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Case Report

Case Report of Granulomatosis with Polyangiitis Formerly Known as Wegener's Granulomatosis

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Abstract

Granulomatosis with polyangiitis also known as Wegener's Disease is involving mostly upper respiratory tract and sometimes kidney. It's very rare that these disease start from kidney and involve both kidney's. We present a case of 50 years old female patient who developed unspecific symptoms in kidney's and after two operation on both kidney's the diagnosis was not sure in histopathology. The extension of diagnosis which include: CT scan, immunology (antinuclear antibodies c-ANCA and p-ANCA) showed that it could be granulomatosis with polyangiitis which symptoms started from kidney's. This publication is showing step by step the path of the patients from symptoms to correct diagnosis.

Keywords: Wegener's Disease; Granulomatosis with Polyangiitis; Renal Mass; Anti-neutrophil Cytoplasm Antibody

Granulomatosis with polyangiitis formerly known as Wegener's granulomatosis (WG) is a systematic disease with necrotic inflammation of small and medium vessels that mainly affects the upper respiratory tract, lung and kidney [6].

At the diagnosis, the kidneys are involved in 20% of cases, but ultimately kidney involvement occurs in 80% of patients [3,4]. Determination of antinuclear antibodies c-ANCA, which is elevated in 90% of patients with Wegener's granulomatosis [9], can help in diagnosis. Renal lesions appear as segmental necrotizing glomerulonephritis, which rapidly leads to renal failure [7,8]. Rarely, this disease manifests itself as a focal lesion in the kidney [1,2,5].

We describe a case in which the dominant element of the disease was multifocal nodular changes in the kidney – very rarely occurring in Wegener's disease.

Case report a 50 year old patient hospitalized for the first time in 2009 in the Department of General and Oncological Urology in Mielec due to suspicion of a left kidney tumor. The ultrasound image of the changes suggested possible inflammatory changes in the dorsal part of the left kidney caused by urolithiasis blocking the middle calyx neck. In performed imaging examination (ultrasound and CT scan with contrast of abdominal and pelvis) stone of 12 mm in diameter, blocking the neck of the middle calyx of the left kidney was found. Additionally, peripherally, the area of weakly strengthening parenchyma 50x51x54 mm with fluid spaces (tumor vs. Abscess) and perirenal abscess were visualized as „focal yellow acute chronic pyelonephritis on the background of urolithiasis of the urinary tract with perirenal abscess. Importantly for the case the right kidney showed no significant changes in the imaging examination. Chest X-RAY showed no patological changes. In laboratory tests performed at the time, there were no significant

abnormalities in blood count. In blood biochemistry, elevated creatinine value: 115 [44-80 qmol/L]. After proper preparation and and anesthetic consultation, the patient was qualified for NSS (nephron sparing surgery) on the left side. An intraoperative change was found in the central part of kidney, with heterogeneous morphology, which did not allow for an unequivocal assessment ad its nature. Liquid content – most likely purulent was collected for microbiological examination. The lesion was completly evacuated and edges were re-marked to assess the resection margins. Additionally, the deposit blocking the calyx and penetrating into the renal pelvis was removed. The procedure and the postoperative course were uneventful. Proteus mirabilis was cultured in the obtained purulent content – antibiotic therapy was carried out according to the culture results. The obtained histopatological result was described as: „mixed – cell inflammatory infiltration without features". The patient was under the care of the Urology Clinic – no deviations from the norm were found.

In August 2012. reported to the Outpatient Clinic due to fever and elevated inflammatory parameters. In the ultrasound performed at the time, a tumor vs. an abscess of the right kidney was suspected. The patient was urgently referred to the hospital in order to extend the diagnosis and therapy. The patient was treated in Department and Internal Medicine and Nephrology - from where she was then referred to the Urology Department. On admission as well as during hospitalization, the patient was not feverish, did not report any dysuria symptoms, but only weakness. Laboratory tests showed an increased level of lecocytes in the blood and high CRP values (>200 ng/ml [<5 ng/ml]). These values systematically decreased, which was correlated with the improvement of the clinical condition. Urine bacteriological culture was performed, which showed the presence of Klebsiella pneumoniae sp. Antibiotic therapy was introduced: Amikacin (empirical) and then Imipenem (targeted). The CT examination of the abdominal cavity and pelvis showed focal changes in the right kidney with a suggestion of an abscess changes in the upper pole of the right kidney (37x42x45 mm and 30x22x29 mm). After treatment the patient was discharged home in good condition for further treatment on an outpatients basis. In September 2012. the patient was referred to the Urology Department again due to ineffective outpatient treatment and increasing weakness. On admission, the patient reported weakness and periodic pain in the right lumbar region and recurrent inflammation of the middle ear. Due to last illness, the patient was treated at the ENT Department of the clinical hospital.

