

Wegener's Granulomatosis: Clinico-Radiological Finding at Initial Presentation

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ABSTRACT

Diagnosis of Wegener's granulomatosis at the early stage is difficult because of the nonspecific symptoms which mimic other disorders. The aim of this paper is to describe clinical and radiological features of Wegener's granulomatosis (WG) in a Serbian population at initial presentation. A retrospective review of 37 patient's case records was carried out. All those patients were diagnosed with WG and they attended the Institute for lung diseases in Belgrade over the period of 15 years. There were 20 males and 17 females, ranging in age from 18 to 73 years (mean age 46.2 years). The mean period from the onset of the first symptoms to diagnosis of WG was 4.59±6.15 months. The criteria of American College of Rheumatology were fulfilled in all patients. Twenty-five of 37 patients had systemic, generalized form of WG and while 12 of them had a limited involvement of upper and lower respiratory system. The frequency of different system involvement was: upper respiratory tract 64.8%, lower respiratory tract 100%, kidneys 67.5%, musculoskeletal system 40.5%, skin 27.2%, eyes 8.1%, and nervous system two patients. ANCA (antineutrophil cytoplasmic antibodies) test was positive in 32(86.5%) patients, and negative in 5(13.5%). All patients were ANA negative. Histological evidence of granulomatous vasculitis was obtained in 34(91.9%), whereas in three patients the diagnosis was based on clinical manifestations and positive c-ANCA test. There are minor variations in our data when compared with those reported in literature.

Key words: Wegener's granulomatosis, radiology, kidney, lung lesion

Introduction

Wegener's granulomatosis is a distinct syndrome characterized by necrotizing granulomatous vasculitis that affects small and medium blood vessels of the involved organs. The cause of disease is not known. It belongs to a group of primary systemic vasculitides of unknown etiology that are associated with antineutrophil cytoplasmic antibodies (ANCA)¹⁻⁴. The most frequently affected organs are upper respiratory tract, lungs and kidneys. The spectrum of diseases extends from a limited or localized involvement to a florid, extensive, and fatal multisystemic illness¹⁻³. A limited form may be presented by pulmonary lesions without kidney lesions. This variant has a good prognosis. The nonspecific symptoms of the disease like fever, arthralgia, skin lesions can mimic other dis-

eases and cause delay in the diagnosis^{1,2}. In addition to clinical and radiographic findings, the diagnosis of WG is confirmed by antineutrophil cytoplasmic antibodies as serological marker and biopsy. Detection of c-ANCA is a useful test in diagnosing WG⁴. With the introduction of ANCA as a serological marker, WG is diagnosed at an earlier stage and with a greater degree of certainty. The test however may be negative in localized form of disease^{4,5}.

The purpose of this report is to show a group of 37 patients with Wegener's granulomatosis (WG) from our Institute, to analyze the clinical and radiological manifestations and compare our results with other published series.

Patients and Methods

The group of patients that were hospitalized at Institute for chest diseases and TB due to respiratory symptoms and non-resolving chest x-ray opacities in the period of 15 years was included in this study. Patients underwent an interdisciplinary clinical examinations to assess the extent of disease. The clinical examination included ear, nose and throat, respiratory and general physical examination. The radiological examination included plain chest x-ray and computerized tomography (CT) of thorax and paranasal sinuses. In one patient with high grade subglottic stenosis, magnetic resonance imaging (MRI) of the neck was done. Ultrasonography of abdomen and kidney were obtained in all patients. Lung function test and bronchoscopy were done in two thirds of patients. The laboratory data recorded were haemoglobin concentration, complete blood cell count, ESR, serum creatinine level, urine examination for red blood cells, proteins and casts. The presence of c-ANCA and p-ANCA was detected by using the indirect immunofluorescence and ELISA method⁴. Tests for antinuclear antibody were done in every patient. Presenting laboratory features and clinical signs at the time of diagnosis were analyzed. The time span from onset of the first symptoms related to Wegener's granulomatosis to the diagnosis was also recorded. In order to confirm the diagnosis we used the criteria introduced in 1990 by The American College of Rheumatology (ACR)⁶. The diagnosis was made by these criteria in conjunction with the clinical symptoms, histopathology findings and blood test results for serum c-ANCA and p-ANCA.

