Rosai-Dorfman Disease – A Case Report

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Case report

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Abstract

Background: The aim is to discuss a case of a meningeal form of Rosai-Dorfman disease, is a rare haematological disease, primarily affecting lymph nodes, but it can also affect most organ systems of the human body, including the central nervous system. So far, there have been over 650 cases of this disease described in literature. **Case presentation:** This report deals with a young, 40-year-old man with a negative history of disease, who observed a loss of smell and taste, without a history of anxiety or an injury. The patient was hospitalized and completely examined. Consequently, a navigational biopsy of the brain was indicated. The findings showed that the patient suffered from Rosai-Dorfman disease with reactive gliosis in adjacent tissue without evidence of amyloid. **Conclusions:** In January, March, and then in May 2018, the patient was checked for the magnetic resonance imaging of the brain, according to which there was a significant regression of the finding, and according to the outpatient reports of the same year, the taste and vision of the left eye are gradually being adjusted. The patient was treated with new corticosteroids, which proved to be effective.

Background

Rosai-Dorfman disease is a rare haematological disease, primarily affecting lymph nodes, but it can also affect most organ systems of the human body, including the central nervous system. The first record of this disease dates back to 1969 when pathologists Juan Rosai and Ronald Dorfman first mentioned it in their study. Since then, more than 650 cases of Rosai-Dorfman disease have been described in the literature. The aim of this report is to discuss a case of a meningeal form of Rosai-Dorfman disease.

Case Presentation

This case report deals with a young 40-year-old man of Slovak nationality living with his family in the Czech Republic and working in a car factory as a welder. So far, he has not been cured for anything and he has not taken any medication. The family history from the neurological point of view includes his father's epilepsy. He has not smoked for 10 years, however, until then he managed to smoke 30 cigarettes a day.

Since May 2017, he has observed a loss of smell and taste, without a history of anxiety or an injury, supposedly to be preceded by a celebration with a bigger amount of alcohol with subsequent nausea and elevated temperatures. In addition, in mid-October, he began to observe the impaired vision of the left eye (the nature of the visual acuity deterioration, the glitter feeling and as if he was looking into the sun), and on the basis of an ocular examination, a reduction in visual acuity of the left eye, which also showed absolute scotoma.

He was hospitalized in the Department of Neurology of the University Hospital of Hradec Kralove (UHHK) in November last year after an outpatient computed tomography (CT) of the brain, where extensive areas of vasogenic edema were described in both frontal and right temporal lobe. When receiving an entry

neurological examination, he did not show signs of cerebral infarction, he was only slightly euphoric. No significant pathological values were found in laboratory samples, including blood count, and a normal record was recorded on the Electro Encephalo Gram (EEG). Supported magnetic resonance imaging confirmed multifocal opacifying deposits in the locations corresponding to the CT scan (consult Fig. 1 below). Differential diagnosis remained wide, among which the possibility of multiform glioblastoma was not excluded.

New corticosteroids were introduced into the therapy and, in agreement with the neurosurgeon, the patient was transferred in a clinically unchanged condition to the Neurosurgery Clinic in the University Hospital of Hradec Králové to complete the open biopsy as the last diagnostic option. According to the neuronavigation examination prior to the medical performance, there was, among other things, a partial regression of the vasogenic edema and the postcontrast opacifying bearings, which can be attributed to the newly applied corticotherapy. It was followed by the performance (open biopsy), which was done without complications, and the samples were subsequently sent for histological verification. After several days, it was concluded that histologically it was a meningeal form of Rosai-Dorfman disease with reactive gliosis in adjacent tissue without evidence of amyloid.

After consultation with the hematologist, a CT examination of the body with a finding of borderline nodes in the retroperitoneum and mild splenomegaly and bone marrow biopsy with the conclusion of hypocellular trilinear hematopoiesis with reactive changes, without significant dysplastic alterations, without convincing tumor structures. From the pathologist's point of view, it is probably histiocytic reactive reaction.

Discussion And Conclusions

General characteristics

Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, is a rare disease characterized by proliferation and accumulation of a specific type of leukocytes (histiocytes) predominantly in lymph nodes, but it can also accumulate in other localizations throughout the body [1]. This disease affects both men and women (although some sources report higher incidence in men). Patients are most likely to be children, adolescents and young adults under the age of 20, but the possibility of occurrence in all age groups is not excluded (as it has been confirmed by this case report) [2-3].

