Tachyarrhythmias in Microscopic Polyangiitis Responding Well to Plasmapharesis Treatment

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Abstract  Microscopic polyangiitis (MPA) is a form of anti neutrophilic cytoplasmic antibody associated necrotizing vasculitis, which may present with a variety of clinical manifestations. Cardiac involvement in vasculitis is common and the manifestations usually include cardiomyopathy, pericarditis and arrhythmias. Our patient presented with dyspnea, hemoptysis and acute kidney injury. The patient developed frequent episodes of supraventricular tachycardia (SVT) and non-sustained ventricular tachycardia during the course of his stay, which subsequently resolved on treatment with plasmapheresis. Mortality rate is significantly high in patients with pulmonary hemorrhage and renal failure; plasmapheresis has proven to be beneficial in such acute case. Our case not only highlights the therapeutic effects of plasmapheresis on MPA but also stresses the importance of prompt plasmapheresis to mitigate MPA complications like SVT.

Keywords: microscopic polyangiitis, plasmapheresis

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1. Introduction

Microscopic polyangiitis (MPA) is a multi-systemic ANCA-associated vasculitis characterized by inflammation of small-sized vessels. Cardiac complications are commonly noted with MPA over the course of their disease. MPA usually presents with constitutional clinical features like dyspnea and acute renal failure, which may worsen acutely with cardiac and neurovascular complications. Cardiac involvement like supraventricular tachycardia (SVT) and pericarditis may dramatically worsen the prognosis and mortality in such patients. Circulating ANCA, antiproteinase-3 (anti-PR3, c-ANCA), and anti-myeloperoxidase (anti-MPO, p-ANCA) have been reported to play a major role in the pathogenesis of ANCA associated small-vascular vasculitis; these antibodies are detectable in 70–90% cases and facilitate diagnosis [1]. Our case demonstrates the effectiveness of plasmapheresis in attenuating the acute complications like SVT by elimination of circulating ANCA may stop the pathological process until corticosteroids and cyclophosphamide suppress inflammation and autoantibody production.

2. Case Presentation

A 73-year-old male with the past medical history of hypertension, prostate cancer status post prostatectomy, and chronic anemia presented to the emergency department with the complaints of worsening shortness of breath and leg swelling for 1 week. He was hemodynamically stable when he presented to ED, though he was in mild distress due to shortness of breath. His baseline investigations revealed hemoglobin of 7.2 g/dl, markedly high creatinine of 16.5 mg/dl and potassium of 6.3 meq/l. The 12-lead EKG did not show any ST or T-wave changes and 3 set of troponins were normal. Chest X ray showed bilateral pulmonary edema. He was given IV Lasix, 1 unit of packed RBC and 3 ampules (150 Meq) of bicarbonate. He underwent emergent hemodialysis for refractory hyperkalemia, acidosis and fluid overload, but his oxygen requirement increased with worsening shortness of breath and leg edema. In the mean time, patient developed massive hemoptysis and gross hematuria with acute blood loss anemia. He developed acute hypoxic respiratory failure for which he was intubated and admitted in the intensive care unit. The CT scan of the chest was ordered which showed extensive scattered ground-glass opacities throughout both lungs with probability of multifocal pneumonia (Figure 1). He was started on vancomycin and Zosyn for healthcare acquired pneumonia.

Due to his hemoptysis, hematuria and acute renal failure, ANCA vasculitis work up with renal biopsy was ordered. He was positive for anti myeloperoxidase antibodies (P-ANCA) and renal biopsy showed focal necrotizing and crescentic glomerulonephritis (RPGN pauci immune type) (Figure 2), thus supporting the diagnosis of microscopic polyangiitis. Anti GBM antibody, ANA, C3 and C4 levels were normal, He was started on cyclophosphamide and steroids, but he continued to have hemoptysis with worsening blood loss anemia requiring frequent blood transfusions. In the mean time, he started having frequent episodes of non-sustained ventricular...
tachycardia (Figure 3) and supra ventricular tachycardia. He did not develop any symptoms during ventricular tachycardia episodes and SVT was treated with adenosine. His EKG did not show ischemic changes and troponins were again normal. His potassium, magnesium and calcium levels were within normal limits. Transthoracic echocardiogram was ordered which showed ejection fraction of 65% with no valvular or wall motion abnormalities (Figure 4).

