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CASE OF GUILLAIN—BARRÉ SYNDROME IN A PATIENT WITH PULMONARY LANGERHANS CELL HYSTIOCYTOSIS

Abstract

Langerhans cell histiocytosis is a rare disease characterized by various clinical patterns: from isolated lung lesions to severe involvement of other organs. This clinical case demonstrates a rare combination of pulmonary Langerhans cell histiocytosis and Guillain—Barré syndrome due to possible common mechanisms of the disease development mediated by the CD1A expression.

Key words: *Langerhans cell histiocytosis, Guillain—Barré syndrome, clinical case*

Conflict of Interests

The authors declare no conflict of interests.

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LV — left ventricle, GBS — Guillain—Barré syndrome, PCR — polymerase chain reaction

Introduction

Guillain—Barré syndrome (GBS) (ICD 10 code is G 61.0) is an autoimmune peripheral neuropathy, which is the most common cause of acute tetraparesis. In the Russian Federation, the incidence corresponds to global data and is on average 1.8 per 100,000 per year [1].

Most authors agree that this refers to post-infectious autoimmune peripheral neuropathy. Because of molecular mimicry of lipopolysaccharide

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structures (ganglioside antigens) of microorganisms and nerve cells, antibody- and cell-mediated immunity is triggered, and the latter plays a key role in GBS [2]. CD1 molecules specialize in the capture and presentation of glycoproteins by T-cells. Meanwhile, CD1a+/CD207+ expression is a key histopathological finding typical for Langerhans cell histiocytosis, a disease characterized by proliferation of CD1a+ dendritic cells with local or multiple organ lesions. CD1 gene polymorphism can influence predisposition to the development of Guillain—Barré syndrome, which is widely discussed [3, 4].

Meanwhile, we found no clinical cases of a combination of pulmonary histiocytosis and Guillain—Barré syndrome in the PubMed database using the keywords "Langerhans cell histiocytosis AND Guillain—Barré syndrome".

Case Report

Lung pathology was revealed in patient G., 56 years old, during regular health check-up. For its clarification, we performed chest computer tomography on 3.04.15, and disseminated lung disease of unknown etiology was diagnosed based on the CT results. From 14.04 to 02.05.2015, the patient was in the Ulyanovsk Regional Clinical TB Dispensary, where video-assisted thoracic surgery (VATS) with biopsy of the right lung was performed on 17.04.2015. In the initial study of the lung biopsy, evidence of histiocytosis was obtained: "Lung tissue with granulomatous pattern of inflammation with signs of vasculitis and bronchiolitis with areas of eosinophilic infiltration, with formation of foci of desquamative interstitial pneumonia, with formation of foci of fibrosis with deposition of coal pigment." No signs of tumor growth and specific inflammation were found. To verify the diagnosis, the sample was sent to the Republican Clinical Oncologic Dispensary of the Ministry of Health of the Republic of Tatarstan. The following conclusion was made: CD1A, S100 positive reaction in cell clusters. CD68 slightly positive reaction. Langerhans cell proliferative activity index (ki67) is 8%. Conclusion: Langerhans cell histiocytosis. Based on the obtained data, Langerhans cell histiocytosis with isolated lung lesion was diagnosed.

The patient has hypertension for a long time; antihypertensive drugs are taken irregularly. There are no cardiovascular diseases in family history. He denies having bad habits, including smoking. Negative PCR (polymerase chain reaction) results on Epstein—Barr virus DNA were obtained.

Since 26.04.15, he has noted numbness in fingers, weakness in the legs. Due to increasing weakness and deterioration of sensitivity in the extremities brain, computed tomography was performed on 29.04.15, and moderate hydrocephalus was revealed. The condition worsened: Numbness of lips and dysarthria occurred. On May 2, 2015, the patient was transferred to the Neurological Department of the Central City Clinical Hospital of Ulyanovsk. During examination in the Neurological Department, consciousness was preserved, the patient was in a forced supine position. Body temperature was 36.9 °C. Body mass index was 28.7 kg/m^2. The skin and visible mucous membranes were clear and of normal color. Peripheral lymph nodes were not enlarged. Vesicular breathing over the lungs, and there were no crackles. Respiratory rate was 19 per minute. Heart tones were muted, heart rate — 104 per minute, blood pressure — 140 and 100 mm Hg on both hands. The abdomen during palpation was soft, painless; the lower edge of the liver was 2–2.5 cm below the costal margin. There was no peripheral edema.

Neurological status: Adequate. No meningeal symptoms. Convergent strabismus OU. Diplopia is stopped with monocular vision. Bilateral weakness of facial muscles up to 2.0 points. Lagophthalmos on the left side. Eyelash sign is positive on the right side. Pharyngeal reflexes are reduced. There are some difficulties with swallowing. Dysarthria. Dysphonia. Dysphagia. Strength in the upper extremities is reduced up to 2.0 points, in the feet — up to 0.5 points, and there is plegia in the hips with diffuse hypotonia. Conductive hypesthesia from the level of Th2–Th3 and down, more pronounced in the distal parts of stocking and glove type. Tendon areflexia. Plantar reflexes are not triggered. There are no pathological plantar reflexes.