Results

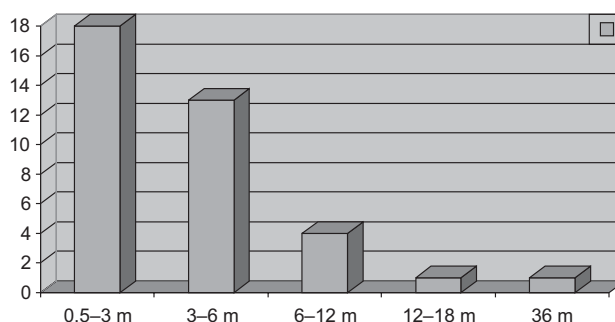
Patient's features

The group had 37 patients, with a majority of male (54%). The mean age was 46.2 years, (range from 18 to 73 years of age). Mean period from the onset of first symptoms to a diagnosis of Wegener's granulomatosis was 4.59±6.15 months (Table 1). In most patients diagnosis of WG was made within six months of the onset of

TABLE 1
DEMOGRAPHIC FEATURES OF PATIENTS WITH WG*

	Our series	Hoffman et al.(2)	Fauci et al.(1)
Number of patients	37	158	85
M:F	1:1.18	1:1	1.6:1
Age			
Mean	46.2	41	43.6
Range	18-73	9-78	14-75
Duration of symptoms before diagnosis(months)			
Mean	4.6	15	8.3
Range	0.5-36	0-120	0.5-120

*Wegener granulomatosis



Graph 1. Duration of symptoms (in months).

symptoms (Graph 1). Twenty-five (67.5%) patients had constitutional symptoms like fever, malaise, weight loss.

Organ system involvement

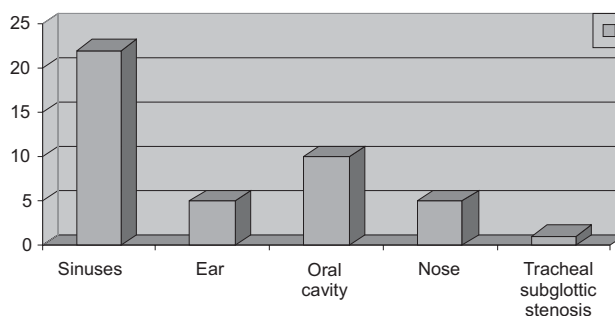
Majority of patients initially sought medical care because of upper or lower airway symptoms (Table 2). Out of 37 patients, 12 (32.5%) had limited WG with involvement of only upper and lower respiratory tract. The classic triad of upper and lower respiratory tract and kidney involvement at the time of diagnosis was seen in 25 (67.5%) patients. The frequency of different system involvement at initial presentation is shown in Table 3.

Respiratory clinical features

Upper respiratory tract involvement was noted in 24 (64.8%) patients. This included nasal, sinus, ear and pharyngeal symptoms (Graph 2). Inflammation of paranasal sinuses was presented with chronic sinusitis. Three of our patients suffered from nasal granulomatous inflammation and two from crusting. All of them have had

TABLE 2
CLINICAL SYMPTOMS IN WEGENER'S GRANULOMATOSIS

Symptoms	Our series	Hoffman et al.(2)	Fauci et al.(1)
Constitutional	25 (67.5%)	36 (23%)	29 (34%)
Cough	37 (100%)	30 (19%)	29 (34%)
Hemoptysis	34 (91.9%)	19 (12%)	15 (18%)
Dyspnea	18 (48.6%)	NR	6 (7%)
Epistaxis	5 (13.5%)	25 (16%)	9 (11%)



Graph 2. Upper respiratory tract involvement.

TABLE 3
THE FREQUENCY OF DIFFERENT SYSTEM INVOLVEMENT AT INITIAL PRESENTATION

	Our series	Hofman et al.(2)	Fauci et al.(1)
Number of patients	37	158	85
Organ involvement, No(%)			
Upper respiratory tract	24 (64.8%)	115 (73%)	57 (67%)
Lung	37 (100%)	71 (45%)	60 (71%)
Kidney	25 (67.5%)	28 (18%)	9 (11%)
Joints	15 (40.5%)	50 (32%)	37 (44%)
Skin	10 (27%)	20 (13%)	11 (13%)
Eye	3 (8.1%)	24 (15%)	14 (16%)
Peripheral nervous system	2 (5.4%)	24 (15%)	1 (1.17%)

bloody nasal discharge for several weeks. The symptoms of ear involvement were earache, hearing loss and tinnitus. The main finding on examination was suppurative otitis media. Ten (27.9%) patients had ulceration of oral mucosa.

