

ORAL PRESENTATIONS - FULL TEXT

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OP-1: Acute Respiratory Failure due to Opportunistic Infection in A Microscopic Polyangiitis Patient under Remission Induction Treatment

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Opportunistic infections are one of the most leading causes of mortality in microscopic polyangiitis (MPA). Patients receiving immunosuppressive therapy should not be ignored for the prophylaxis of pneumocystis jirovecii infection. Herein, we aimed to report a severe opportunistic lung infection in MPA patient on remission induction treatment.

CASE REPORT

A 70 years old female patient with diabetes mellitus and hypertension was admitted to the hospital with dyspnea, high fever, weight loss and oliguria. In her history, she had weakness, loss of appetite, weight loss and she was being followed up without any medication due to interstitial lung disease for 1 year. In admission to the hospital, fever (38.5°C), decreased breath sound in the lower zones of the left lung, coarse rales in the both lungs and systolic murmur on aortic area were seen on her physical examination. In laboratory assesment; leukocytosis (11500/ uL), anemia (hemoglobin: 7.8 g/dl), increased in creatinine (14 mg/dl), and acute phase reactants [CRP (168 mg/L), sedimentation (86 mm/h)] and proteinuria (300 mg / dl) were found. Computerized tomography (CT) of thorax (**Figure 1**) showed bilateral pleural effusion and consolidated areas in the upper lobe of the left lung. In her previous evaluation she had normal creatinine levels and p-ANCA seropositivity of 1 year ago.

In hospital admission of the patient, hemodialysis was started due to oliguric acute renal failure and antibiotic treatment (sulperazone and moxifloxacin) was given for possible pneumonia. In repeated autoantibody tests, p-ANCA and anti MPO were found to positive again and intravenous 1000 mg methylprednisolone pulse (3 days) treatment was initiated with the diagnosis of "MPA". In her follow-up, oliguria improved, creatinine (4 mg/dl) and CRP (13 mg/ l) levels decreased and hemodialysis was discontinued. She was discharged by starting monthly intravenous cyclophosphamide (750 mg) treatment.

After second cycle of cyclophosphamide treatment, she presented to our rheumatology outpatient clinic with progressive dyspnea and fever. Physical examination revealed tachipnea (36/ per min), fever (39°C), hypoxia (SO₂:70%) and bilateral diffuse coarse rales in the both lungs. In her laboratory tests, leukocytosis

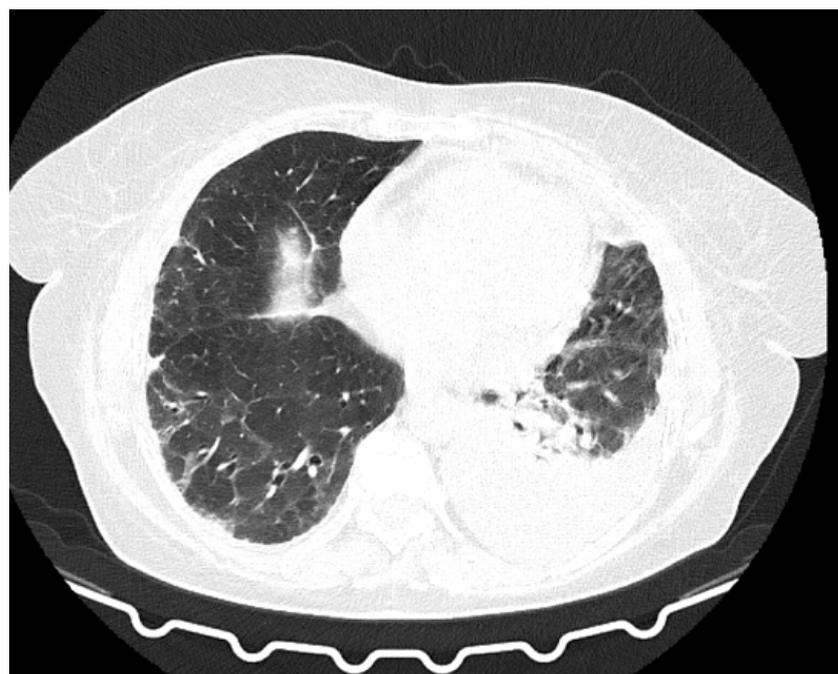


Figure 1: Thorax CT, performed before MPA diagnosis showing bilateral pleural effusion and consolidated areas in the upper lobe of the left lung.

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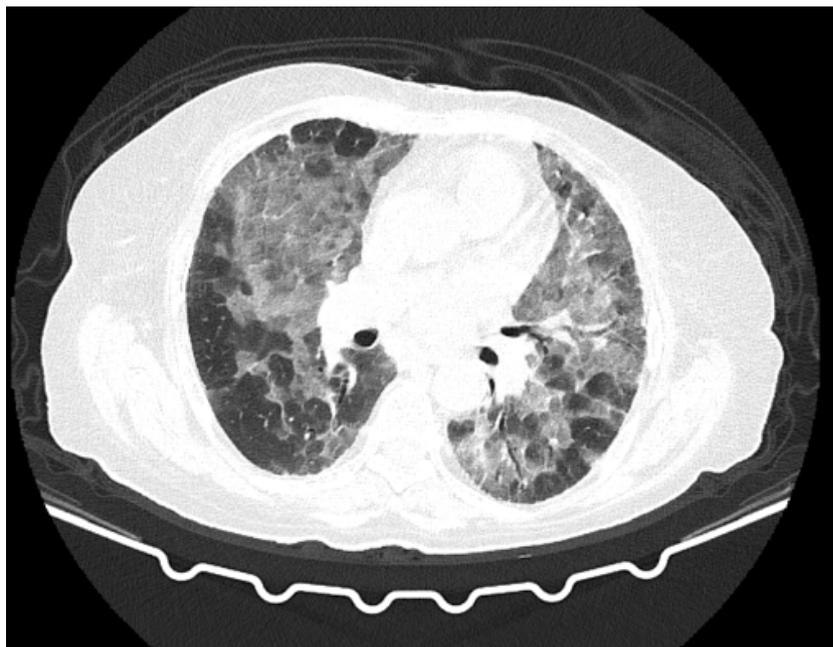


Figure 2: Thorax CT, performed due to acute respiratory failure showing diffuse ground glass and traction bronchiectasis in both lungs.

