

OPIS PRZYPADKU
CASE REPORT

CHURG-STRAUSS SYNDROME: CLINICAL CASE AND ITS FEATURES

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ABSTRACT

Introduction: The article describes the clinical case devoted to the features of the clinical course of Churg-Strauss syndrome (eosinophilic granulomatous with systemic necrotizing vasculitis). Churg-Strauss syndrome is a disease that is rarely diagnosed because of presenting by high-variability clinical syndromes, that causes difficulties in diagnostic process, especially on the first visit of patient to the general practitioner.

The aim of the article is to show the clinical course features of the Churg-Strauss syndrome in the time of disease progression, as well as peculiarities and possibilities of in-time diagnosis of this disease.

Clinical case: It was described a clinical case of Churg-Strauss syndrome in a 46-year-old woman, presenting with successively appeared signs of the disease that resulted in some difficulties of making in-time diagnosis.

Conclusions: As Churg-Strauss syndrome is a low-spread disease presenting with high-variety of clinical signs and despite of the presence of clear diagnostic criterias Churg-Strauss syndrome was diagnosed with delay in this clinical setting. General practitioner should remember that if the disease debuts by the signs of pneumonia with further appearance of signs of bronchial obstruction and eosinophilia, its necessary to perform diagnostic approaches with the aim to carry out the presence of the Churg-Strauss syndrome.

KEY WORDS: Churg-Strauss syndrome, diagnostic, treatment

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INTRODUCTION

Nowadays, Churg-Strauss syndrome is considered as an inflammatory-allergic lesion of small and medium vessels (capillaries, venules, arterioles), which leads to the formation of necrotizing granules. It has typical clinical manifestations in the form of bronchial asthma, eosinophilia and pulmonary infiltrates [1].

It should be noted that there are some difficulties in diagnosing this pathology in our time. There are both clinical situations as with hyperdiagnosis as with the presence of a long-lasting course of the disease, when for some reasons this syndrome remains not diagnosed in time [2,3].

Correct and in-time diagnosis of this disease is critically important to initiating and providing appropriate treatment. So it's necessary for practicing doctors to have relevant knowledges about Churg-Strauss syndrome and also to have some kind of alertness about the possibility of this disease.

From our point of view, described clinical case should be interesting and usefull, first of all, for general practitioners, as they predominantly have the first contact with patients suffering on Churg-Strauss syndrome. So, they should provide correct diagnostic approaches in a timely manner and forward patients to the next stage of medical care.

THE AIM

To describe the clinical case of Churg-Strauss syndrome, marking its clinical features, and emphasizing the attention of general practitioners to the possibility of early and timely diagnosis of this syndrome that is necessary for initiation of specific therapy.

CLINICAL CASE

Patient T., female, 1963 year of birth, having III-rd group of invalidity, considered herself as being ill since 2006 year when some complaints occurred for the first time. Patient suffered on dry cough, breathlessness during rigorous physical activity, elevated body temperature more than 37.5 C, general weakness, increased sweating, asthmatic attacks for up to 2 times / day (treated by taking 1 tablet of theofilline), pain-feeling in large joints, that increased during physical exertions.

After a first physical examination performed on out-patient settings, it was diagnosed «Untreated pneumonia», but later diagnosis was changed to «Chronic obstructive pulmonary disease with concomitant arthrosis, osteoarthritis and gonarthrosis». But as patient began to suffer on progressive shortness of breathing, predominantly in the morning hours, pulmonologist, changed the diagnosis to the «First occured bronchial asthma».

On 12th October 2016, chest MRI was performed. It showed slightly expressed fibrous lesions in the parame-diastinal divisions of S5 of both lungs, solid calcifications at the level of S6 of the right lung and at the S9 of the left lung. Changes in pulmonary tissue were considered as the consequences of chronic bronchitis. Lipomas of the anterior cardio-diaphragmatic angles and atherosclerotic aortic cardiosclerosis were also found.

As the disease had progressive course a repeated review by the doctor-pulmonologist was performed on 24.10.2016. Diagnosis of «Bronchial asthma of moderate severity, mixed chronicall form with controlled course» was established. So, patient was referred to specialized pulmonological department of the regional hospital on 24.11.2016. There was established diagnosis of «Persistent form of bronchial asthma of moderate severity with partially controlled course. Pneumosclerosis. Gastroesophageal reflux disease. Erosive gastropathy. Reflux-esophagitis. Chronic hepatitis, persistent form, exacerbation phase. Primary osteoarthritis, polyostearthritis with lesions of the lower leg, shoulder and knee joints».

