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Case Report

Granulomatosis with Polyangiitis that Presented wıth Hearing Loss: A Case Report

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Abstract
Wegener's Granulomatosis is an ANCA-associated necrotizing vasculitis characterized by moderate-sized vascular involvement and often affects renal and pulmonary functions. The disease can appear in systemic and localized forms. The patients usually apply with weakness, fever, weight loss, artrhalgia, rhinosinusitis, cough and dyspnea, urinary abnormalities with or without renal failure, neurological abnormalities, these symptoms can all be seen in ANCA associated vasculitis. Herein, we report a case of Wegener's granulomatosis presented with hearing loss and pulmonary syndrome, diagnosed by renal biopsy and whose condition improved after initial treatment.

Keywords
Wegener's Granulomatosis, Hearing Loss

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Introduction

Wegener's Granulomatosis is an ANCA-associated necrotizing vasculitis characterized by moderate-sized vascular involvement and often affects renal and pulmonary functions. C-ANCA is usually positive in these patients. In 2011, the name of Wegener's Granulomatosis was changed to Granulomatosis with Polyangiitis (GPA) by the American College of Rheumatism (ACR), the American Society of Nephrology (ASN) and the European Leauge Against Rheumatism (EULAR). The disease can appear in systemic and localized forms. Localized form is a little more frequent in female and younger patients, usually remains limited in the upper respiratory tract [1]. Although patients that are diagnosed with systemic GPA are usually older patients, the disease can be seen in all age groups [2]. Gender- based distribution is similar in both male and female patients; the disease is seen more in the white race [3].

Patients usually apply with weakness, fever, weight loss, arthralgia, rhinosinusitis, cough and dyspnea, urinary abnormalities with or without renal failure, neurological abnormalities, and these symptoms can be seen all ANCA associated vasculitis [2]. Ear, nose, throat related symptoms can all be seen in ANCA associated vasculitis, these symptoms are reported in GPA more frequently [2]. The hearing loss can be seen in patients with affected ear, nose, throat and these situations can be temporary or permanent [4].

In this case, we present a patient who has Wegener's Granulomatosis who applied with hearing loss and pulmonary-renal syndrome.

Case Report

A 48-year-old female patient without any known disease underwent tympanostomy because of sudden onset hearing loss twice in another medical center approximately 2 months ago. The reason of hearing loss was not found. After that, the symptoms such as cough, hemoptysis started, antibiotherapy was initiated with a preliminary diagnosis of pneumonia. Because mass lesions were detected in thorax CT, PET CT was performed; pathological lesions were detected in nasopharynx, lungs, bilateral hilar regions, spleen and bone marrow. During the follow-up of the patient, after the appearance of anemia and renal failure, the patient, anticipating the need for intensive care, was transferred to our medical center. Blood test values upon the arrival of the patient were determined as follows: urea: 139mg/dl, creatinine: 9.4mg/dl, wbc: 18000uL, Hb: 7.9gr/dl, pH: 7.38, HCO3: 21.1mmol/L. Hemodialysis is planned. The patient had urine output; postrenal acute renal failure was excluded with urinary USG. Because of procalcitonin and CRP elevation, infection in urinalysis test and fever, meropenem therapy was initiated. In the situation of this patient, vasculitis with pulmonary-renal syndrome was considered. Although the fast antigen tests were negative, the Quantiferon test was performed for tuberculosis. ARB test was planned for 3 consecutive days in sputum. The tests results were negative. Owing to preliminary diagnoses of vasculitis, C-ANCA, P-ANCA, ANA, and other serological tests were performed. The pulse steroid therapy (3 days, 1gr/day methylprednisolone) was planned but because of the elevated infectious parameters, the pulse steroid therapy could not be given, plasmapheresis was performed for 7 consecutive days.

The infectious parameters and urine leukocyte count regressed under the antibiotherapy, the kidney biopsy was performed.

PET CT performed in the other center was interpreted again. Pathological hypermetabolic lesions were detected in nasopharynx, bilateral lungs, bilateral hilar regions, and spleen, common diffusely lesions in bone marrow and bilateral kidney lesions were detected. These lesions suggest Granulomatosis with Polyangiitis (GPA).