Figure 1. CT scan of the chest shows extensive scattered ground-glass opacities throughout both lungs

Figure 2. Renal Biopsy (H & E stain) shows focal necrotizing and crescentic glomerulonephritis
He was started on plasmapheresis treatment primarily for his pulmonary hemorrhage and renal failure, which resulted in improvement of kidney functions and pulmonary hemorrhage. His arrhythmias completely resolved with 2 sessions of plasmapheresis. No further arrhythmias were seen throughout the remaining duration of his hospital stay. He was started on maintenance steroids and cyclophosphamide.

3. Discussion

Antineutrophil cytoplasmic autoantibody vasculitis like MPA and Wegener’s granulomatosis are potentially fatal diseases secondary to multiorgan involvement. MPA commonly involves the pulmonary, renal and the integumentary system and is noted by ANC A positivity, pulmonary capillary inflammation and crescentic glomerulonephritis. Initial treatment with cyclophosphamide and prednisolone results in disease remission in about 80–90% of patients; however, those patients who present with advanced renal failure or pulmonary hemorrhage have significantly worse outcomes. Thus, in these patients stepwise introduction of more aggressive immunosuppressive induction therapy might be beneficial [2].

MPA often affects the heart in 10 to 20% patients. Cardiovascular involvement in MPA may lead to congestive heart failure in 17.6% patients, pericarditis in 10% and subclinical myocardial infarctions. The myocardium is primarily affected due to vasculitis of coronary arterioles. Cardiac involvement may dramatically worsen the prognosis, particularly for MPA patients. Death can be caused by acute heart failure, arrhythmia or massive myocardial infarction, and can occur during the initial acute phase, or later during the course of the disease, due to refractory residual cardiac failure [3]. Occasionally heart disease can be secondary to uncontrolled hypertension secondary MPA. A case of cardiac tamponade secondary to acute necrotic inflammation by MPA has been reported [4].

While aortic valve insufficiency due to large vessel involvement in MPA without relative endocarditis have also been noted in the literature [5]. MPA without prompt treatment carries a very high mortality, with a 5-year survival of 10% to 20%. Renal failure and acute pulmonary hemorrhage are the leading cause of mortality in such patients. High dose steroids and cyclophosphamide are usually the first line of treatment and can greatly improve the prognosis. Other medications have been used including methotrexate, azathioprine, TNF alpha blockers, rituximab, mycophenolate mofetil, IVIG. Maintenance therapy is required after successful induction, although the optimal duration is not clear [4].

Plasmapheresis has been shown to be effective in removal of circulating antibodies which assists in resolving the acute complications and further deterioration of the patient’s condition. Plasmapheresis facilitates rapid restoration of the renal function without complications. The combination of plasmapheresis with immunosuppressive medication adds much to the recovery of renal function in cases with serious acute renal failure. Plasmapheresis should be considered for all patients with severe alveolar hemorrhage, those with increasing hemoptysis despite conventional immunosuppressive treatment, and those with advanced renal failure (creatinine >5.7 mg/dL and/or the need for hemodialysis) [6].

Our case demonstrates the typical presentation of MPA developing cardiac conduction defects as acute complication of MPA, which was successfully treated with a combination of oral steroids, cyclophosphamide therapy and plasmapheresis. Although the efficacy of plasmapheresis compared to systemic drug therapy is debatable, plasmapheresis provides acute relief in cardiac and pulmonary complications. It also assists in rapid recovery of renal function covering the lag time period the systemically administered corticosteroids and immunosuppressive agents need to be effective, hence plasmapheresis should be considered as an adjunctive therapy for MPA. Plasmapheresis might also be a useful in patients with
MPA who are not responding to standard therapy. This case shows that early diagnosis and interventions like plasmapheresis is crucial in vasculitis associated glomerulonephritis and can reverse cardiac complications like SVT and VT. The primary reason for plasmapheresis in our patient was severe pulmonary hemorrhage and renal failure, though it also resolved cardiac arrhythmias in our patient.

4. Conclusion

Acute cardiac arrhythmias secondary to MPA may prove to be life threatening and lead to increased mortality. Immunosuppressive therapy along with prompt plasmapheresis must be considered in patients with acute renal failure and diffuse alveolar hemorrhage. The fact that plasmapheresis resolved arrhythmias in our patient indicate that plasmapheresis might be a treatment option in vasculitis patients presenting with life threatening cardiac arrhythmias. Our case highlights the rapid therapeutic effects demonstrated by plasmapheresis and emphasizes its utility in providing adequate protection at a critical time.

References