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Case Presentation

Castelman Disease Plasma Cell Variant Case Report

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Abstract

Castelman disease is also known as non-malignant vascular nodular hyperplasia or giant lymph nodes. Clinically the localized and multifocal form is distinguished. The authors present the case of a 34-year-old patient diagnosed with Castelman tumor within the abdominal cavity. The patient was treated surgically. In the two-year follow-up, there was no recurrence of the proliferative process. This article presents current recommendations for diagnostic procedures and treatment possibilities.

Keywords: Castelman disease; Lymphadenopathy; Lymphoma

Introduction

Castleman disease-rare clinical entity, characterized by non-cancerous cell proliferation associated lymph proliferative systems, is often connected with HHV-8 or HIV infection [1].

Existing synonyms, found in literature, include inter alia benign giant lymphoma and hamartoma of lymph nodes.

Deferential diagnosis includes Hodgkin's lymphoma, Kaposi's sarcoma, Non-Hodgkin's lymphoma, POEMS syndrome. Therapy selection depends on the clinical type. Treatment methods of CD include surgery, chemotherapy, radiotherapy or biological drugs [2].

Case Presentation

We present a case of a 34-year-old female patient diagnosed at the Maria Skłodowska Curie Institute of Oncology in 2015 due to mild anemia-stage G1, increased inflammatory parameters and nonspecific pain in the abdominal cavity. Deviations in laboratory tests and subjective symptoms were observed in patient examination for about a year.

As a result of diagnostic imaging including CT of the neck, chest, abdomen and pelvis, pathological mass was found in the vicinity of the lesser curvature of the stomach and near the right branch of the diaphragm.

The radiological picture was ambiguous - to the differentiation of ganglioneuroma *vs.* PNET. Exclusive viral background of lesions (Hepatitis B and C and HIV). Cancer markers, including chromogranin in control assays, were negative. Finally, the patient was qualified for surgery.

The patient was operated outside our Centre, in the histopathological material collected by the laparotomy; the lymph node was transformed with the stimulation of the reproductive centres.

Due to unambiguous results of histopathological examination, the collected material was subjected to path morphological reassessment; the patient was diagnosed with an early phase of cell-associated variant of Castleman's disease-The patient was left for close observation, in two years observation until now without any ailments. Normalization of inflammatory and morphological parameters was observed.

Discussion

First defined in 1956 by Benjamin Castelman, the localised and multifocal form of CD is clinically distinguishable [3].

The incidence of this disease in the United States is estimated at 30,000 to 100,000 per year, while no data is available for Europe, including Poland. For this reason, Castleman's disease is rarely considered in differential diagnosis [4].

Histopathologically, we distinguish vas-schizophrenic, plasma-cell and mixed form. In pathogenesis, a large role is attributed to the overproduction of pro-inflammatory cytokine-IL-6, which is also responsible for the systemic symptoms in this disease. Due to the unspecific microscopic image, cooperation between a clinician and a pathomorphologist is extremely important [5].

We can distinguish between large and small diagnosis criteria for Castleman's disease. The former include characteristic histopathological weave (in the case of our patient, it was an increased number of plasma cells in the inter-and intra-lateral region) and enlarged lymph node >1 cm in at least 2 locations, while the latter consist of elevated levels of CRP, ESR, anaemia, hypoalbuminaemia, renal failure and hypergammaglobulinemia

For the diagnosis of Castelman's disease, the coexistence of 2 large and 2 small criteria is required.

Depending on the form of the disease, patients require different therapeutic treatment. The limited form is characterised by a good prognosis (90-95% complete cure), the treatment of choice is the surgical removal of lesions [6] the diffuse form is characterised by a very aggressive course of illness and the average survival is 26 months [7]. High mortality in multifocal form is mainly associated with infectious complications, secondary tumours and local progression of nodal changes. In multifocal form, the treatment procedure depends on the coexistence of infection with the human HHV8 herpes virus. Chemotherapy and radiotherapy should primarily be considered for treatment [8]. The mechanism of action of new drugs used in the treatment of disseminated Castleman's disease is based on blocking the binding of IL-6 to its receptors - both soluble and membrane-bound [9,10].

The biological drugs used include: Siltuximab - chimeric

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monoclonal antibody - registered in 2014 in Europe for the treatment of patients who have excluded inf. HIV I HHV8 [11] and Tocilizumab - recombinant humanised monoclonal antibody, originally registered for the treatment of RA, currently also used to treat Castelman's [12].

Summary

Due to the rarity and difficulties associated with the accumulation of a representative research group, work on new treatments is of limited nature. Patients with diagnosed.

Castleman's disease should remain under the constant supervision of both an internist, and an oncologist. However, there are no detailed guidelines for monitoring the course of the disease. It seems that periodic physical examination together with performing imaging and laboratory tests, including the IL-6 level determination, should be the standard of conduct in case of these patients.

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