(15100 /uL), high CRP (185 mg/l) and mildly elevated creatinine (1.3 mg/dl) level were found. Thorax CT showed “diffuse ground glass and traction bronchiectasis in both lungs” (**Figure 2**). She was admitted to the intensive care unit due to rapidly developing respiratory failure, O₂ support was started with non-invasive mechanical ventilation. Bronchoscopic evaluation was done, alveolar hemorrhage excluded and *Pneumocystis Jirovecii* PCR test were found as positive in the bronchoalveolar lavage (BAL) fluid. She was diagnosed as *Pneumocystis pneumonia* (PCP) and Trimethoprim-sulfamethoxazole (TMP-SMX) (320/1600 mg/day-IV) treatment was commenced. Her tachypnea regressed and oxygen requirement (2 lt / min) decreased on the 6th day of the treatment. In her follow-up, regression of consolidation showed on imaging and antibiotic treatment was discontinued on the 21st day.

DISCUSSION

MPA is a necrotising vasculitis on the spectrum of ANCA-associated vasculitis and characterized mainly pulmonary and renal involvement. The main lung manifestation of MPA is diffuse alveolar haemorrhage (DAH) due to pulmonary capillaritis [1]. Lung fibrosis and interstitial abnormalities has been reported in approximately 30-92% of MPA patients. While interstitial lung disease is mostly found at the same time with MPA symptoms, it can occur 1-12 years before the diagnosis of MPA in 10-30% out of patients [2]. In some studies, MPA ranging has been reported as 1.7-25.7% during the follow up of p-ANCA positive idiopathic pulmonary fibrosis patients [3,4]. Management of ANCA-positive interstitial pneumonia cases with immunosuppressive and/or antifibrotic treatments is still controversial.

Differential diagnosis of acute respiratory failure during the treatment in MPA can be difficult. BAL examination is very important for the diagnosis. *Pneumocystis jirovecii* is one of the most important opportunistic infections and increases with the use of high doses glucocorticoids for more than 1 month and prophylaxis is recommended in these cases. TMP-SMX is the most preferred agent in prophylaxis, but as in our case, severe renal failure limits its use [5,6]. Alternative prophylactic treatment regimens (Dapsone, pentamidine) should be preferred in such cases.

KEY MESSAGES

- *It should be kept in mind that ANCA positive interstitial pneumonia may be a precursor of MPA. These cases should be evaluated carefully and multidisciplinary for immunosuppressive and antifibrotic treatments.*
- *In acute respiratory failure that develops during treatment in MPA, the differential diagnosis of disease activity and infections should be made carefully. The importance of prophylaxis in the prevention of opportunistic infections, which is one of the main causes of mortality, should not be forgotten.*

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OP-2: An Intra-Abdominal and Renal Mass in A Patient Followed with The Diagnosis of Granulomatosis Polyangiitis: A Malignancy or Activation of The Disease?

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Granulomatosis polyangiitis (GPA) is an autoimmune vasculitis that is associated with anti-neutrophil cytoplasmic antibodies and affects small and medium-sized arteries and causes internal organ involvement [1]. Clinical manifestations of kidney involvement may range from asymptomatic form to kidney failure which increase the mortality of the disease [2]. Here, we would like to present our case diagnosed with granulomatosis polyangiitis which occurred with bilateral renal masses and which might be confused with renal cell carcinoma (RCC), a rare form of kidney involvement.

CASE REPORT

A 59 years old age male patient applied to our rheumatology outpatient clinic with epistaxis and hemoptysis in October 2016. The patient with cavitating pulmonary nodules in lung computerized tomography (CT) and a positive PR3-ANCA was diagnosed with GPA after performing a biopsy from the mass in the nasopharynx. After achieving remission through the induction treatment with cyclophosphamide, the treatment was continued with azathioprin. Due to the remission after two years therapy, the treatment of the patient was tapered and finally stopped in June 2019. In February 2020 the patient admitted with newly onset cough, hemoptysis, arthralgia, weight loss and elevated acute phase reactants, cyclophosphamide and corticosteroid was administered due to the possibility of the activation of the disease. The patient was hospitalized because of an increase in hemoptysis, the onset of the abdominal pain, a rise in cavitory lung lesions and the continuing elevation of acute phase reactants. There were no significant findings in physical examination. In laboratory assessment the patient had moderate leukocytosis (11.000 / mm³), normocytic anemia (12, 1 g/dl), elevation in ESR (48 mm/hour) and CRP (101 mg/L), and PR3-ANCA level of 162 U/ml. The patient also had isolated hematuria in his urine analysis; with no cast, and proteinuria (160 mg per day). In abdominal ultrasonography, solid formations with a diameter of almost 4 cm were observed at the bottom poles of both kidneys. Several massive lesions contrasted heterogeneously and coherent with renal RCC were spotted at the bottom pole of the left kidney, with a size of 57X48 mm and at the bottom pole of the right kidney with a size of 46 X