Complete blood count (04.05.15): hemoglobin — 170 g/L, RBC — 5.1×10^{12}/L, WBC — 19.0×10^{9}/L,

ESR 5 mm/h. Blood chemistry (02.05.15): Creatinine — 72.4 mmol/L, alanine aminotransferase — 86 IU/L, total protein — 71 g/L, albumin — 41.21 g/L, total bilirubin — 8.2 mmol/L, glucose — 8.51 mmol/L, cholesterol — 4.52 mmol/L, potassium — 3.79 mmol/L. Prothrombin index — 88%, activated partial thromboplastin time — 30 s, international normalized ratio — 1.1. Glycemic profile (09.05.15): glucose — 6.7–9.7–9.2–6.3 mmol/L. Cerebrospinal fluid was not examined.

Electrocardiography (02.05.15): Sinus rhythm with a heart rate of 104 bpm. PQ — 160 ms. QRS — 90 ms. QTc — 414 ms.

Echocardiography (04.05.15): concentric left ventricular (LV) hypertrophy (LV myocardial mass index — 126 g/m^2). Systolic LV function is preserved, ejection fraction is 53%. Diastolic LV dysfunction of impaired relaxation pattern.

Chest radiography (04.05.15): A chain of metal clips at the level of the third intercostal space of the right lung. Pulmonary pattern is enhanced in the lower basal regions of the right lung. Sinuses are clear. The mediastinum is not displaced.

Ultrasound of the kidneys and bladder (20.05.15): Coral calculus in the left kidney, 18-mm cystic calculus.

Based on patient's complaints, history, examination data, typical neurological status, laboratory and instrumental data, which can exclude focal pathology of the brain, acute demyelinating polyneuropathy (Guillain—Barré syndrome) was diagnosed. Bulbar palsy. Peripheral tetraparesis. The syndrome of oculomotor disturbances.

Hypertension of the 2nd stage, achieved the 1st degree of HT, risk 4. Left ventricular hypertrophy. Urolithiasis. Coral calculus of the left kidney. Cystic calculus. Hyperglycemia.

In the Neurological Department, treatment was performed, including plasmapheresis, intravenous immunoglobulin, and systemic corticosteroids (pulse therapy with metipred 1,000 mg due to histiocytosis) and oral metipred 40 mg, thioctic acid (Berlithion), anticholinesterases (neostigmine, ipidacrine), B vitamins, and antihypertensive drugs (amlodipine, bisoprolol).

The patient's condition stabilized within 4 days after transfer to the Neurological Department: strength in the hands began to grow, range of motions increased, swallowing restored, strength in the hands increased up to 4.0 points, in the hips — up to 3.0 points, and in the feet — up to 4.0 points. On 20.05.15, the patient was discharged with improvement, and considering high rehabilitation potential, he was referred to the Rehabilitation Department.

It is recommended to take corticosteroids with a gradual daily dose reduction to 8 mg of metipred, thioctic acid, B vitamins, and anticholinesterases.

When the patient was contacted by telephone after 3 years, it was found that he feels himself satisfactory, disturbance of sensitivity in the fingers of the lower extremities remained, and there were no complaints related to the respiratory system. He is socially adapted, works, and performs daily physical activity (running). He is under neurologic follow-up and receives annual health resort treatment. Recommended antihypertensive drugs, unfortunately, are taken irregularly.

Discussion

In recent years, the etiology and pathogenesis of histiocytosis have been actively discussed. In addition to infectious histiocytosis, the malignant nature of the disease has been discussed [5]. The favorable course of the disease within 4-year follow-up of the patient, in our opinion, gives evidence against the oncological etiology. On the other hand, no obvious infection was detected while the patient was in the TB dispensary.

It seems to us that in tour patient activation of CD1A typical for Langerhans cell histiocytosis, regardless of the etiological cause, triggered the immune mechanism of the development of autoimmune peripheral neuropathy (Guillain—Barré syndrome). Anti-ganglioside antibodies play a key

role in the development of histiocytosis, and they are also found in the serum of patients with autoimmune neuropathy — ganglioside complexes play an important role in the pathogenesis of Guillain—Barré syndrome [2].

Damage to the central nervous system in histiocytosis is extremely rare and is an inflammatory process resembling paraneoplastic encephalitis [6]. As a rule, pulmonary histiocytosis is isolated and is not accompanied by a multi-organ lesion [5]; such patients are not classified as high-risk according to the prognosis [1, 7]. However, the presented case suggests that even when mono-organ Langerhans cell histiocytosis exists severe life-threatening complications, such as Guillain—Barré syndrome may develop.

Conclusion

This clinical case is of interest from the perspective of possible general mechanisms of both Langerhans cell histiocytosis and Guillain—Barré syndrome. In addition, the presented case shows that patients with isolated pulmonary Langerhans cell histiocytosis require special attention due to the possible development of multi-organ lesions and the development of life-threatening complications, including Guillain—Barré syndrome.

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