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Castleman Hastalığı; Boyun Kitlelerinin Nadir Nedeni

Castleman Disease; A Rare Mass of Neck

Özge Çağlar[1], Aytaç Kılıç[1], Oğuz Güçlü[1], Ceren Canbey Göret[2], Fevzi Sefa Dereköy[3]

[1]Çanakkale Onsekiz Mart Üniversitesi, Kulak Burun ve Boğaz Hastalıkları Kliniği, Çanakkale, Türkiye
[2]Sancaktepe Şehit Prof.Dr. İlhan Varank Eğitim ve Araştırma Hastanesi, İstanbul, Türkiye
[3]Medipol Üniversitesi, Kulak Burun Boğaz Bilim Dalı, Kulak Burun ve Boğaz Hastalıkları Kliniği, İstanbul, Türkiye

Öz

Castleman hastalığı, lenfoproliferatif hastalıkların heterojen bir grubu olup tek bir hastalık olarak düşünülmemelidir. Bu hastalık çeşitli klinik şekillerde ortaya çıkmaktadır. Üç histolojik çesidi olan hastalığın etyopatogenezinde IL-6, HİV ve HHV- 8 yer almaktadır. Bu hastalığı anlamada önemli yollar kat etmiş ve tedavi tecrübelerimiz rituximab, tokilizumab ve siltuximab gibi ilaçlarla artmış olsa da, halen etyolojisi, prognozu ve tedavisi hakkında cevapsız sorular bulunmaktadır. Aşağıdaki olgumuzda, meduller tiroid kanser tanısı ile takipte olan hasta boyunda kitle nedeni ile tarafımıza başvurmuş, boyun diseksiyonu sonrası patoloji sonucu castleman hastalığı olarak rapor edilmiştir.

Anahtar Kelimeler: Castleman Hastalığı, Lenf Nodu, Boyun Kitlesi

Abstract

Castleman disease is a heterogeneous group of non clonal lymphoproliferative disorders, so it should not be considered as a single disease. This disease has a wide spectrum of clinical expressions. Three histological types have been reported. IL-6, HIV and HHV-8 are involved in the etiopathogenesis of Castleman disease. Despite recent significant advances in our understanding of this disease and the increasing therapeutic experience with rituximab, tocilizumab and siltuximab, there are still difficult questions concerning its etiology, prognosis and optimal treatment. In this paper, follow up patient with medullary thyroid cancer was admitted because of mass in the neck and neck dissection was performed, pathology result was reported as Castleman disease.

Keywords: Castleman Disease, Lymph nodes, Neck Mass

INTRODUCTION

Castleman disease (CD) is a rare disease of lymph nodes and related tissues. It was first described by Dr. Benjamin Castleman in the 1950s. It is also known as Castleman disease, giant lymph node hyperplasia, and angiofollicular lymph node hyperplasia (AFH) (1). This disease is described as non-caseous lymphoid proliferation. Two types were determined according to the presence of lymph node spread such as; unicentric and multicentric (2). Pathologically, there are 4 subtypes with different diagnoses and treatments: 1. Unicentric CH (UCH), 2.Multicentric (MCH) HHV 8 and HIV-related 3.Multicentric (MCH) - HPV 8 related but not HIV related 4.Multicentric (MCH) (idiopathic) that is not associated with any virus (3). Diagnosis of multicentric disease (MCH) is difficult due to its nonspecific clinic and very low incidence. Great advances have been made to understand the pathophysiology of Castleman disease. There are several theories based on the repetitive antigenic stimuli of B lymphocytes formed against some biological agents (4). The most accepted theory is based on abnormal overproduction of IL-6 by B lymphocytes in the mantle zone. These cells are stimulated by viral antigens or yet unknown endogenous and exogenous factors. Inflammatory mediators and infections such as HHV-8 and HIV are important for explaining the etiopathogenesis and symptoms of Castleman disease. Although this disease is very common in the literature, there is still no definitive treatment. However, various drugs have been emphasized recently (5).

İletişim Bilgisi / Correspondence
Dr. Aytaç Kılıç, Çanakkale Onsekiz Mart Üniversitesi, Kulak Burun ve Boğaz Hastalıkları Kliniği, Çanakkale - Türkiye
E-mail: serkanaytackilic@hotmail.com
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CASE REPORT

The patient underwent thyroidectomy for medullary thyroid cancer. Postoperative 3rd year follow-up scintigraphy showed a large hyper metabolic lymph node in the right upper cervical chain. Right lateral neck dissection (Level 2, 3, 4) was performed but no metastasis was detected. 4rd year follow-up the patient was found to have lymphadenopathy approximately 5 cm in size at right level 5 and was reported as a necrotic lymph node in a fine needle aspiration cytology (FNAC). Operation was planned. During operation specimen was sent for frozen section result was malignant (presence of coarse atypical cells between the lymphoid elements) (Figure-1) therefore functional neck dissection was performed. The patient's final pathological result was reported to be compatible with Castleman disease (hyaline vascular variant) (Figure-2). The patient did not experience any postoperative complications, and there was no problem during the follow-up and no additional treatment was required.