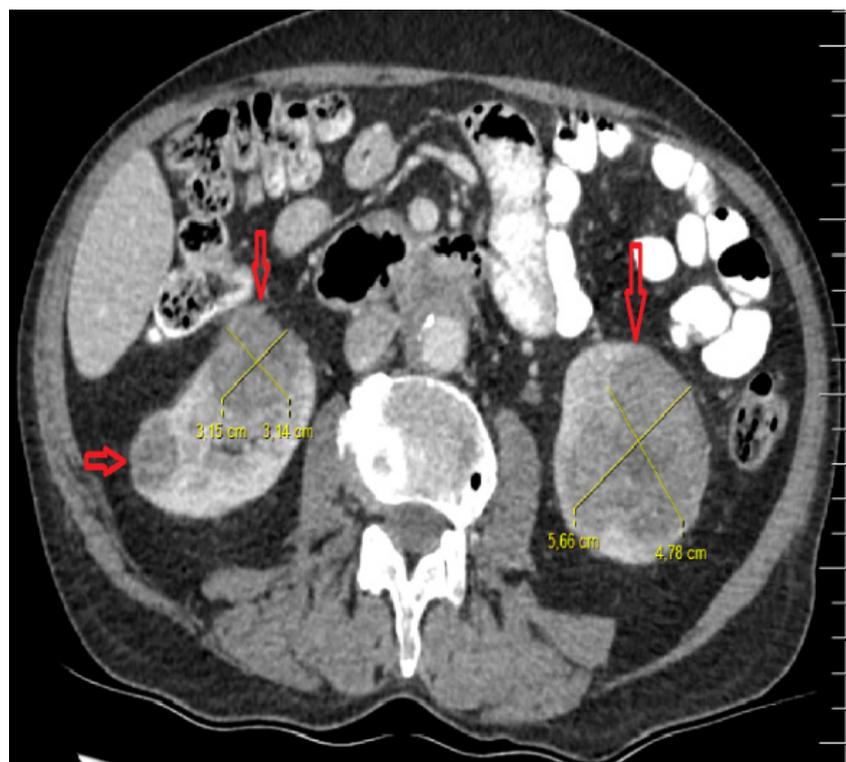


Figure 1: A massive lesion in the left kidney, 57x48 mm and several massive solid lesions with differing sizes in the right kidney.

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Figure 2: A massive solid lesion with a thickness of 18 mm, surrounding abdominal aorta at the infrarenal level.

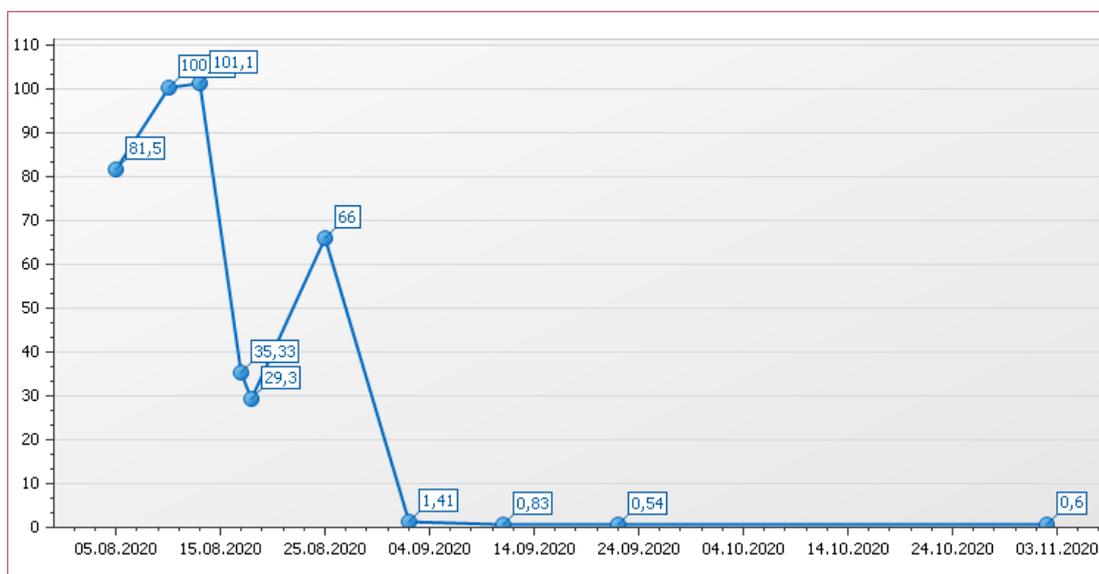


Figure 3: CRP response after pulse steroid and rituximab treatment.

37 mm in the abdominal computerized tomography (**Figure 1**). Also, massive lesions with a thickness of up to 18 mm and surrounding the abdominal aorta at the infra-renal level which were assessed in favor of metastasis detected (**Figure 2**). The biopsy findings of the renal mass were compatible with GPA. After intravenous pulse steroid and rituximab administration, clinical and laboratory remission was achieved (**Figure 3**).

DISCUSSION

The presentation of renal involvement in GPA with a kidney mass is a very rare case. There is only a few case report in the literature about GPA accompanied with bilateral renal multiple mass [3]. Radiological features of renal inflammatory pseudotumor can mimic renal carcinoma or lymphomas and surgical approach, such as nephrectomy, may perform mistakenly. Also, in our case the radiological images of renal masses were reported as RCC. The massive lesions in periaortic and inguinal regions were evaluated as metastasis. However, the biopsy revealed granulomatosis inflammation and vasculitis. These renal masses were determined to be related to active disease of granulomatosis polyangiitis with high acute phase levels and high Birmingham Vasculitis Activity Score (BVAS/WG). The masses in the kidney may have been emerged by the patient’s not using the maintenance treatment for 1 year. On the other hand, the data supporting the association between the development of RCC and GPA and

cyclophosphamide administration led us to be suspected from malignancy in the patient applied with renal mass. If the masses cannot be distinguished from RCC radiologically, and the malignancy can be excluded by pathological examination, GPA should be considered in patients with the other clinical characteristics of GPA.

KEY MESSAGE

- *Though, the appearance of GPA with renal masses is rare, it should be kept in mind. As in the current case, disease flare of GPA should be thought in a patient with abdominal and renal masses, beside solid.*

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OP-3: If A Patient of Granulomatosis with Polyangiitis (GPA) Gets Pregnant under Rituximab Treatment

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Anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitides are wide spectrum of diseases that can lead to life-threatening multi-organ involvement. Drugs used in the treatment of the disease, especially in individuals of reproductive age, may have harmful effects on the fetus. Herein, we present a case of granulomatous polyangiitis (GPA) who became pregnant while under rituximab treatment and developed preeclampsia in the follow-up.