Etiology

The exact cause of the disease is unknown (= idiopathic); the most likely cause is infection (Klebsiella, polyomaviruses, Epstein-Barr viruses, parvovirus B19, human herpesvirus 6); other factors involved in immunodeficiency, autoimmune diseases, environment. Inflammatory activated monocytes can produce colony-stimulating factor for macrophages, resulting in complex signal transduction, which subsequently leads to the histopathology characteristic of Rosai-Dorfman disease [4-5].

Clinical picture

Symptoms are very variable and individual, depending inter alia on the affected organ. The disease can be asymptomatic, with only lymph nodes (= lymphadenopathy - usually cervical, which is in more than 78%, axillary, inguinal, mediastinal, ...), both lymph nodes and extranodal tissues, or only organ (16%), nasal and paranasal cavities (11%), eyes / eyelids / orbit (11%), bones (9%), salivary glands (isolated form) (5%) and CNS (7%), in rare cases kidney (2%), respiratory tract or liver [3]. The most common symptoms include, in addition to painless enlargement of the lymph nodes, overall symptoms such as fever, paleness, weight loss, malaise and chronic rhinitis, in rare cases hepato/splenomegaly [2,4].

Diagnostics

Due to the wider differential diagnosis, which includes both malignant and non-malignant diseases, the most important diagnostic method is biopsy of the affected tissue with microscopy and immunohistochemistry with S100, CD68, CD163, CD14 and MAC-387 positivity and CD1, CD21, Mr B and Mr T with histiocyte negativity [5-6].

Treatment

First, it is important to emphasize that treatment is strictly individual. The decision on treatment is difficult due to the different forms, course and degree of the disease. In some cases, the symptoms resolve over months or years (spontaneous remission in 70-80% of cases) and therefore clinical observation without treatment is often recommended (if possible). In some cases, treatment is important [4]. Therapeutic options include treatment for specific symptoms - surgical (lymph nodes, central nervous system lesions, localized skin lesions), pharmacological (steroids, alpha-interferon, chemotherapy) and radiotherapy [2]. The basic chemotherapeutics to treat Rosai-Dorfman disease include vinblastine, 6-mercaptopurine, methotrexate or thalidomide and classical rule applies here - take the lowest possible dose of the chemotherapeutic agent so as to keep the disease under control while ensuring a good quality of life [5,7]. In the case of increased immunoglobulins, plasmapheresis is another therapeutic option. Furthermore, the symptomatic treatment in general is also important. Rosai-Dorfman disease does not usually pose a threat to life-threatening illnesses, or there is usually no significant organ failure, and the prognosis is good despite the damage to the tissue (but worsening with the number of affected nodes) [5].

In January, March, and then in May 2018, the patient was checked for the MR of the brain, according to which there was a significant regression of the finding, and according to the outpatient reports of the same year, the taste and vision of the left eye are gradually being adjusted. Therefore, it was recommended to continue in the corticotherapy as a prevention of the relapse, as well as in patient dispensarisation.

List Of Abbreviations

CT – Computed Tomography

EEG – Electro Encephalo Gram

MRI – Magnetic Resonance Imaging

UHHK – University Hospital of Hradec Kralove

Declarations

Ethics Approval and Consent to Participate

The Ethics Committee of the University Hospital in Hradec Kralove does not give consent to case reports because it is provided in the informed consent of the patient on his/her admission to hospital. The informed consent to participate in the study was obtained from the participant and it is available on request.

Consent for publication

The patient provided written informed consent for the publication and the use of his images.

Availability of data and materials

The data that support the findings of this study are available from the University Hospital of Hradec Kralove, but restrictions apply to the availability of these data, so are not publicly available. The data are however available from the authors upon reasonable request.

Competing interests

The authors declare that they have no competing interests to declare.

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Not applicable.

Author's contribution

OM, PK, MB, PK, BK, and MV equally contributed to the drafting, analyses and final version of the whole manuscript. All authors read and approved the final manuscript.

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Figures

Figure 1

CT of the Native Brain - Large areas of indicated vasogenic edema in both frontal and right temporal lobe and MRI of the brain - extensive hyperintense non-expanding deposits side-by-side symmetrical bilaterally (including the bublus olfactorius area) subcortical vasogenic edema affecting the subkalous gyrus and marginally extending into the genus of the caudal body and chiasm more left, similar temporal right (without hippocampus)

Supplementary Files

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