The diagnosis of vasculitis was detected by a kidney biopsy, C-ANCA test results were positive, tuberculosis was excluded. The therapy with methylprednisolone (1gr/day, 3 days), after that methylprednisolone 1x60mg maintenance therapy, IVIG 1x0.4gr/kg for 5 days, after that cyclophosphamide 500mg and MESNA 400mg once a week, the therapy was planned 3 times to the patient. In the follow up of the patient, intermittent hemodialysis was performed. The need for high flow mechanical ventilation and hemodialysis were regressed. The patient's general situation improved dramatically. Patient's hearing loss was documented with audiometric tests and regressed after therapy.

Discussion

Granulomatosis with Polyangiitis (GPA) frequently presents with urinary and pulmonary symptoms. Ear, nose and throat lesions can be seen more commonly in this type of ANCA associated vasculitis.

Depending on ACR, nasal and oral inflammation, radiographically abnormal thorax findings, abnormal urinary sedimentation and showing arterial and perivascular inflammation with biopsy were determined criteria of GPA. The presence of 2 or more of these findings has a %88 of sensitivity, %92 of specificity [5]. The lung and kidney biopsy samples of GPA patients have geographical necrotizing vasculitis. In %50 of biopsies taken from patients who have a limited disease in the upper respiratory tract, chronic inflammation findings are observed. In %15 of biopsies taken from the upper respiratory tract, granulomatous inflammation, vasculitis, and necrosis are detected [1]. In these patients, early diagnosis and treatment are very important in terms of prognosis [6]. Our patient applied to another center with sudden onset hearing loss, then hemoptysis, cough, and renal failure occurred. In patients with hemoptysis, tuberculosis and malignant disease must be considered in the differential diagnosis. In our patient tuberculosis is excluded with the help of ARB tests, Quantiferon test, and fast antigen tests, the pathological diagnoses determined as vasculitis. In the patients that applied with pulmorenal syndrome, the diagnosis of vasculitis must be considered, it should be remembered that complications such as renal failure, hearing loss can be permanent.

This case underlines that in patients applying with hearing loss, renal and pulmonary findings the diagnosis of vasculitis must be considered in the differential diagnosis. It also emphasizes the importance of early treatment for avoiding high mortality that may occur, reducing complications such as pulmonary, renal, and otolaryngological failure that may be permanent.

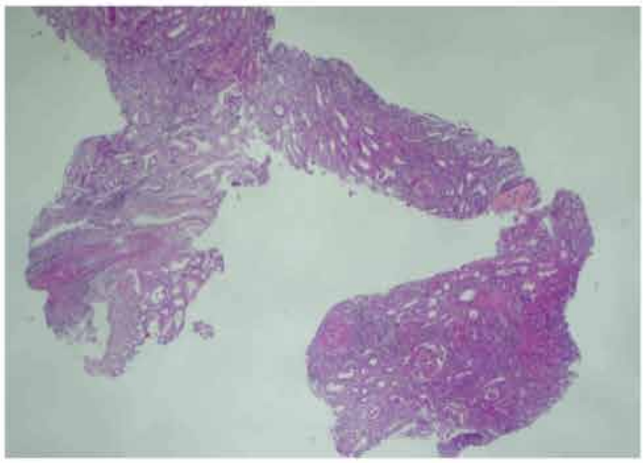


Figure 1. Intense inflammation of the interstitial area, glomeruli and 1 vascular structure were noted in the sections. (H&Ex40).

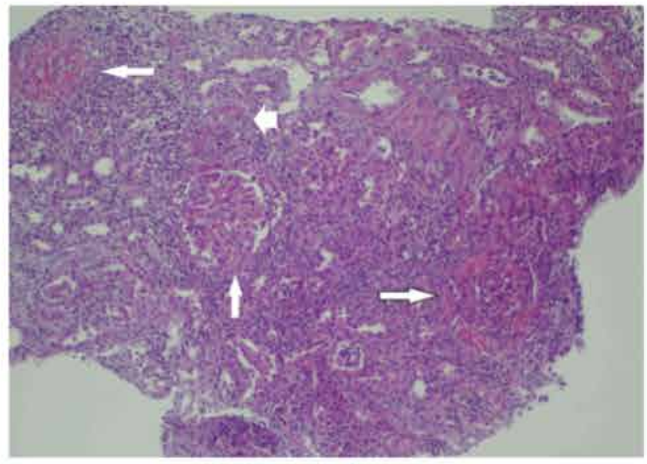


Figure 2. Fibrinoid necrosis is observed in the glomeruli (long white arrow) and arteriole wall (short white arrow).(H&Ex100)