CASE REPORT

A 26-year-old female patient had admitted to another hospital with an enlarging mass in her left eye in 1999. She was followed up with a retrobulbar mass detected on orbital magnetic resonance imaging (MRI) (compatible with the pseudotumor orbita). As a result of biopsy taken from the mass in 2005, she was diagnosed with GPA. The biopsy was reported as “damage in the vessel wall and fibrinoid necrosis in granulomas observed around the vessels”. In 2006, she admitted to Hacettepe University Faculty of Medicine, Department of Pediatric Nephrology, with ulcerous wounds on her body, shortness of breath, and proteinuria in the urine (404 mg/day). She was taking prednisolone 1mg/kg/day and methotrexate 15mg/week on admission. The kidney biopsy of the patient taken at our hospital was reported as necrotizing glomerulonephritis. The ANCA profile was negative, Birmingham Vasculitis Activity Score-Wegener Granulomatosis (BVAS-WG): 3, Five Factor Score (FFS): 0, Vasculitis Damage Index (VDI): 2. Medical treatment of the patient was revised as prednisolone and 2x50 mg oral cyclophosphamide during two years.

In the control evaluation; the mass was similar size in control orbital MRI and proteinuria was in regression (126 mg/day). The disease was accepted in remission and cyclophosphamide treatment was discontinued (BVAS-WG: 1). 13 years after the diagnosis, in 2012, she admitted with shortness of breath and hoarseness under the maintenance treatment with 2x50 mg azathioprine. In physical examination, subglottic stenosis was detected and paranasal sinus computed tomography (CT) showed inflammatory changes compatible with chronic sinusitis. In addition, urinary proteinuria was increased (639 mg/

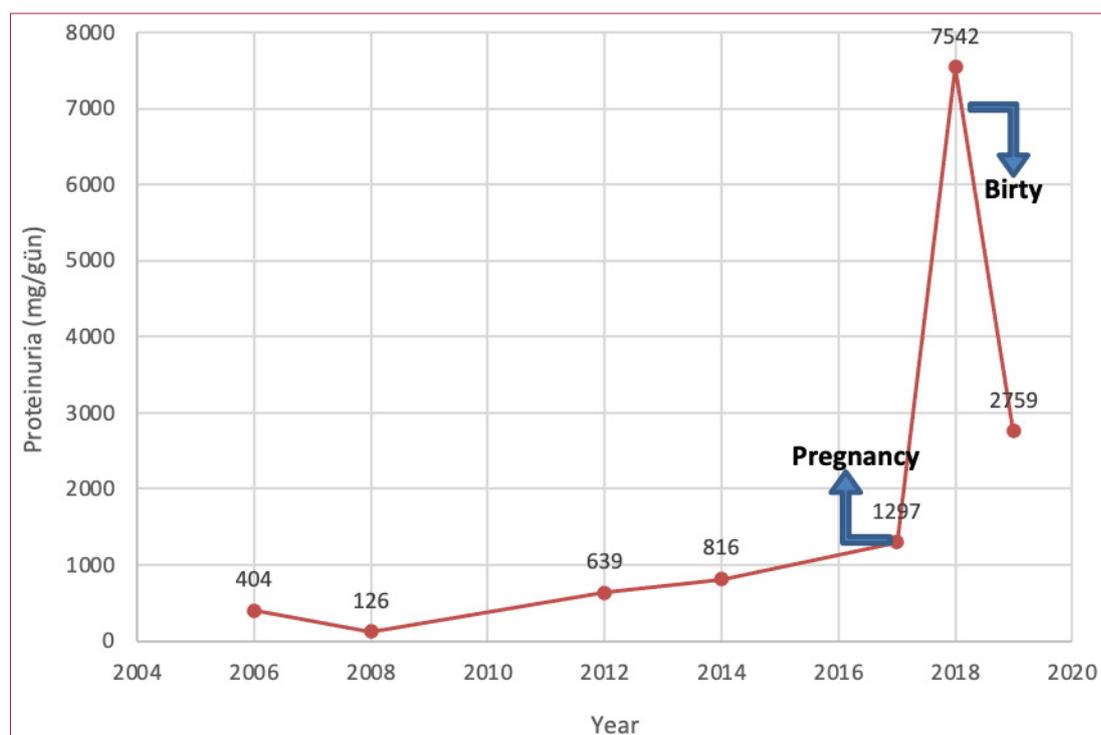


Figure 1: Follow-up values of proteinuria

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day) (BVAS-WG: 3). Induction therapy was restarted with rituximab due to disease flare. In 2012, laryngotracheal dilatation was applied to the patient for subglottic stenosis. In the evaluation performed after 4 gr RTX treatment, although proteinuria continued (816 mg / day), it was interpreted as a sequelae. Also, there were not any significant changes on control orbita MRI. Therefore, disease was accepted in remission and RTX was discontinued (BVAS-WG: 3). She was followed up with enalapril 5 mg/day, prednisolone 5 mg/day and 150 mg azathiopurine.

In 2017, when the patient was under these treatments, urinary proteinuria increased from 582 mg/day to 1297 mg/day, the patient was accepted as GPA disease exacerbation, and RTX induction therapy was started again (BWAS-WG: 6). Although she was told that she should comply with the contraception rules during the treatment process, she admitted to our clinic with 6 week pregnancy after she received the first 1000 mg RTX treatment. Rituximab induction treatment was discontinued and she was informed about the progression risks of the current disease and the possible risks it may pose to the fetus. However, the patient said that she wanted to continue her pregnancy. She was followed up with 20 mg/day prednisolone until the third trimester. She was hospitalized with suspect of GPA disease flare or preeclampsia due to the increase in proteinuria (7542 mg/day) and high blood pressure in the outpatient clinic control at 28 weeks of pregnancy. In the evaluation; GPA exacerbation was not considered initially due to the absence of new or worsening major and minor signs and stable BVAS/WG score (BVAS/WG: 3). Obstetrics and gynecology department evaluated the patient as preeclampsia due to hypertension and increase in proteinuria. Cesarean section was performed at the 32nd week because of blood pressure could not be controlled despite anti-hypertensive treatments. A healthy girl weighing 1980 grams was born. Congenital malformation or neonatal infection was not observed. In the postpartum period, blood pressure values returned to normal and proteinuria regressed. The patient was discharged with 20 mg/day prednisolone and 20 mg/day enalapril treatment.