Lower respiratory system was involved in all patients at initial presentation. Pulmonary symptoms included cough in all patients, dyspnea and hemoptysis (Table 2). Five patients (13.5%) had only lung involvement. Lung function tests were done in 24 patients and showed restrictive defect related to space-occupying lesions in the chest in 12 (50%) patients, air flow obstruction in six, and mixed pattern of restriction and obstruction in three patients. Normal lung function test was in three patients. In other patient these tests were unavailable. Twenty-seven (73%) patients underwent bronchoscopy which was abnormal in 22 (81%) cases. Pathologic endobronchial finding consisted of inflammation in 19 cases,

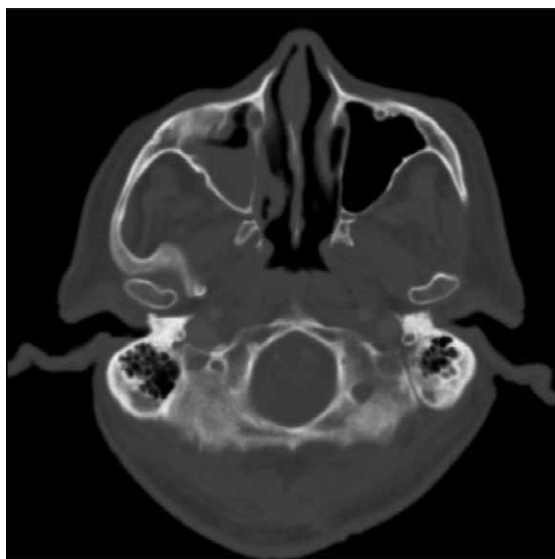


Fig. 1. CT of sinuses shows mucosal thickening and air-fluid level of the right maxillary sinus.

TABLE 4
THE FREQUENCY OF RADIOLOGICAL FEATURES IN THE LUNGS

Radiological features	Our series	Hofman et al.(2)	Kumar et al.(5)
Nodules and mass	2+3 (13.5%)	35 (22%)	5 (20 %)
Cavitation	12 (32.4%)	NR	9 (36%)
Consolidation	19 (54.0%)	36 (23%)	7 (28%)
Pleural effusion	1 case with consolidation	16 (10%)	0
Number of patients	37	158	25

isolated hemorrhage in two and compression of the bronch by tumor mass in one case. Bronchial biopsies showed mild nonspecific chronic inflammation in 9 cases and granulomatous vasculitis in 10 cases. Lung involvement was confirmed by open thoracotomy in five patients and by lung biopsy in four cases.

Upper and lower airways imaging

Plain film and CT of paranasal sinuses revealed mucosal thickening with opacifications in 19 patients and air-fluid levels in three cases (Figure 1).

Pulmonary symptoms were associated with chest x-ray abnormalities in all of our patients at the time of presentation (Table 4).

Radiographs or CT of the thorax showed bilateral opacities in 62.1% and unilateral in 37.9%. In 20 (54%) patients areas of consolidation were observed. These opacities spared the apices and ranged from focal consolidation in five cases to frank bilateral consolidation in 12 patients (Figure 2). Cavitations of the consolidations occurred in 5 (25%) patients. Diffuse, bilateral, and low density opacities occurred in three patients (Figure 3). One patient with focal consolidation had accompanying



Fig. 2. Thoracic CT scan with soft tissue window demonstrates ill-defined parenchymal consolidation bilateral, with a small cavitation in the left lung.

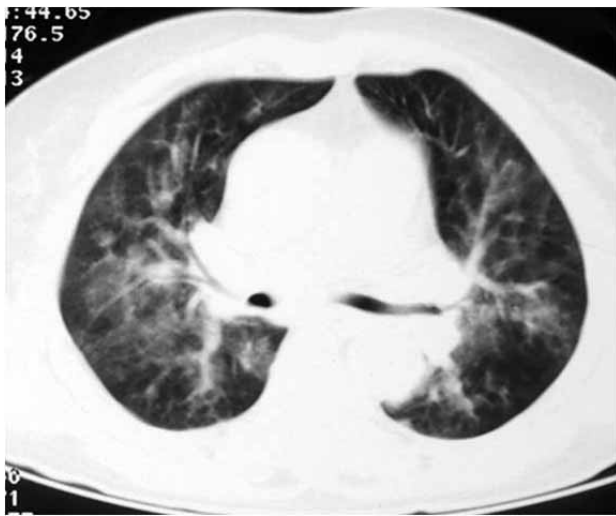


Fig.3. Chest CT scan with lung window demonstrates ground glass opacities in both lungs with small nodules along with bronchovascular bundle.