From 10.12.2016 to 20.12.2016, patient received treatment at the district hospital having the diagnosis of «Left-sided pneumonia». During this hospitalization it was found an increase in the number of eosinophils in blood sample up to 64% and increased ESR 35 mm/h. So, patient was referred to the hematologist for diagnosis clarification.

At that time several laboratory and instrumental diagnostic investigations were performed. Blood sample (22.12.2016): leukocytes - $29.6 \cdot 10^9/l$, eosinophilic cells - 54%, ESR-45 mm/h. Myelogram (22.12.2016) showed increased eosinophilic granulocytes presence up to 63%. On ultrasound examination it was detected enlargement of the liver (16.7x3.4 cm). Chest X-ray investigation (22.12.2016) showed the signs of chronic bronchitis and pneumosclerosis. Endoscopy of the stomach estimated erythematous gastropathy and gastro-esophageal reflux with pH 1.0.

On 12.22.2016 hematologist established the diagnosis of «Leukemoid reaction of eosinophilic type».

The patient's condition had no signs of improvement by the time goes. As progressioun of arthritis syndrome occurred, patient was hospitalized to the rheumatologic department. Patient stayed there for more than 20 days, from 27.12.2016 till 18.01.2017. On 12.28.2016 MRI of the neck, thoracic cavity, abdominal cavity, retroperitoneal space, with contrast enhancement was performed. It found clinically significant enlargement of liver and spleen, pathological lesions in the spleen (one them with signs of calcification) and several liver cysts, But enlargement of lymphatic nodes within the scanned zones (neck, chest, abdomen) was not present.

On 12.30.2016 MRI of the brain was performed. It concluded the absence of signs of oncological, inflammatory and demyelinating processes of the brain. Cysts of the left maxillary sinus were found. Also MRI showed slightly expressed osteochondrosis, straightening of the cervical lordosis and right vertebral artery hypoplasia.

On 12.30.2016 MRI of the lumbar-sacral zone was performed. It was found the signs of diffuse explosions of intravertebral discs in segments L3-L4, L4-L5, L5-S1, numerous hemangiomas of the vertebrae, osteochondrosis in the lumbar zone, deforming spondylosis and spondyloarthritis.

On 30.12.2016 ultrasound examination was performed. Hepatomegaly splenomegaly, chronic cholecystitis and cysts of the spleen were detected

On 25.01.2017 patient was examined by hematologist. Leukemoid reaction of eosinophilic type was diagnosed. In blood cells analysis performed on 26.01.2017 number of eosinophilic cells was 9%.

After this examinations it was diagnosed «Deforming osteoarthritis with lesions of the left knee and left shoulder joints. Endoprosthesis of the right knee joint (2014). Leukemoid reaction of eosinophilic type. Gastroesophageal reflux disease with esophagitis. Chronic gastroduodenitis with increased acid-secretion function of the stomach. Fatty liver dystrophy. Non-alcoholic steatohepatitis of moderate activity. Chronic cholecystitis on stage of unstable remission. Osteochondrosis. Primary hypothyroidism, first detected. Secondary iron deficiency anemia of mild degree. Eosinophilia (28.12.2016 – eosinophilic cells - 72%, ESR 33 mm/h, 10.01.2017 – eosinophilic cells - 9%, ESR 24 mm/h).

At the same time the presence of Churg-Strauss syndrome was considered. So, patient was referred for an expert consultation to the National Scientific Centre «Institute Of The Cardiology named after M.D. Strazhesko». On 3 February 2017, after provided consultation by experts diagnosis was changed to «Systemic granulomatous vasculitis of Churg-Strauss, chronic disease course, II level of disease activity with lesions of the lungs vessels (a syndrome of bronchial asthma, with frequent attacks of suffocation (October-December 2016), allergic rhinitis, polyarthritis, blood eosinophilia (up to 72% in December 2016).

The prescribed treatment included 12 mg of Medrol per day for duration of 1.5 months with further reduction of dose on 1 mg every 2 weeks to the daily dose of 8 mg / day.

In 2017 the patient was examined and treated twice in the rheumatologic department of the regional hospital, and quarterly consulted at the National Scientific Centre «Institute Of The Cardiology named after M.D. Strazhesko». Due to continuous observation and obtained results of repeated diagnostic investigations it was diagnosed «Systemic granulomatous vasculitis of Churg-Strauss, chronic course, activity II, with lesions of the lungs' vessels (a syndrome of bronchial asthma, with frequent attacks of breathlessness (October-December 2016), allergic rhinitis, cardiac arrhythmia (myocardiofibrosis, insufficiency of the tricuspid valve of II degree, pulmonary hypertension of the II degree, chronic heart failure). Polyarthritis. Arthralgia associated with polyostearthritis and spondylarthritis with secondary radicular syndrome, left gonarthrosis. left metatarsophalangeal arthritis with recurrent synovitis. Right knee arthroplasty (2011). Hypereosinophilia (up to 72% - December 2016). Subclinical hypothyroidism, moderate severity, stage of unstable sub-

compensation. Discirculatory encephalopathy, II degree, with bilateral reflex-pyramidal insufficiency, vestibular dysfunction, persistent asthenoneurotic syndrome. Osteochondrosis with more predominant lesions in lumbar and sacral zones, deforming spondylosis, protrusions of L3-S1.