DISCUSSION

In the treatment of GPA, rituximab or cyclophosphamide treatment is recommended in combination with glucocorticoids in life-threatening situations and severe organ involvement [1]. Rituximab treatment given during pregnancy passes through the placenta and causes B cell depletion and hypogammaglobulinemia in the fetus [2]. Although there is no strong evidence for its safety in pregnancy, in the 2020 American College of Rheumatology guideline for reproductive health, it is stated that rituximab can be given during pregnancy only in life-threatening organ damage [3].

It is difficult to make the differential diagnosis of preeclampsia and GPA exacerbation. Complete blood count, kidney function tests, liver function tests, ANCA titer, sedimentation, C-reactive protein (CRP) can be used. Unfortunately, none of these tests are specific. An increase in proteinuria is seen in both GPA exacerbation and preeclampsia. Auzary et al. reported that the presence of hypertension may guide the differentiation of preeclampsia from GPA activation in a series of 5 cases [4]. In our patient, preeclampsia was considered initially because BVAS-WG score did not increase, ANCA profile was negative, blood pressure was high, and sedimentation and CRP levels were normal. Postpartum blood pressure values returned to normal and proteinuria decreased.

KEY MESSAGES

- *Effective contraception should be provided definitely for patients with rheumatic diseases in reproductive age. When pregnancy develops in individuals under active treatment, the risks that may occurs in the fetus and in the patient should be evaluated together with the patient and individualized decision should be made.*
- *Disease activity scores can be used in the differential diagnosis of preeclampsia and disease exacerbation during the course of pregnancy.*

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OP-4: Severe Covid-19 Pneumonia in A Granulomatosis Polyangiitis Patient under Rituximab Treatment: Difficult Aspects of Immunosuppression in the Pandemic

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Systemic involvement of granulomatosis with polyangiitis (GPA) can be presented in a wide spectrum and the degree and variety of organ involvement can be comprehensively evaluated in the aspects of diagnosis, treatment and prognosis by disease specific activity scores. During the Covid-19 pandemic, granulomatosis polyangiitis patients are facing with an important infection risk as in many patients with other rheumatological diseases. Herein we present a case of GPA with pulmonary, ophthalmologic and nasopharyngeal involvement, who was diagnosed with Covid-19 pneumonia despite the absence of additional comorbidity and immunosuppressive therapy being titrated according to disease activity scores.

CASE REPORT

A 57-year-old male admitted to our clinic due to bloody nasal discharge for months. Progressive droopy eyelids; redness, discharge, and pain in both eyes; intermittent hemoptysis and constitutional symptoms such as fatigue, night sweats, involuntary weight loss (approximately 6 kg in 3 months) were also present. Physical examination revealed scleral hyperemesis, nasal septum ulcers and rales in bilateral upper pulmonary zones. Neutrophilia ($6.5 \times 10^3 / \mu\text{L}$) and thrombocytosis ($411 \times 10^3 / \mu\text{L}$), increase in ESR (81 mm / h) and CRP (7.6 mg / dL) and p-ANCA positivity (IFA 1/320) were present in the laboratory examination. Creatinine and glomerular filtration values were in the normal range. He had stage 1 proteinuria (373.15 mg / g creatinine) in spot urine and had nodular opacities on chest radiography.

Orbital magnetic resonance imaging (MRI) revealed that both lacrimal glands had mild size increase, hyperintensity and heterogeneity. Computed tomography (CT) of thorax showed bilateral multiple nodular lesions of which the largest was located in the right upper lobe and had sizes of 17x15 mm. Nasal septum biopsy revealed signs of active and chronic inflammation in small vessels. Patient's BVAS score was 11 and the BVAS-WG score was 6.

The patient was started on rituximab (1000 mg every 15 days) and prednisolone (60 mg) with diagnosis of activated GPA and was discharged by decreasing prednisolone dose (36 mg). In his follow-up, BVAS and BVAS-WG scores decreased and the rituximab dose scheme was arranged to be 500 mg every 4 months at the last outpatient visit.