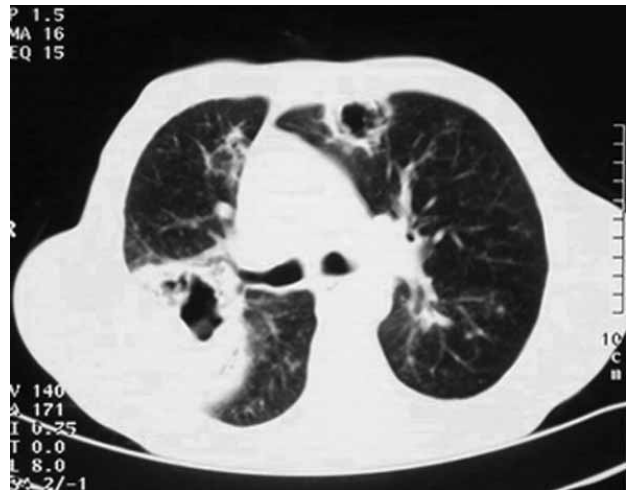


Fig. 5. Chest CT scan with lung window demonstrates a large cavitary mass with thick irregular borders in the right upper lobe, and the smaller one in the left lung.

pleural effusion. Nodules were present in 17 (45.9%) patients, five without cavities (Figure 4) and 12 (64.7%) with cavities (Figure 5). They were bilateral in 10 (58.8 %) patients. The number was from two to innumerable and the size varied from 1.0 to 10 cm. Hilar and mediastinal adenopathy was rare finding, just in one patient. One patient with massive bilateral opacities had associated high grade subglottic tracheal stenosis (Figure 6).

Kidney involvement

Twenty-five of our patients had kidney disease. All of them had hematuria and proteinuria at initial presentation. Blood biochemistry analysis revealed elevated levels of serum urea and creatinine in 25 patients and azotemia

in 19 of those (67.5% and 76.0% respectively). Median serum creatinine level was 414.8 $\mu\text{mol/L}$. Ultrasonography of kidneys confirmed morphological changes in 17 (68.0%) patients. Large echogenic kidneys on ultrasound without specific Doppler abnormalities were most common finding in our patient. Diagnosis of WG was confirmed in 8 patients by percutaneous renal biopsy.

Clinical features of the other organ involvement

Musculoskeletal symptoms were present in 15 (40.5%) patients with arthralgia and myalgia. Skin disease manifestations occurred in 10 (27.02%) patients: four with purpura, three with ulceration, maculae and nodules one each and one with mastitis. In five patients skin biopsy showed necrotizing vasculitis.

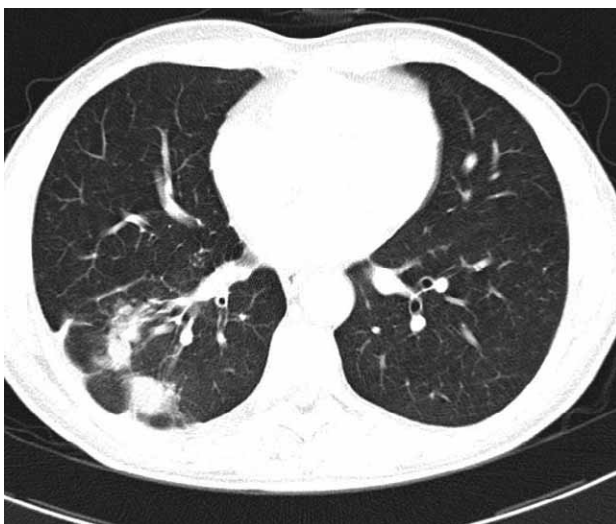


Fig. 4. Chest CT scan with lung window shows multiple nodules in right lower lobe.



Fig. 6. MRI of the neck in sagittal view shows high grade subglottic stenosis.