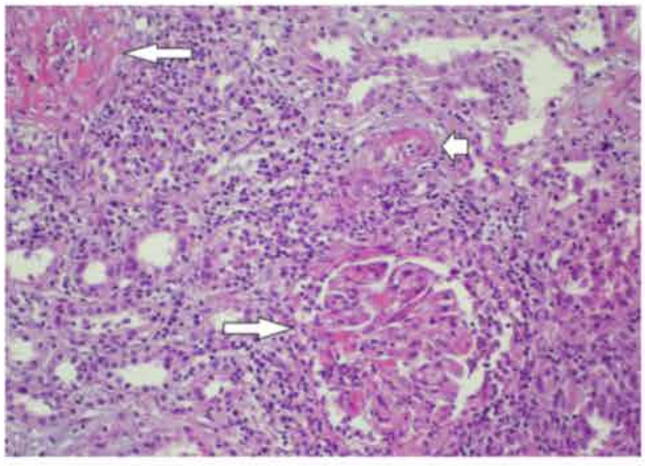


Figure 3. Figure 2x200 (H&E)

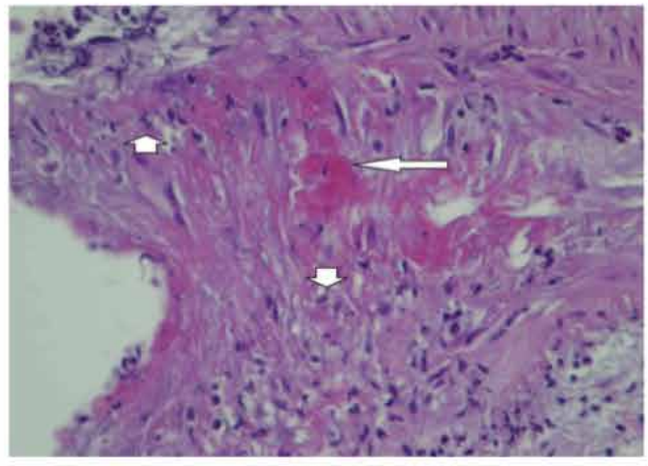
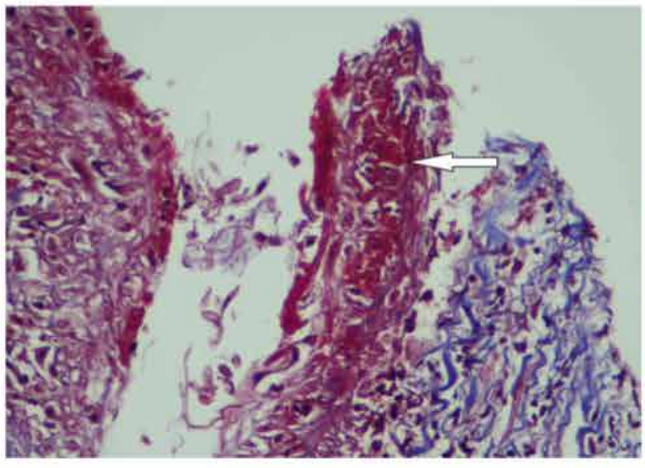


Figure 4. Fibrin (long arrow) deposition in vascular wall and inflammatory cells were noted.(H&Ex400)



