treatment involves immunosuppression. Induction agents include pulse dose steroids with either rituximab or cyclophosphamide. Steroids alone are not effective for induction. Avacopan is a newer agent used with traditional regimens and trial results suggest similar efficacy with less steroid use.²

Maintenance immunosuppression includes azathioprine, rituximab (maintenance dose every 6 months) and steroid taper. Glucocorticoids have several side effects that occur with relative frequency, such as infection, hyperglycemia,

hypertension and psychiatric disturbances. As a result, recent trials have focused on reducing cumulative steroid doses and attempting steroid-sparing regimens.^{2,3,4} Typically, glucocorticoids are tapered and stopped by 6 months after treatment, if there have been no relapses or flaring of disease.¹

Our case, which describes a patient with severe disease that responded to induction with rituximab and steroids with continued improvement on glucocorticoid maintenance alone, presents unique nuances in the treatment of AAV.

First, her responsiveness despite an elevated serum creatinine of 6.93 mg/dL, severe IFTA and requiring one-time dialysis suggests that there was likely underlying active disease and inflammation amenable to steroid treatment. Certain trials excluded AAV patients with sCr >4 mg/dL or who required dialysis, but this case suggests that even very severe cases of AAV may respond to treatment.³

Second, her relapse requiring an increase in steroid dosing presents some challenges, given the current recommendations to reduce steroids quickly in AAV treatment. However, relapses occur in 30–50% of patients, and glucocorticoid withdrawal is an independent risk factor for relapse.¹ Relapses are typically treated by temporarily increasing steroid doses or giving additional dosing of cytotoxic medications, as needed for severity of relapse.⁵ For example, in the RAVE trial, patients with non-severe relapses were treated with increasing prednisone dosing to a median of 17.5 mg/day (range: 2–80 mg/day).^{6,7}

Third, her improvement and stabilization with steroids alone offers some treatment options for patients who cannot receive additional cytotoxic medications.

Ideally, our patient would have received maintenance therapy with additional rituximab or azathioprine; however, due to her preferences, she stayed on steroid therapy alone. Avacopan was not an option due to cost issues. Without traditional maintenance immunosuppression, she has remained dialysis free and although she had one relapse, this improved with only a slight increase in steroid therapy to 10 mg from 5 mg. She did not have any reported side effects from steroids and this could be from the lower dose.

In conclusion, our case of AAV treated with induction with rituximab and maintenance with glucocorticoid therapy alone presents a unique treatment option for a severe case AAV. She did not want rituximab/azathioprine due to concerns of risk

of lowered immunity during the covid-19 pandemic. This case reflects the importance of informed decision making and patient choice in complex treatment plans.

Conflict of interest

The authors have stated there are no conflicts of interest.

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