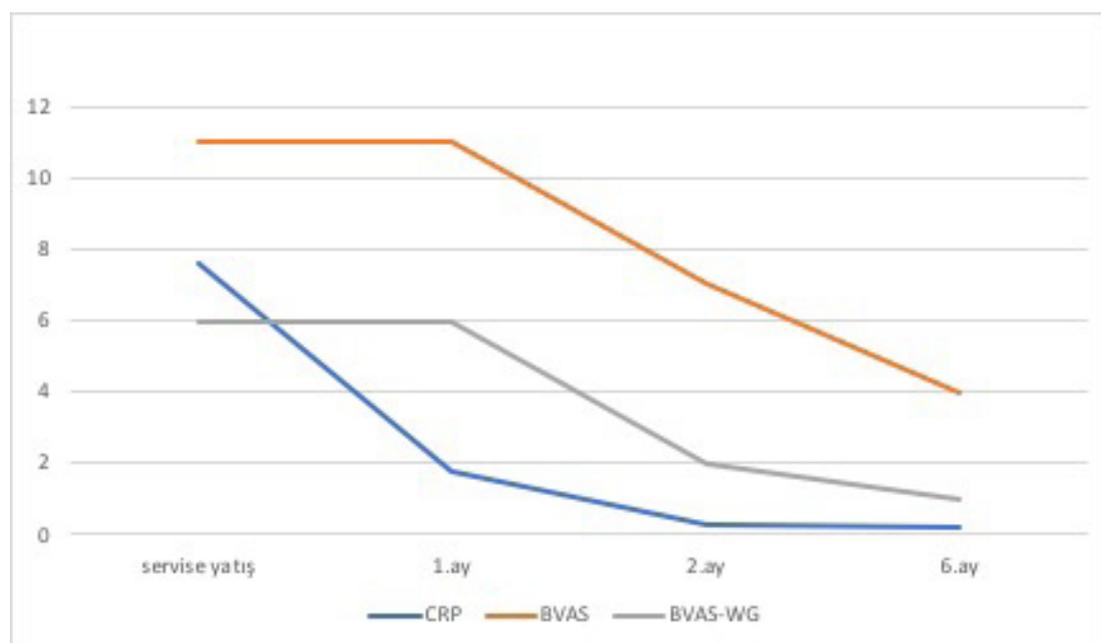


Figure 1: Disease activity score and CRP course

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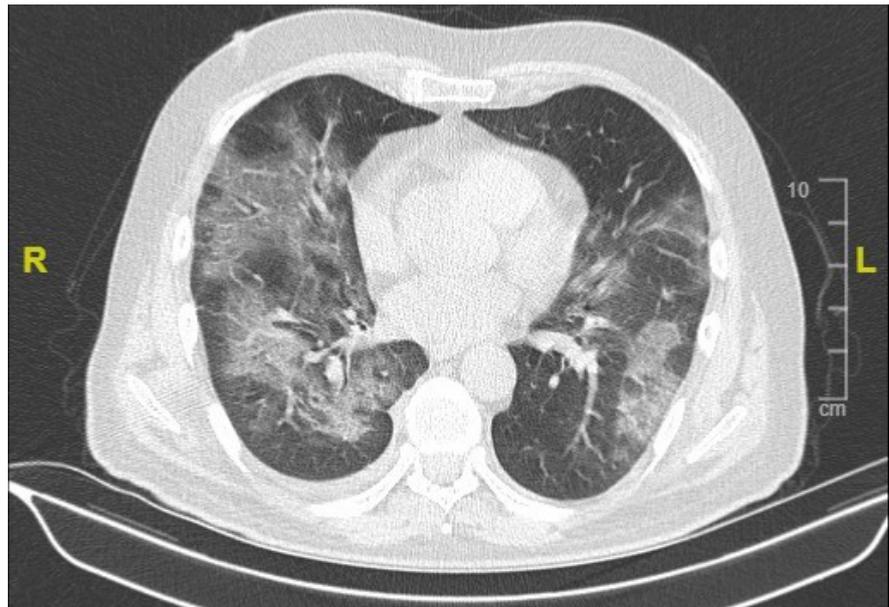


Figure 2: Bilateral ground-glass opacities consistent with Covid-19

One week after the last rituximab treatment, patient's complaints of weakness and widespread muscle pain started, and his Covid-19 nasopharyngeal PCR swab sample revealed as positive. He was started on favipiravir treatment and isolation was recommended. Patient developed shortness of breath during the follow-up and applied to emergency department. Patient was started nasal oxygen therapy and due to the detection of bilateral ground glass densities compatible with severe Covid-19 pneumonia in his thorax CT under rituximab and prednisolone (5 mg) treatment, was decided to be followed in the intensive care unit under dexamethasone therapy, in where he was needed short-term non-invasive mechanical ventilation. After his symptoms decreased and need for oxygen therapy waned with the negative result of Covid-19 nasopharyngeal PCR swap at the 10th day, he was discharged with his remission steroid therapy.

DISCUSSION

During the Covid-19 pandemic, epidemiological studies have been carried out in various centers on the frequency of Covid-19 infection in rheumatological diseases and the effect of immunosuppressive treatments on infection outcomes [1]. While Gianfrancesco et al. stated that as the steroid treatment dose increased, hospitalizations secondary to Covid-19 infection increased, they also reported that hospitalizations were seen at a lower rate in patients using biological disease modifying agents [1]. However, Guilpain et al. reported a case of Granulomatosis with Polyangiitis (GPA) complicated by Covid-19 pneumonia requiring mechanical ventilation under rituximab therapy [2]. Similar to eosinophilic Granulomatosis with Polyangiitis (GPA) cases [3], the clinical and radiological confusion of the pulmonary involvement of the disease with Covid-19 pneumonia and delay in the diagnosis of infection constitute another problem encountered in granulomatosis polyangiitis during the current pandemic.

The BVAS and BVAS-WG scores are prognostically validated scoring systems in patients with ANCA-associated vasculitis and granulomatosis polyangiitis, respectively. These scores play a critical role in making the right amount of immunosuppressive treatment decision for the right patient for rheumatologists who need to keep the risk of Covid-19 infection to a minimum while simultaneously trying to control the underlying rheumatological disease. As seen in our case, encountering severe Covid-19 pneumonia even in patients who do not have additional comorbidities and whose immunosuppressive treatment is appropriately followed by disease scoring systems, emphasizes the importance of this issue once again.

KEY MESSAGES

- *In GPA patients, immunosuppression status and COVID-prevention recommendations may differ depending on the immunosuppressive treatment regimen; groups under rituximab and cyclophosphamide treatment have the highest risk.*
- *Although our patient was under RTX treatment and developed COVID-19 pneumonia, successful results were obtained with early and effective supportive treatment.*

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OP-5: Treatment Resistant Subglottic Stenosis in Granulomatosis with Polyangiitis: Case Report

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The frequency of subglottic stenosis (SGS) in granulomatosis with polyangiitis (GPA) is between 16-23% [1]. Since SGS can occur at any time regardless of GPA disease activity and is an important cause of morbidity and mortality, early diagnosis and treatment of SGS is important [2]. Here, we present a case of GPA with SGS that occurred during the course of the disease and required both immunosuppressive and subglottic balloon dilatation treatments.