Empirical antibiotic therapy (Amikacin) was applied and microbiological diagnostics (bacteriological cultures of urine and blood) were started. In the CT of the abdominal cavity and pelvis with contrast: thick-walled lesions with fluid content were found in the right kidney: 60 mm in diameter in the upper pole, in the lower pole by a diam. 40 mm and second smaller diam. 10-15 mm. Moreover, there was a change in the left kidney with an average ad 15 mm – also an abscess. As a conclusion in the imaging examination it was found: "disseminated inflammatory process". Importantly, laboratory tests did not show any deviations correlating to inflammation. During hospitalization symptoms characteristic of *C. difficile* infection appeared, which was confirmed by microbiological tests. After consultation with the Infectious Diseases Doctor, the Patient was transferred to the Department of Infectious Disease in order to continue the treatment in a sanitary regime. 15 October 2012 the patient returned to the Urology Department.

On the day of admission, control laboratory tests were performed, in which the following deviations were found; HgB 8,9g/dl [12-16 g/dl]; creatinine 117 qmol/l [44-80 qmol/l] at GFR 44 ml/min/1,73m^2. Due to clinical condition and anemization, 2 units of red cell concentrate (230ml each one) were transfused. Due to the unchanged ultrasound image of the right kidney and due to clinical condition and the lack of effectiveness of treatment, the patient was qualified for surgery in order to verify the changes intraoperatively. After proper preparation and anaesthesiological consultation, the procedure was performed via retroperitoneal access. During the procedure, the kidney was found to be with nodular changes – no abscesses were confirmed. Therefore, a decision was made to remove the organ. In the cross- section of the specimen, the entire organ is occupied by solid nodular changes (Figure 1). The intraoperative course was uneventful. A few hours after the surgery, the patient developed a fever – symptomatic treatment and empirical antibiotic therapy were introduced. In the microbiological tests collected at the time (urine and blood specimen), Staphylococcus haemolyticus was found in the bacteriological culture – target antibiotic therapy (Vankomycin) was implemented. As a result of the treatment, the clinical condition was stabilized. Postoperative process ran without any other complications. The patient was discharged home in good general condition. The obtained histopathological result revealed: "diffuse infiltration of histiocytic cells and presence of numerous

necrotic abscesses surrounded by giant multinucleated cells. The inflammatory infiltrate contains plasmocytes, lymphocytes and granulocytes. The infiltration is present in the kidney's fibrous capsule and in the perirenal fat tissue. This could correspond to the diagnosis of pyelonephritis xantogranulomatosa".

Figure 1: Right kidney with nodular changes.

The patient was under the care of the Urology and Nephrology Clinics.

The available documentation shows that the patient was hospitalized in 11.2012. at the Clinical Department of Internal and Metabolic Diseases with the Nephrology Subdivision of the Medical University in Warsaw (Poland). Then, in the interview, the patient also reported: recurrent infections of the middle ear, increasing weakness, joint and muscular pains, paresis of the VII cranial nerve and visual acuity disturbances in the left eye. As part of hospitalization, the following were performed: CT of the chest showing lumpy changes in the lungs; CT scan of the paranasal sinuses showing massive granulation tissue in mastoid and sinus processes. Serological tests revealed increased levels of p-ANCA and c-ANCA. Based on the overall clinical data, the occurrence of the above symptoms provided the basis for an initial diagnosis of primary systematic vasculitis of Wegener's granulomatosis. In addition, all specimens obtained during previous operations were analyzed again.

Based on the above analyzes, the patient remained under specialist treatment focused on Wegener's disease. The left kidney allows it to function without dialysis. At the present, the patient's condition is stable.

It should be discussed whether in 2009 lesions removed from the left kidney might suggest the above diagnosis. The answer should be the medical opinion of Professor Renata Langfort (Head of the Department of the Pathomorfology at the Institute of Tuberculosis and Lung Diseases in Warsaw): (…) It should be assumed that in 2009 the process corresponded to inflammatory changes related to the presence of urolithiasis, while it was found in the 2012 sections can be associated with Wegener's granulomatosis (…). The clinical data available at the time of diagnosis by diagnosing physicians did not provide a basis for establishing such a difficult diagnosis, which entails very burdensome therapeutic consequences".

Summing up, granulomatosis with polyangiitis (WG) is most often multi organ disease and only 30% of patients may experience symptoms in one organ but then the upper respiratory tract or the lungs are affected. Renal changes accompany the generalized form are most often manifested by glomerulonephritis with the formation of crescents and focal necrosis of the vascular loops – very rarely in the form of macroscopic changes.

Due to its non-specific nature, symptoms, the above diagnosis is very difficult to establish in the initial stage of the disease development and requires the participation of many specialists: Radiologists, Surgeons, Pathomorphology, Internal Medicine, Laryngologists, Pulmonologists. Good diagnosis in the early stage of disease is practically impossible.

Nevertheless, we hope that the presented case will allow you to remember that this primary vasculitis may also manifest its presence in macroscopic changes in the kidneys, which should be differentiated from kidney tumors.

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