UDC 616 – 005.2

POLYCYTHEMIA VERA AN EXAMPLE OF A CLINICAL CASE

Tomina E. E.[1], Kamenskaya E. P.[2], Lebedynets P. V.[1], Tomakh V. V.[1]
[1] V. N. Karazin Kharkiv National University, Kharkiv, Ukraine
[2] STPI "Central clinical hospital «Ukrainian railway», Kharkiv, Ukraine

The article concerns a clinical case management of patients with polycythemia vera. The data are given from the laboratory and instrumental diagnostic methods, clinical diagnosis, selection of the optimized treatment and modification of the habit of life.

KEY WORDS: polycythemia vera, secondary arterial

ІСТИННА ПОЛІЦИТЕМІЯ НА ПРИКЛАДІ КЛІНІЧНОГО ВИПАДКУ

Томіна О. Є.[1], Каменська Є. П.[2], Лебединець П. В.[1], Томах В. В.[1]
[1] Харьковскій національний університет імені. В.Н.Каразіна, м. Харків, Україна
[2] ДЛПЗ Центральна клінічна лікарня «Укрзалізниці», м. Харків, Україна

Розглянуто клінічний випадок ведення пацієнта з істинною поліцитемією. Наведені дані лабораторних та інструментальних методів дослідження, описана діагностика, постановка клінічного діагнозу, вибір оптимальної тактики лікування та модифікація способу життя.

КЛЮЧОВІ СЛОВА: істинна поліцитемія, вторинна артеріальна гіпертензія

ИСТИННАЯ ПОЛИЦИТЕМИЯ НА ПРИМЕРЕ КЛИНИЧЕСКОГО СЛУЧАЯ

Томина Е. Е.[1], Каменская Э. П.[2], Лебединец П. В.[1], Томах В. В.[1]
[1]Харьковский национальный университет имени В.Н. Каразина, г. Харьков, Украина
[2] ГЛПУ Центральная клиническая больница «Укрзалізниці», г. Харьков, Украина

Рассмотрен клинический случай ведения пациента с истинной полицитемией. Приведены данные лабораторных и инструментальных методов исследования, описана диагностика, постановка клинического диагноза, выбор оптимальной тактики лечения и модификация образа жизни.

КЛЮЧЕВЫЕ СЛОВА: истинная полицитемия, вторичная артериальная гипертензия

INTRODUCTION

Polycythemia vera or erythremia, Vaquez disease – is a chronic neoplastic myeloproliferative disorder with bone marrow involvement on the cell level, precursor of myelopoesis, characterized by the unbounded proliferation of all three hematopoietic lineages, which predominantly are erythroid and megakaryocytic, granulocytic in a smaller extent [1, 2].

The disease is quite rare: 0,5-1,7 cases per 100,000 in full set population [3].

The main difficulty and danger pose that the disease is most commonly developed over the years (an average of 15 –20years) with a gradual increase of symptoms, that all this time has not been specific to such a disease. This leads to the late diagnosis and treatment, which results in the reduced quality of life of the patient. That poses a danger of the squeal occurrence, such as: myocardial infarction, the stroke, the syndrome of the disseminated intravascular coagulation and others [1-3].

Thus, such factors necessitate early detection of polycythemia vera cases and their forehanded treatment. This article focuses on one of such cases.

CLINICAL CASE

The patient I., a man born in 1983, was admitted to the CCH UZ cardiology department in October, 2013 with complaints about general weakness, periodic headache of pressing nature, often perceiving in the temporal regions and accompanied by noise in

the ears; dizziness accompanied by the increased blood pressure; pains in chest of compressive, heating character without irradiation and dependence on any physical activity, compensated spontaneously after 5-10 minutes of its occurrence.

HISTORY OF DISEASE

Such complaints for the first time occurred in the spring of 2013. In October 2013 at a time when emerged intense headache, it had been registered the increasing numbers of the blood pressure to 180/110 mm Hg , that was the reason why he had been hospitalized in one of the city hospitals. Received therapy: magnesia, Dibazolum tablets without any positive effect. By agreement, the patient was transferred to Ukrainian railway central clinical hospital (CCH UZ) cardiology department for further examination and treatment. Additionally cases of nosebleeds on a background of normal blood pressure were diagnosed.