CASE REPORT

A 44-year-old female was diagnosed with GPA in 2016 due to otalgia, rhinorrhea, hemoptysis, hematuria, dyspnea, 410 grams of proteinuria in 24-hours urine, cytoplasmic antineutrophil-cytoplasmic-antibody positivity (c-ANCA), cavitory lesions on thorax computed tomography, necrotizing granulomatous vasculitis findings on nasal mucosa biopsy. Cyclophosphamide (CYC) and high dose corticosteroid (CS) were started as induction therapy. In the 6th month of treatment, CYC (total of 6.5 grams) was discontinued and rituximab (two-1000 mg IV infusions separated by 2 weeks (one course) every 6 months) was started due to persistent otalgia, dyspnea and wheezing. Three months later, patient's previous pulmonary findings improved significantly in control thorax CT but there was no decrease in dyspnea. On physical examination, there was prolonged inspiration and stridor. She was consulted to the Otorhinolaryngology clinic, video laryngoscopy revealed a subglottic stenosis (**Figure 1**) and balloon dilatation was performed. The patient's dyspnea and stridor improved. However, dyspnea and stridor recurred 3 months after dilatation. Balloon dilatation was performed three times in two years due to recurrent subglottic stenosis. On the other hand, rituximab and low dose CS treatment was continued. In 2020, the cavitory lesions disappeared in thoracic CT, however, soft tissue thickening in the subglottic area on neck CT continued, although it regressed compared to previous imaging. The patient's subglottic stenosis was considered a sequela, and rituximab treatment (8 grams in total) was discontinued. Although balloon dilatation or tracheoplasty or tracheostomy was recommended by otolaryngologists, the patient did not accept this.

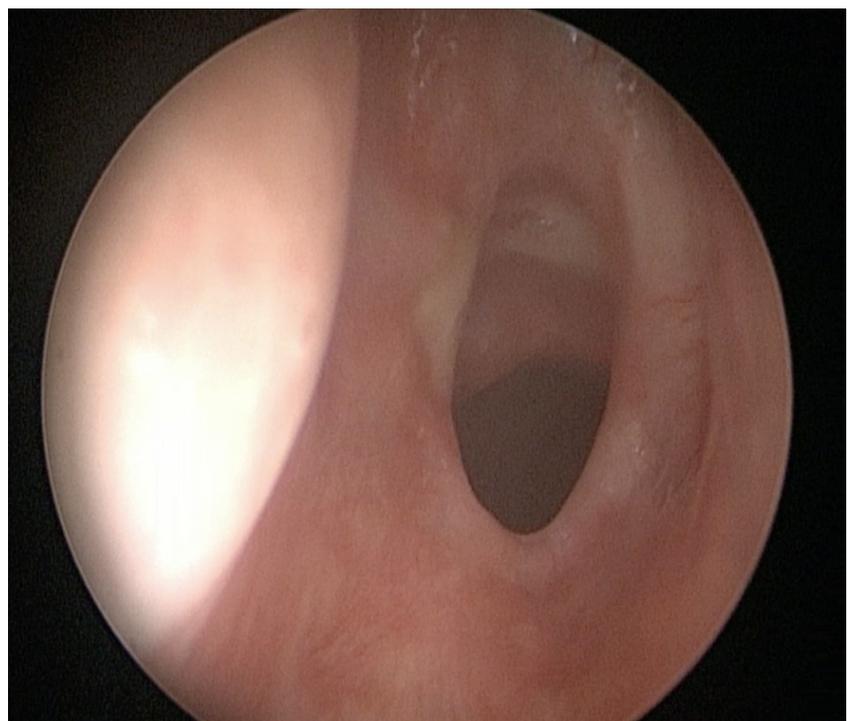


Figure 1: View of subglottic stenosis in video laryngoscopy

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DISCUSSION

Subglottic laryngotracheal stenosis is a relatively rare but important complication of GPA, which usually requires immunosuppressive therapy, surgical intervention, or both. As a surgical intervention, balloon dilation, a therapeutic endoscopic procedure, has been popular in recent years. Relaps of SGS is common despite immunosuppressive and surgical treatment. Since SGS or SGS recurrence may not be associated with active systemic vasculitis, patients should be regularly reviewed in the clinic for symptoms such as increased dyspnea, decreased exercise tolerance, and stridor [3]. Decisions regarding immunosuppression, local surgery, or a combination of these in the treatment of SGS should be made on a patient's basis. When managing patients with SGS or other life-threatening GPA involvement, a holistic and multidisciplinary approach should always be applied, and the best solutions should continue to be sought in collaboration with relevant departments.

KEY MESSAGES

- *Optimal treatment of SGS in GPA patients requires a multidisciplinary approach that includes both rheumatologist and otolaryngologists.*
- *Although various treatments have been tried for SGS in GPA, there is no standardized treatment method (medical, surgical or both), so prospective studies are needed for the optimal treatment of SGS.*

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OP-6: Eosinophilic Granulomatosis with Polyangiitis: Three Cases with Cardiac Involvement

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Eosinophilic granulomatosis with polyangiitis (EGPA) is an ANCA-associated systemic vasculitis characterised by eosinophil-rich and necrotizing granulomatosis inflammation. This necrotizing vasculitis predominantly affects small-to-medium vessels and is associated with asthma and eosinophilia. EGPA often involves the ear, nose, throat (ENT) and respiratory tract. In particular, cardiomyopathy due to eosinophilic myocarditis is not uncommon in EGPA and can be life threatening. Here, we describe the clinical and serological manifestations of cardiac involvement of 3 EGPA patients from a single centre.

CASE REPORTS

Case 1. A 44-year-old-man with a history of late onset asthma, nasal polyps and intermittent chest pain was referred our vasculitis center. On admission, his laboratory tests revealed eosinophilia and negative ANCA. His echocardiography was normal, but his cardiac MRG showed subendocardial patchy contrast enhancement and myocardial linear areas on the inferolateral wall (**Figure 1**). Pulse-corticosteroids and cyclophosphamide treatment were given for induction therapy and then switched to methotrexate maintenance. The patient has been following for 4 years in stable condition.