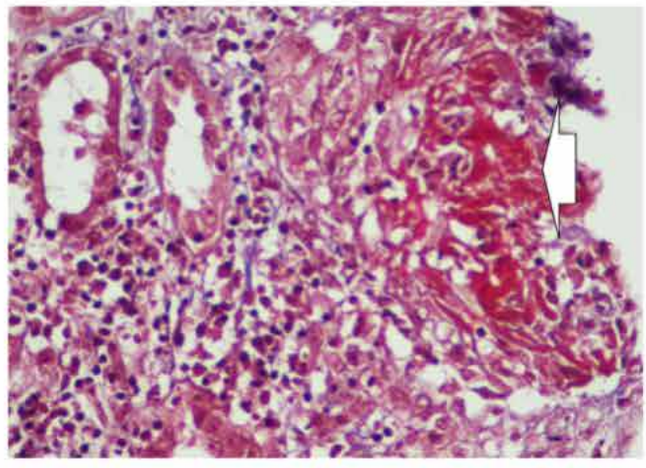
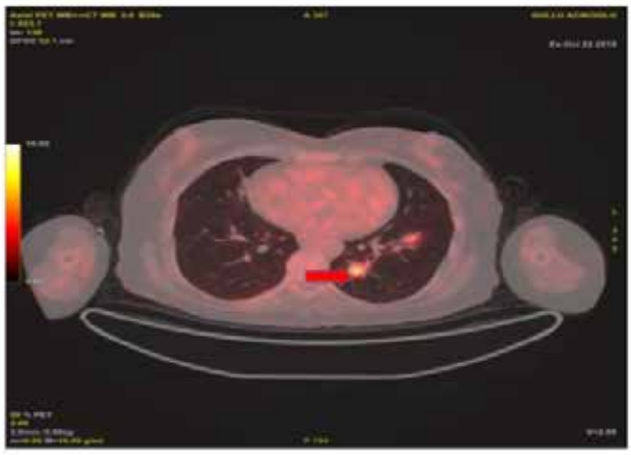


Figure 5. In fibrin deposits in the vessel wall (EN) (long arrow) and glomerular ball (B) (short arrow) fuchsinophilic and red staining with trichrome dye was noted.(MTKx400).



Firuge 6. Increased nodular hypermetabolic activity is observed in the left lung.

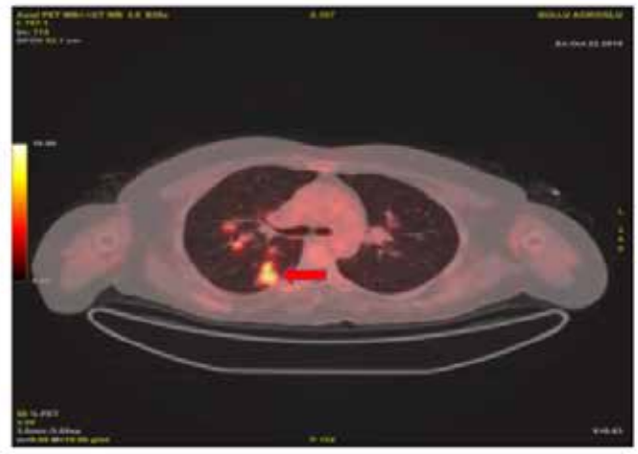


Figure 7. Increased nodular hypermetabolic activity is observed in the right lung.

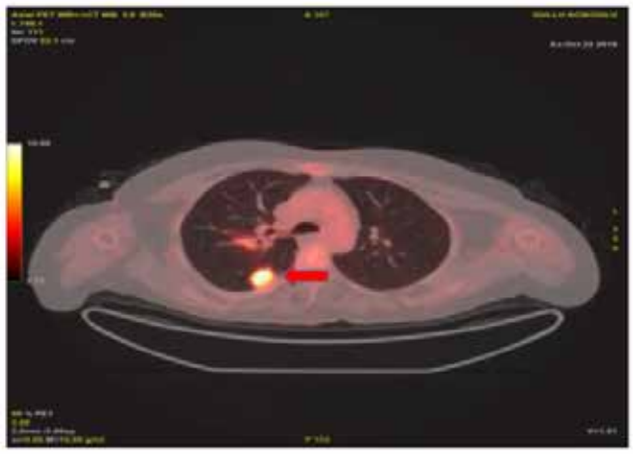


Figure 8A. Increased nodular hypermetabolic activity is observed in the right lung.

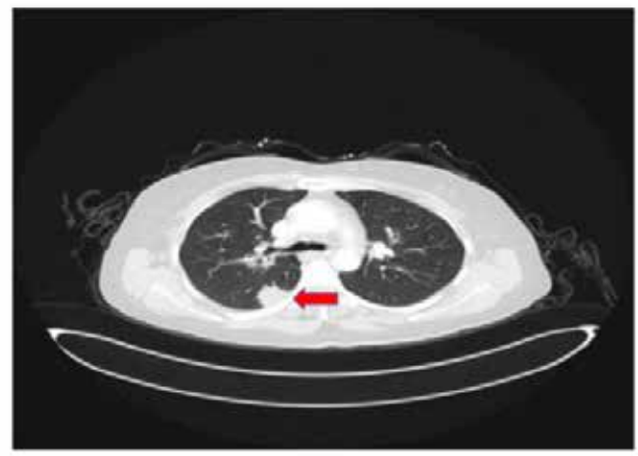


Figure 8B. CT image of the nodule showing hypermetabolic activity in the right lung is observed.