ANAMNESIS VITAE

Leads a healthy lifestyle, patient does not smoke or drink, getting enough nutrition, has good living conditions. Grew and developed according to the sex and age. There are no injuries and surgical operations occurred in the past. In the childhood suffered the chickenpox. Hereditary and allergic anamneses are not burdened. Viral hepatitis, tuberculosis, venereal diseases, diabetes mellitus, HIV are absent.

PHYSICAL EXAMINATION

General condition is satisfactory, conscience is clear, position is active. The body type is normosthenic. Skin is clean, normal colored, without any scars, traces of scratching and venous lakes, moderate hyperemia of the cheeks is observed, visible mucous membranes of pale pink color, derivatives of the skin are without any visible changes. There is no edema. Subcutaneous fat tissue is developed satisfactorily, uniformly, respectively to the sex and age. Peripheral lymph nodes are not enlarged, with normal consistency. Thyroid gland isthmus is palpable. Musculoskeletal system is without any singularities. Auscultation over the lungs is clear, vesicular breathing. The heart rhythmical sounds are clear, heart rate - 67 beats/min, blood pressure - 150 /90 mm Hg. Abdomen is normally sized, soft and painless. Liver and spleen remain impalpable. Tapping symptom is negative on both sides.

REFERRAL DIAGNOSIS

Somatoform autonomic dysfunction

RESULTS OF LABORATORY AND INSTRUMENTAL DIAGNOSIS

All the represented testing was conducted on the basis of CCH UZ in October, 2013.

Complete blood count: polychromemia 179 g/L, erythremia 5.67 x 10^{12}/L, increased hematocrit 51.1, other indices are within normal limits.

Biochemical analysis of blood: imperceptible hypercholesterolemia 5.62 mmol/L, other indices within normal limits.

Electrocardiography (ECG) showed regular sinus rhythm, heart rate (HR) 73 beats per minute, axis deviation to the left.

Veloergometry: the total amount of work done 5508 kgf (54.02 kJ), the power load maximum proposed 150 V, complaints during the test of general fatigue, the reason for stopping is the achievement of submaximal heart rate , during the trial and recovery period the ECG did not show any coronary insufficiency. The test is negative.

Holter ECG and blood pressure monitoring: monitoring showed regular sinus rhythm with average HR 73 beats per minute, paroxysmal arrhythmias and any ischemic changes are not diagnosed. During the day and the night indices of systolic blood pressure and diastolic blood pressure are typical for normotension. Average BP is 108/64 mmHg.

Echocardiography: abnormal chord were diagnosed in the left ventricular, myocardial hypertrophy of the left ventricular, no akinesia zones have been identified, the indicators of myocardial contractility of the left ventricle are saved.

Ultrasound of the kidneys, adrenal glands and bladder: increased blood flow velocity in distal thirds of the renal arteries.

Check ultrasound of the renal arteries showed no clear data of the significant dopplerographic hemodynamic narrowing of the renal arteries on both sides.

Ultrasound of the thyroid gland: increased echogenicity, diffuse changes in the thyroid gland.

Ultrasonography of the abdomen: diagnosed liver and pancreas diffuse changes.

Consultation of neurologist: Dysfunction of autonomic nervous system.

Has been carried the brain magnetic resonance imaging (MRI): MRI of the brain did not diagnose any organic pathology.

RECOMMENDATIONS FOR FURTHER EXAMINATION

Complete blood count with the diagnosis of serum erythropoietin (in favor of polycythemia vera there would be indicated the plethora on the background of normal or even low concentration of serum erythropoietin).

Determination of blood oxygen saturation for diagnosing with secondary erythrocytosis (in favor of polycythemia vera there would be indicated normal blood oxygen saturation - more than 92 %).

Determining whether a mutation V617F persists in the gene JAK2, which is etiological factor of polycythemia vera. Consultation with the hematologist to clarify the diagnosis [4].

Lipidogram.

Thyroid panel determination of TSH, T3,T4 [5, 6].

BASIC CLINICAL SYNDROMES

- Pletorhycal: erythrocythaemia, arterial hypertension, headaches.
- Hyper blood viscosity with the microcirculation disorders: anginal aches and headaches.

CLINICAL DIAGNOSIS

First stage of polycythemia vera: of a few symptoms up to 5 years [2, 3].

Secondary arterial hypertension, II stage - with the presence of objective evidence of target organ damage without symptoms on their part or dysfunction - left ventricular hypertrophy, moderate 3 degree hypertension with systolic figures of blood pressure between 160 and 179 mm Hg, heart failure is absent [7].