Case 2. A 40-year-old male admitted with chest pain, tingling and numbness in hands and feet. The patient had been followed in another center with frequent asthma attacks and eosinophilia for 2 years. The patient was hospitalized, a negative ANCA, a raised troponin and ischaemic changes on ECG and dilate cardiomyopathy on echocardiography was found. Electromyography showed severe distal sensorimotor axonal polyneuropathy. He was diagnosed with EGPA with respiratory, cardiac, neurologic involvement and eosinophilia. Pulse-corticosteroids and cyclophosphamide treatment was started to the patient, but there was no improvement cardiac status. During the follow-up bi-ventricular ICD was inserted, but after 1.5 years of cardiac involvement, the patient died by sudden cardiac arrest.

Case 3. A 33-year-old-man previously diagnosed with EGPA for six-years admitted with chest pain and dyspnea. ENT and respiratory involvements were the initial presentations with negative ANCA serology and the patient was treated with methotrexate. On admission, a widespread T wave and ST segment changes were found on electrocardiography. There were global hypokinesia and dilated cardiomyopathy on echocardiography and cardiac fibrosis on MRI. Treatment with pulse-corticosteroids and cyclophosphamide was started immediately and patient followed-up as a stable disease with heart failure for three years.

DISCUSSION

The present cases highlight the possibility of heart involvement in EGPA at disease onset or during the course of disease. Cardiac involvement is seen up to 60% of EGPA patients and may occur with different clinical presentations such as chest pain, myocardial infarction or heart failure [1]. Even in asymptomatic EGPA patients subclinical myocardial involvement has been shown by using either echocardiography or MRI [2].

Of three types of AAV, cardiac involvement is the most common in EGPA, especially in ANCA – subsets [3]. Cardiac involvement, which is also used in the Five Factor Score, is common and poor prognostic factor in EGPA patients [4]. Therefore, risk stratification using cardiac imaging may be rational in EGPA patients.

KEY MESSAGES

- Cardiac involvement is frequently seen in EGPA patients and cardiomyopathy is one of the most important predictors of a negative outcome.
- Cardiac imaging should be done for early diagnosis of cardiac involvement in EGPA patients, even for asymptomatic cases.

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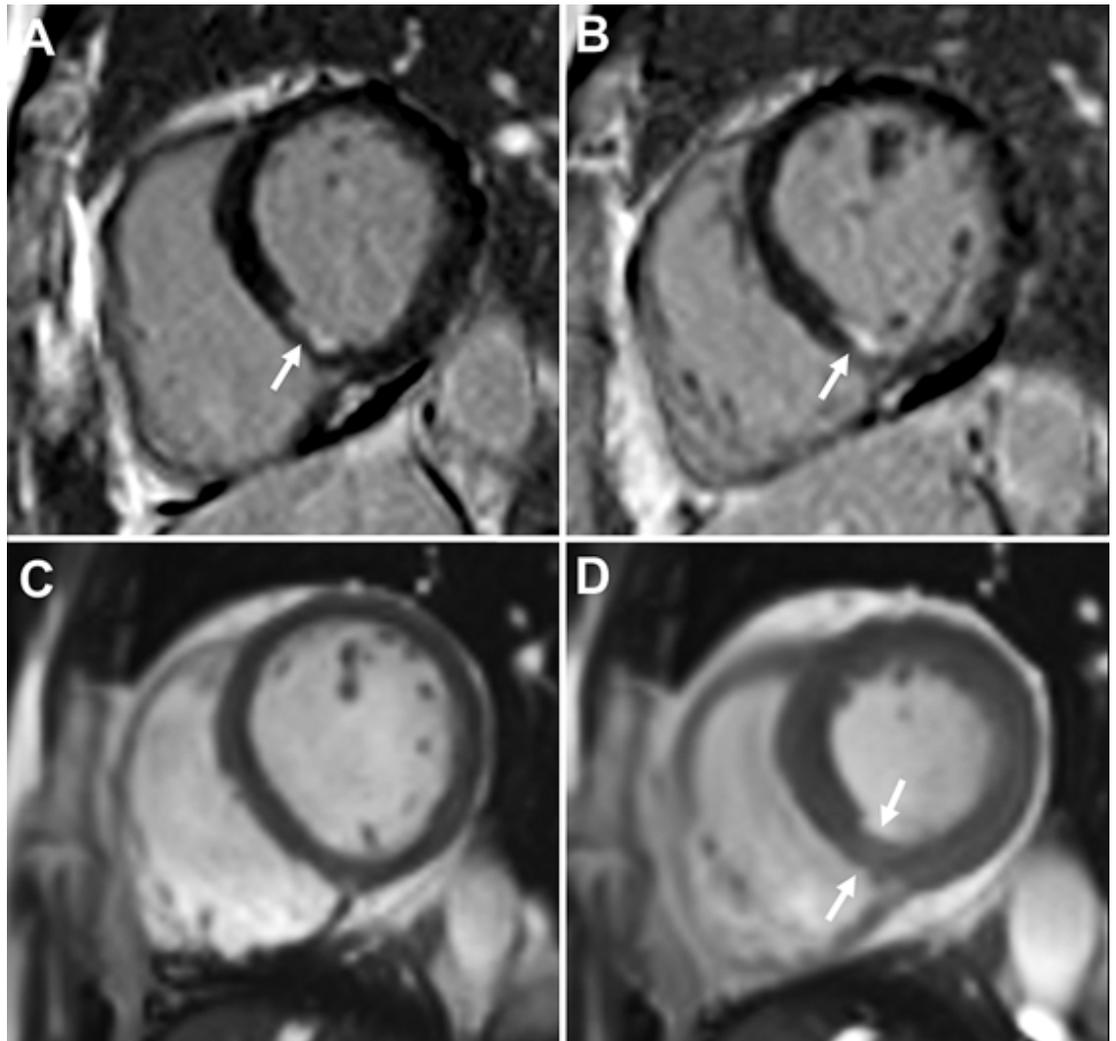


Figure 1: 48 years-old male patient with EGPA. A, B : Subendocardial contrast enhancement in the phase-sensitive inversion recovery (PSIR) sequences of inferoseptal wall in the sections through midventricular and short axis at late-contrast phase. C, D: Inferoseptal hypokinesia (D, arrows) at short axis end-diastolic sine SSFP (C) and end-systolic (D).

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