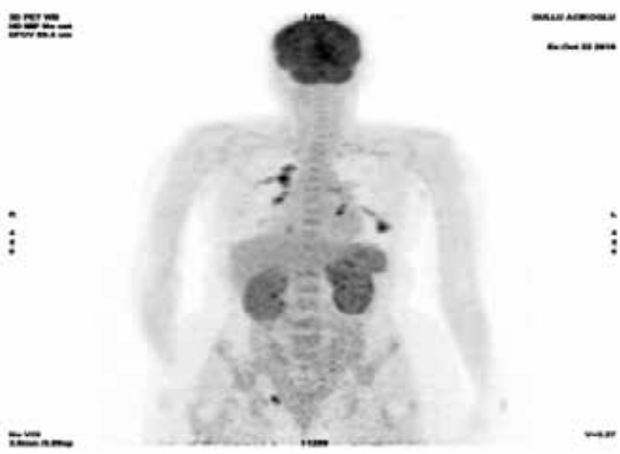


Figure 9. In the whole body coronal section, diffuse hypermetabolic activity is observed in bilateral hilar regions, spleen and bilateral kidneys.

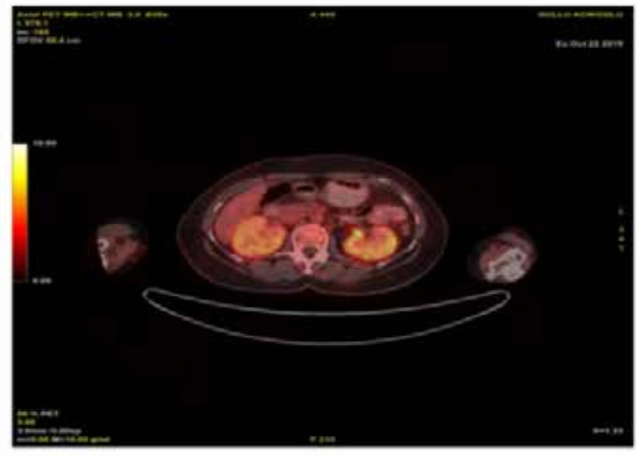


Figure 10. Hypermetabolic activity is observed in bilateral kidneys.

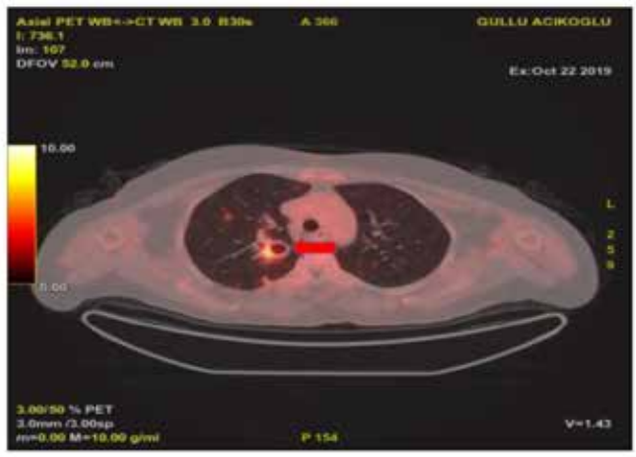


Figure 11A. A cavitary lesion with hypermetabolic activity is observed in the right lung.

