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Two Different Faces from Unicentric Castleman Disease (UCD): A Case Series

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Abstract Castleman disease is a rare lymphoproliferative disorder that was first reported in 1954. The most common type is unicentric and is a benign form of lymphatic disorders, which its treatment has been successfully reported. The etiology of this disease is still unclear, but there is a relationship between this disorder and overproduction of IL- 6. In our study, after treating one of the patients with UCD, due to the unexpected transformation of this disease to DLBCL, the new treatment was performed with combination chemotherapy and is still ongoing. We suggest continuous follow-up of patients during and after treatment. As well, more investigations are needed on the pathogenesis of disease that will lead to better treatment for these patients.

Keywords: *castleman disease, unicentric, DLBCL, lymphoproliferative*

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1. Introduction

Castleman disease (CD) that was first reported in 1956 by Benjamin Castleman, is an uncommon lymphoproliferative disorder [1]. The remarkable clinical characteristics of the CD are fever, weakness, organomegaly, malaise, anemia, pulmonary, and additionally CNS involvement [2]. There are two distinctive sub-groups CD: multicentric (MCD) and unicentric (UCD). The most common form of the disease is UCD that is limited to a single lymph node area and histologically, is hyaline vascular. The patients have mostly asymptomatic characteristics [3]. MCD is systemic form of the disease that less frequent and more invasive than UCD. Also, MCD has a higher mortality [4]. Understanding the pathogenesis of CD is not clear, but one of the main pathogenic processes in this disease includes the high secretion of cytokine interleukin-6 and infection with HHV8 Virus [5]. However, it has a higher frequency in HIV-infected patients [6]. Diffuse Large B-Cell Lymphoma (DLBCL) disorder is the most frequent type in non-Hodgkin lymphoma that include about 31% of the total NHL in European countries and 37% B-cell tumors around the world [7]. Recent studies have reported patients of Castleman disease with DLBCL [8]. We report two patients with Castleman disease who are UCD type, but despite their similarity in the type of disease, they have unique features and different type of treatment.

2. Case Presentation 1

The patient was a 37-year-old woman who referred to the Clinic of Hematology and Oncology, Kermanshah, Iran, with a feeling of the enlarged mass in the neck and without any systemic symptoms in June 2015. Initial clinical examinations and ultrasound results from the axillary, thyroid and cervical sections indicated a very large lymph nodes around the neck. The pathology report of excisional biopsy revealed a lymph node with overall preserved architecture, containing numerous follicles of varying sizes and based on the immunohistochemistry (IHC) findings below, our diagnosis was Castleman disease.

CD3, CD20 and Ki67 were positive, but BCL2 was negative. Also, the results of blood tests showed a low level of WBC (3.400 x 10^6/L) and Viral tests were negative for EBV, HHV, HIV, and CMV. Therefore, treatment with Chlorambucil and Prednisolone was started and continued for one year. After 19 months, due to her normal condition, maintenance therapy was started. Moreover, during this time, the results of bone marrow examination and several times the lymph node biopsy showed that there was no lymphomatous involvement.

Until April 2017, the patient felt a mass in the left side of the neck and the doctor requested computerized tomography (CT) scan of the pelvis, cervical, abdomen and biopsy of the mass in the left-side the neck. In examining CT, scan multiple lymphadenopathies with a diameter of 35 mm in the left side of the cervical and in right axillary with a diameter of 20 mm were seen. Also, the results of the biopsy and IHC were as follows: CD20, CD3, CD5, CD30, PAX5, BCL6 and Ki67 were positive, while CD23, CD43 and EMA were negative. Unexpectedly, these results revealed the change in Castleman disease to Diffuse Large B-Cell Lymphoma

(DLBCL). Therefore, the combination chemotherapy with rituximab and cyclophosphamide, doxorubicin, vincristine and prednisone (R-CHOP) started to treat the patient and is still ongoing.

3. Case Presentation 2

Our second case was a 35-year-old woman who admitted to the clinic with symptoms such as fever and coughing and a feeling of the enlarged mass in the right axillary region in February 2015. After clinical examinations, the excisional biopsy of the lymph nodes in right axillary and the IHC test was performed and the results are as follows: CD20 and LCA: positive, Ki67: poorly positive, CD10, CD30 and CD15 were negative. According to these results, her disease was diagnosed with CD. Further examinations, such as electrophoresis of serum proteins and Complete Blood Count and CT scan results were normal. Therefore, due to her normal condition, no specific treatment was performed and only the patient is under continuous followed-up.

4. Discussion

Castleman disease is also known as angio-follicular lymph node and is divided into both localized (UCD) and systemic (MCD) [9]. Due to the rarity of the disease, it is difficult to calculate the incidence [10]. UCD is the most frequent type of benign lymphoproliferative disorders that is usually treatable [11]. Chest, neck and abdomen are the most frequent sites involved in the disease, respectively [12]. Despite the important advances in past decades, our knowledge of pathogenesis and etiology of the disease is ambiguous [13]. One of the reasons for the pathogenesis of the disease is infected with the HHV8 virus, which in infected cells, it produces viral homologues of interleukin-6 and by binding to its receptors, activates the pathways of lymphocyte proliferation [14]. The standard treatment of the UCD is excision biopsy and surgical [10,15]. In this study, we showed two different faces of Castleman disease who both of them were localized and hyaline-vascular type. In the first patient after two years of treatment, the disease suddenly changed to DLBCL, which this condition lead to a change in her treatment using combination chemotherapy, such as R-CHOP but in the second patient, due to her relatively favorable condition, with the advice of a doctor, she was under constant follow-up. It is significant that unlike previous reports which described DLBCL in patients with MCD [8], in this study, we observed first this lymphoma in a patient with UCD after two years of treatment.

5. Conclusion

The most important results of this study are multi-faceted of Castleman disease despite the similarity in the type of disorder and non-specificity of occurrence of DLBCL in the type of multicentric. Also, we illustrate the importance of continuous follow-up with CT scan, which is a useful guide to diagnose and evaluate the effectiveness of treatment.

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