TABLE 5
BIOCHEMICAL FEATURES IN PATIENTS WITH WG

Abnormality	Our series	Hofman et al.(2)	Kumar et al.(5)
Elevated ESR	23 (62.1%)	126 (80%)	25 (100%)
Anemia	16 (43.2%)	115 (73%)	22 (90%)
Leukocytosis	20 (54.0%)	NR	25 (100%)
Proteinuria	25 (67.5%)	28 (18%)	18 (72%)
Hematuria	25 (67.5%)	28 (18%)	18 (72%)
ANCA	32 (86.5%)	139 (88%)	17 (70%)

Ocular abnormalities were seen in 3(8.1%) patients with signs of scleritis, conjunctivitis and retinal vasculitis. Nervous system was involved in two patients manifesting with mononeuritis multiplex. There were no patients in our series with central nervous system involvement.

Laboratory findings

Usual laboratory tests revealed the raised erythrocyte sedimentation rate and leukocytosis in more than half patients, anemia and leukocytosis in 11 patients, leukocytosis in 9, and only anemia in five. Twenty-five patients were c-ANCA positive, six patients had positive p-ANCAs with anti MPO antibodies. One patient was both, p- and c-ANCA positive. Negative ANCA test results were in 5 (13.5%) patients. All patients were ANA negative. Presenting features are shown in the Table 5.

Diagnosis

Diagnosis of WG was based on the evaluation of clinical manifestations and histological analysis of involved organs. American College of Rheumatology criteria were fulfilled in all patients. In three of 37 patients, the diagnosis was established on the basis of clinical manifestations and positive c-ANCA tests with blood serum titers from 1:32 to 1:1024. Pathohistological verification was achieved in 34 (91.9%) patients from biopsy of involved organs (bronchi 10 cases, lung 9 cases, kidneys 8 cases, skin 5 and oral mucosa in two cases). Positive both ANCA test and biopsy were found in 29 (78.3%) patients while negative ANCA test with positive biopsy was registered in 5 (13.5%) patients.

Discussion and Conclusion

Patient's characteristics

Wegener's granulomatosis (WG) is systemic, necrotizing granulomatous vasculitis primarily affecting the upper and lower respiratory tracts and kidneys, but it may also involve any other organ. The onset of disease is usually between 25 and 50 years, but it can occur in childhood. The mean age of our patients (46.2 years) is similar to that of other series. The sex ratio was about equal. The course varies from acute systemic disease with lung and

renal manifestations to subacute or even indolent. There is usually a delay in the diagnosis in patients with a mild disease and indolent progression⁵ which occurred in two of our patients. Diagnosis of Wegener's granulomatosis in the early stage is difficult because of the nonspecific symptoms which mimic other infectious, granulomatous diseases, vasculitides and neoplastic disorders. Initial nonspecific symptoms include fever, malaise, weight loss, arthralgia, and myalgia. These symptoms were more frequent in our patients than previously reported^{1,2}.

Upper and lower airway manifestations

Most patients have initial upper airway symptoms, such as nasal discharge, sinusitis, or epistaxis^{5,7,8}. The degree of involvement differs ranging from rhinorrhea and nasal discomfort to nasal mucosal ulceration. Sinusitis is present at initial presentation in about one half to two thirds of patients with WG⁷. Sinusitis can be mild or involve the bony wall of sinuses. Radiologic imaging studies are helpful in diagnosing Wegener's granulomatosis, including sinus radiographs and CT. A computerized tomography is more informative than plain radiographs in cases with destructive and erosive bone changes. Otitis media is usually the result of blockage of the Eustachian tube. Suppurative otitis, sensory neural deafness and mastoiditis can occur^{2,5,9}. Oral lesions are rare at initial presentation. The disease may remain localized to the oral cavity for several weeks or months before multi-organ involvement occurs¹⁰. This manifestation of WG was more frequent in our patients than previously reported^{1,2,15}. Characteristic pathologic changes of WG are often absent from the biopsy specimens of upper airways. Upper respiratory tract involvement in our series is lower than previously reported^{2,11,12,13}.