CASE MANAGEMENT

At one of the hospital's department, before the establishing diagnosis, the patient has been treated as follows:

1. Noliprel (perindopril arginine + indapamide) tab. 1. morning 5/1.25mg.
2. Pentilin (pentoxifylline) 5.0 ml / in cap.
3. Eufillin 5.0 ml of 2 % + 50 % Analginum 2.0 ml / cap.
4. Noobut (Phenibutum) 250mg, 1 tab. 2 times a day.
5. Lysine aescinat 0.1 % 5.0 ml / intven. drip.
6. Relaxil (hydroalcoholic extract of dry valerian 0.8 % + dry mint peppermint + Melissa extract dry) 1 capsule (0.125 / 0.025 / 0.025) 2 t / d.
7. Psychotherapy.
8. Therapeutic exercise.
9. Physiotherapy.

RECOMMENDATIONS FOR TREATMENT AFTER DIAGNOSIS

1. Bloodletting (phlebotomy): effusion of 200-300 ml, every day or every second day to achieve the target hematocrit level - 0.42-0.45.
2. At run time of phlebotomy - taking aspirin (100 mg / day).
3. Angiotensin converting enzyme inhibitor (perindopril) 2 mg/day under the control of blood pressure (at home), after 2 weeks-checking visit to correct the dose.

Due to a significance of combination of various risk factors on the development of disease, a term risk factors has been accepted for general use instead of a singular risk factor term. To evaluate the effect of risk factors on the prognosis, the course, and the outcomes of the disease, it is important to introduce a global index as an integral measure [8].

RECOMMENDATIONS FOR LIFESTYLE MODIFICATION

Bring in the diet following principles of sensible nutrition:

1. Reduce intake of fats by one-third, mainly due to saturated ones.
2. Increase mono- and polyunsaturated fatty acids in nutrition.
3. Increase consumption of fruits and vegetables, rich in potassium and magnesium.
4. Limit the usage of salt.

Aerobic exercise of moderate intensity: jogging, swimming, cross-country skiing, biking.

PROGNOSIS

Polycythemia vera is a chronic disease, hence the prognosis relatively recovery is poor. But considering the detection of the disease at early stages, as well as previously initiated treatment, the prognosis for the patient's life is favorable. It should be mentioned that without any treatment mortality within 18 months after the diagnosis of the disease is about 50 %. With adequate and timely therapy the median

survival exceeds 10 years, the patients of young and middle-age – several decades.

The most common cause of death is thrombosis, the second by importance - complication of myeloid metaplasia with the transition of the disease in leukemia, therefore the further case of the patient is to be directed concerning these two aspects. For the first it is disaggregated control therapy of thrombosis and it's further consequences. For the second case, unfortunately, at this stage there is taken no precaution, though timely started treatment will provide a benign clinical course of the disease [1-4].

REFERENCES

1. Vorobyov A.I. Guide to Hematology / A.I. Vorobyov. – M.: Newdiamed, 2003. – 247 p.
2. Spivak Jerry L. Polycythemia vera: myths, mechanisms, and management / Jerry L. Spivak // Blood. – 2002. – № 100 (13). – P. 4272-4290.
3. Mamayev N.N. Hematology / N.N. Mamayev. – M.: SpecLit, 2008. – 558 p.
4. Girodon F. Presence of Calreticulin Mutations in JAK2-Negative Polycythemia Vera / F. Girodon, J. Broseus, J.-H Park-Alexandre et.al.// Blood. – 2014. – № 124 (21). – P. 1819.
5. Ayalew T. Polycythemia vera and essential thrombocythemia: 2013 update on diagnosis, risk-stratification, and management // T. Ayalew. – American Journal of Hematology. – 2013. – №88. – P. 507-516.
6. Passamonti F. How I treat polycythemia vera / F. Passamonti // Blood. – 2012. – № 120(2). – P. 275-285.
7. ESH/ESC Guidelines for the management of arterial hypertension: The Task Force for the management of arterial hypertension of the European Society of Hypertension (ESH) and of the European Society of Cardiology (ESC) // Journal of Hypertension. – 2013. – Vol. 31, Is. 7. – P. 1281-1357.
8. Yabluchanskiy M. I. Internal Diseases: The Time Of Global Somatic Risk / M. I. Yabluchanskiy, A. M. Yabluchanskiy, O. Y. Bychkova et.al. // The Journal of Kharkiv V. N. Karazin` National University, Series «Medicine». – 2013. - № 1044 (25). – P.5-7.