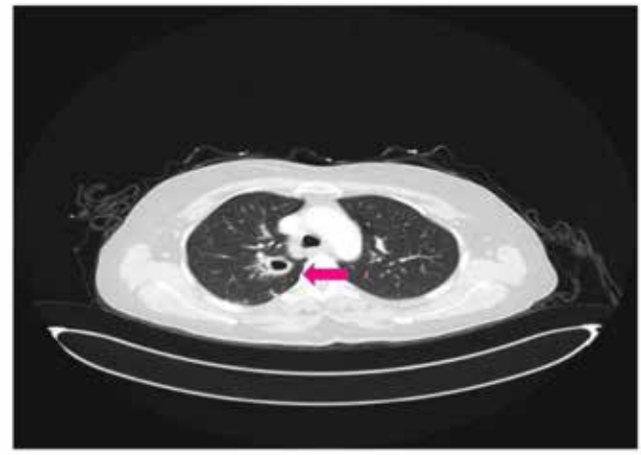
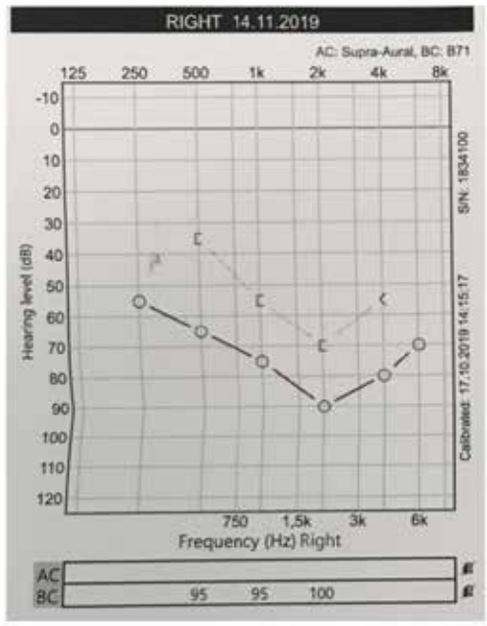
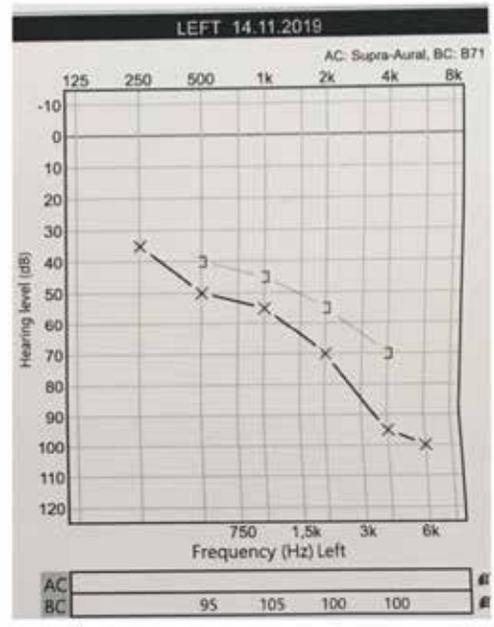


Figure 11B. CT image of the cavitary lesion showing hypermetabolic activity in the right lung is observed.





Scientific Responsibility Statement
The authors declare that they are responsible for the article's scientific content including study design, data collection, analysis and interpretation, writing, some of the main line, or all of the preparation and scientific review of the contents and approval of the final version of the article.

Animal and human rights statement
All procedures performed in this study 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. No animal or human studies were carried out by the authors for this article.

Conflict of interest
None of the authors received any type of financial support that could be considered potential conflict of interest regarding the manuscript or its submission.

References
1. Devaney KO, Travis WD, Hoffman G, Leavitt R, Lebovics R, Fauci AS. Interpretation of head and neck biopsies in Wegener's granulomatosis. A pathologic study of 126 biopsies in 70 patients. Am J Surg Pathol. 1990; 14(6):555-64.
2. Jennette JC, Falk RJ. Small-vessel vasculitis. N Engl J Med. 1997; 337(21):1512-23.
3. Falk RJ, Hogan S, Carey TS, Jennette JC. Clinical course of anti-neutrophil cytoplasmic autoantibody-associated glomerulonephritis and systemic vasculitis. The Glomerular Disease Collaborative Network. Ann Intern Med. 1990; 113(9):656-63.
4. Seo P, Stone JH. The antineutrophil cytoplasmic antibody-associated vasculitides. Am J Med. 2004; 117(1):39-50.
5. Hunder GG, Arend WP, Bloch DA, Calabrese LH, Fauci AS, Fries JF, et al. The American College of Rheumatology 1990 criteria for the classification of vasculitis. Introduction. Arthritis Rheum. 1990; 33(8):1065-7.
6. Trimarchi M, Sinico RA, Teggi R, Bussi M, Specks U, Meroni PL . Otorhinolaryngological manifestations in granulomatosis with polyangiitis (Wegener's). Autoimmun Rev. 2013; 12(4):501-5.

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