Pulmonary symptoms like cough, hemoptysis and dyspnea were more frequent in our series than in the literature data^{2,11,12,14}. That can be explained by the fact that all patients were collected from the Institute for chest diseases and TB. Lung function test were done in 64.8% patients at initial presentation and the most common abnormality was restrictive pattern. Fiberoptic bronchoscopy in most patients with WG shows nonspecific inflammatory lesions. Lung biopsy specimens, especially if obtained by an open-lung procedure, usually reveal necrotizing granulomas and vasculitis^{11,12}.

Radiographic changes in the lungs are found in 50–75% of patients. The chest CT is more useful for further characterizing the image features. It can reveal the lesions which were not seen on the chest x-ray. Furthermore, CT often shows previously unsuspected necrotic content in nodules and consolidations. Most characteristic pulmonary findings are opacities that vary in the character from nodular masses to ill-defined areas of consolidation, both of which may cavitate^{28–33}. The nodules are rounded, range from a few millimeters to several centimeters in size, the largest being usually excavated. This finding is highly suggestive of WG. The number of nodules is usually less than ten but may be innumerable^{5,12–14}. The focal consolidations are commonly pleural based and

have a wedge shaped configuration with or without air bronchogram. The patchy and of low density infiltrates have also been described^{11,14–16}. As with three of our patients, diffuse, bilateral and low density patterns may occur. Pathologic examinations of the lung in such cases reveal diffuse hemorrhage and capillaritis¹⁷. This is a serious manifestation of lower respiratory tract involvement. In patients with pulmonary hemorrhage concurrent kidney failure may be present. It may result in the erroneous diagnosis of Goodpasture disease despite the absence of antiglomerular basement membrane antibody¹⁷. Some times there is a mixed pattern of nodules and consolidation. The present series shows difference in radiological features in the lungs when compared with reported series. In our study, nodules and areas of consolidation were observed at the same frequency. In the studies by Kumar⁵, Lee¹⁴ and Cordier¹⁵ the most common finding of WG were nodules or masses followed by consolidation. In all of these studies, one half to two thirds of nodules and masses cavitated. The study by Lohrman et al.¹⁶ found the nodules in 89% patients and mixture of consolidation, nodules and bronchial wall thickening in about a half of patients. The finding similar as in our study was found by Hoffman et al. in a series of 158 patients².

Peribronchovascular thickening also has been reported. In the study by Lee et al.¹⁴ this finding was detected on CT in 73% of patients. Although the large airway manifestations of Wegener's granulomatosis are not frequent at initial presentation, tracheal narrowing is not unusual finding³. Tracheal stenosis is usually subglottic as occurred in one patient in our series. It manifests on CT or MRI as a mural thickening and narrowing of involved part¹⁶. Laryngeal involvement has been described in up to 30% of patients^{14,19}. Lesions less often affect true vocal cords and epiglottis and may be ulcerative or proliferative. In one of our patient, ulceration affected true vocal folds. In other reported studies frequency of large airway involvement was as similar as in our study². A variety of less common intrathoracic manifestations in WG have been reported. Pleural effusions, pneumothorax and hydropneumothorax occur rarely and vary in severity^{16,20,21}. Hilar and mediastinal adenopathy have been detected in up to 2% of cases, typically with other pulmonary manifestations of WG¹⁸. This finding was seen in one of our patients.

Renal manifestations

The proportion of patients with kidney involvement at disease presentation has varied between studies from less than 20% to 80%. Kidney involvement in Wegener's granulomatosis heralds a more severe outcome²². Microscopic or macroscopic haematuria with proteinuria is usually the only evidence of renal involvement^{22,23}. It was noted in all patients with kidney failure manifestations in our series. The renal involvement in our study was much higher than in other reported series. In studies by Hoffman² and Fauci¹ renal involvement was present in less than 20% of patients. Study by Aasarod K et al. revealed in 34.3% of 108 patients a various degrees of renal

involvement at disease presentation²². Large echogenic kidneys on ultrasound without specific Doppler abnormalities are the most common early finding. Although this is a non-specific finding, it is the most common sign of early glomerulonephritis, implying involvement of the kidney by Wegener's granulomatosis. Renal biopsy specimens reveal changes typical of focal segmental glomerulonephritis or a necrotizing proliferative glomerulonephritis with crescent formation^{2,22}.