Patient received treatment with regularly titrated doses of Medrol.

During the first months of 2018, signs of the involvement of the kidneys to the pathological process have occurred and were associated with clinically significant progression of disease. Patient suffered on the pain in the lumbar region, frequent urination, puffiness of the face in the morning hours. It was found progressively increasing levels of protein in the urine samples (up to daily proteinuria - 0.450 g). At the same time blood pressure increased to 190/100 mmHg. Due to this findings it was concerned that patient had secondary nephropathy as a manifestation of kidney damage secondary to Churg-Strauss syndrome.

DISCUSSION

The Churg-Strauss syndrome was first described in 1951 as a triad of symptoms, which included hypereosinophilia, asthma and vasculitis, with asthma being the most typical sign of this syndrome.

The first descriptions by Churg and Strauss distinguished the high spectrum of clinical and pathological signs. Nowadays, it is concerned that the definition of this disease is too narrow, so, resulting in dramatically limitation of diagnostic capabilities [4,5].

In recent years the diagnostic criteria for Churg-Strauss syndrome were established. They include the presence of the following signs - asthma, the presence of more than 10% of eosinophils in the blood sample, mono- or polyneuropathy due to systemic vasculitis, infiltration of the pulmonary tissue, anomalies of the nasal sinuses, extravascular eosinophils accumulation evaluated in samples obtained by tissue biopsy. By performing histological analysis it is usually observed necrotizing vasculitis of small vessels with inflammatory infiltration by eosinophils accompanied with necrotizing granulomas.

Anti-neurotrophic cytoplasmic antibodies (ANCA) can be detected in 40% of patients with Churg-Strauss syndrome. So, we can suggest the existence of 2 types of Charge-Strauss syndrome: ANCA-positive and ANCA-negative. ANCA-positive patients usually have renal impairment, neuropathy, alveolar hemorrhage and purulent vasculitis. ANCA-negative patients have predominantly lesions of heart and lungs. However, these two types are impossible to differentiate in many cases. Also, there is no evidence that these two types may have a different prognosis. Cerebrovascular complications (chronic brain ischemia and stroke) are on the first place among the other causes of death in patients suffering on Churg-Strauss syndrome [6,7]. Non-specific signs may also include such manifestations as fever, general feeling of weakness and fatigue, loss of appetite, weight loss and

muscle pain. Peripheral nerves, kidneys and gastrointestinal tract may be also affected.

The average age of the disease debut is 38-49 years of old [8].

It is important to notice that clinical symptoms are so non-specific in some clinical cases that simulate other pathological conditions and diseases..

Clinical course of Churg-Strauss syndrome is divided on three consecutive stages – prodromal stage, eosinophilic stage and vasculitis.

The prodromal stage, lasting from 6 months to two decades, may precede the development of the vasculitis and is characterized by a variety of allergic manifestations. Usually patients have signs of asthma with frequent cough, dry wheezing and shortness of breath (as described in this clinical case).

The eosinophilic phase is characterized by the accumulation of eosinophils in various tissues of the body as the consequence of continuous hypersynthesis of eosinophils with their infiltration of different tissues (predominantly, the lungs, the gastrointestinal tract, and the skin).

The third phase is vasculitis, that manifests as inflammation of various blood vessels and leads to the occurrence of cardiovascular disorders and to the development of aneurysms. So, the risk of bleeding is high.

In this clinical case, vasculitis manifested as involvement of renal vascular system (glomerulus) that resulted in secondary nephropathy with malignant arterial hypertension syndrome.

At the same time, we are sure that timely prescribed therapy prevented the development of severe complications of the Churg-Strauss syndrome and normalized the number of eosinophils in the blood.

Primary prevention for this disease is not established yet. Secondary prophylaxis consists of providing adequate supportive therapy and long-term observation [9,10].

CONCLUSIONS

Due to the low-prevalence of this disease in general population and due to the variety of clinical signs, even despite the existence of clear diagnostic criteria, the true diagnosis for this patient was established with significant delay. General practitioner should be aware that if disease is beginning like pneumonia with symptoms of bronchial obstruction and hypereosinophilia, it's important to provide diagnostic examinations to carry out the presence of Churg-Strauss syndrome.

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