Other organ manifestations

Arthralgia and arthritis of small and large joints are seen in 30–68 % of cases and may be misdiagnosed as rheumatoid arthritis unless other features typical of WG are present^{2,5,24}. Our patients had non specific musculoskeletal symptoms with arthralgia and myalgia. Skin is also involved in some cases. Urticaria, palpable purpura, papules, vesicles, erythema and petechiae are the common manifestations^{1,2,5,25}. Skin manifestations of WG were more common in our study than in reported series^{1,2,15}. Patients with histologically confirmed necrotizing vasculitis in cutaneous lesions have worse prognosis than those with a predominant granulomatous reaction²⁵.

Ocular abnormalities like proptosis, conjunctivitis, episcleritis, sclerocorneal ulcerations, uveitis, retinal vasculitis, central retinal artery occlusion are present at the beginning of disease in 15–30%^{2,12,26}. Only three (8.1%) of our patients had mild ocular symptoms with scleritis, conjunctivitis and retinal vasculitis.

Neurological conditions like mononeuritis multiplex, peripheral neuropathy are seen in about 8% of cases at early stage while central nervous system abnormalities (cranial nerve palsies, cerebral infarcts, transverse myelitis and seizures) occurred rarely^{1,2,27,28}. Only two of our patients had neurological disorders with mononeuritis multiplex.

Biochemical analyses

Usual laboratory tests are nonspecific because they only point to an inflammatory syndrome with a raised erythrocyte sedimentation rate, leukocytosis and anemia^{2,15}. The association between Wegener's granulomatosis and the presence of antineutrophil cytoplasmic antibodies (ANCA) in the blood has been reported by numerous investigators^{4,29}. Even though a positive c-ANCA test is of great value in confirming a diagnosis of WG, it is still negative in some cases, particularly in those with localized form of the disease^{2,30}. It was found negative in five (13.5%) patients in our series (four with limited and one with generalized form of WG).

The results of our study show that clinical symptoms and radiologic features of lung diseases at initial presentation of WG are slightly different from the reported series^{2,5,11,12,22,13–16}. It could be explained by relatively small number of patients and by the fact that all patients were recruited from a referral center.

Today WG is more frequently diagnosed in Serbia at an early stage of disease because of awareness and avail-

ability of diagnostic techniques. In patients who have non-resolving chest x-ray opacities a thorough clinical examination and all relevant investigations should be performed. Upper and lower respiratory tract symptoms along with active urine sediment should raise the clinician's awareness of Wegener's granulomatosis.

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WEGENEROVA GRANULOMATOZA: KLINIČKE I RADIOLOŠKE ZNAČAJKE NA POČETNOJ PREZENTACIJI

SAŽETAK

Dijagnoza Wegenerove granulomatoze u ranoj fazi je teška zbog nespecifičnih simptoma koji imitiraju druge bolesti. Cilj ovog rada je opisati kliničke i radiološke značajke Wegenerove granulomatoze (WG) u srpskoj populaciji na početnoj prezentaciji. Proveden je retrospektivni pregled podataka za 37 pacijenata. U svih ovih pacijenata je dijagnosticirana WG i oni su bili na Institutu za plućne bolesti u Beogradu tijekom razdoblja od 15 godina. Bilo je 20 muškaraca i 17 žena, u dobi od 18 do 73 godina (prosječne dobi 46,2 godina). Prosječno razdoblje od početka prvih simptoma do dijagnoze WG je $4,59 \pm 6,15$ mjeseci. Kriteriji Američke Visoke škole za reumatologiju su ispunjeni u svih bolesnika. Dva-deset i pet od trideset i sedam bolesnika imalo je sustavan, generalizirani oblik WG, dok je njih 12 imalo ograničenu zahvaćenost gornjeg i donjeg dišnog sustava. Učestalost zahvaćenosti različitih sustava je: gornjih dišnih putova 64,8%, donjih dišnih putova 100%, bubrega 67,5%, mišićnoskeletnog sustava 40,5%, kože 27,2%, oči 8,1%, a živčanog sustava u dva bolesnika. ANCA (Antineutrofilna citoplazmatska antitijela) test je bio pozitivan u 32 (86,5%) bolesnika, a negativan u 5 (13,5%). Svi bolesnici su bili ANA negativni. Histološki dokaz granulomatozna vaskulitisa ostvaren je u 34 (91,9%), dok se u tri bolesnika dijagnoza temelji na kliničkim manifestacijama i pozitivnom c-ANCA ispitivanju. Postoje manje razlike u našim podacima, u usporedbi s onima u literaturi.