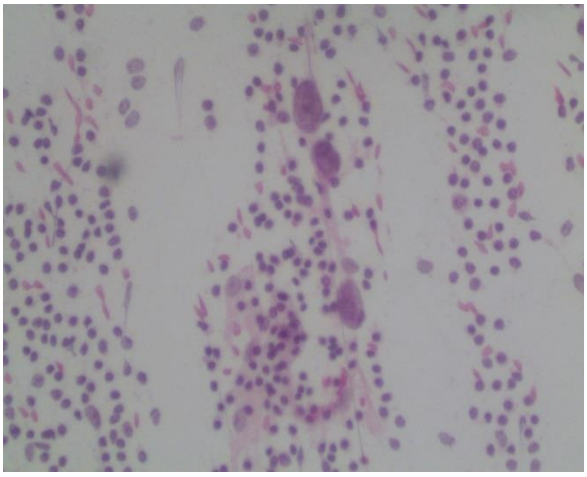

Figure 1. Frozen Section: Atipical cells (H & E x 400)

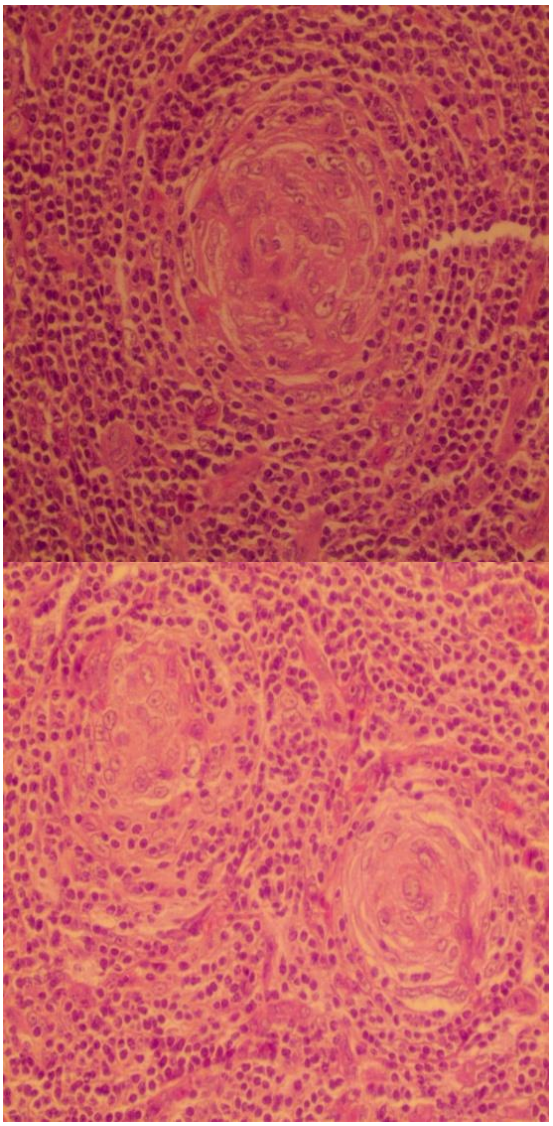

Figure 2. Central sclerotic blood vessels, concentrated lymphoid elements (H & E x 400): Castleman's Disease

DISCUSSION

Castleman disease defined as benign large hyperplastic lymph nodes by Castleman et al. (1). Although the etiology is unknown, many theories have been proposed. Histopathologically there are 3 subtypes; hyaline vascular, plasma cell and mix type. The hyaline vascular type is the most common type seen in 80-90% of cases (6). In addition, this disease has; unicentric (localized) and multicentric (generalized) types. The prognosis of the multicentric type is known to be the worst which is seen in 3-4-decade men. Unicentric type is localized form of disease that has benign subtypes. (7). This disease is usually characterized by a large lymph node which is

generally asymptomatic. On the other hand, in multicentric type systemic symptoms such as fever, weight loss, splenomegaly can be observed (8). Our case was reported to be localized hyaline variant by pathology, and since it was not multicentric type, systemic symptoms were absent. Although this disease is not yet well understood, many theories have been proposed. The most supported of these is excessive lymphoproliferation as a result of continuous stimulation by virus or chronic inflammation. This theory has been proved by a strong relationship between Castleman disease and HHV 8, HIV and EBV (9). Another strong theory is based on the relationship between IL-6, TNF and multicentric Castleman disease (9). In general, the prognosis of Castleman disease depends on the subtype. Benign localized form is treated by excision of the lesion (10). Although operation is the first choice of treatment in operable cases, they can also be referred for radiotherapy. Conversely, multicentric form can only controlled by medical treatment because of the aggressiveness of this subtype (9). Recently, experiences with chemotherapy, steroids, rituximab, anti-interleukin 6 and antivirals have increased (5). The most important part in the treatment of these patients is performing frequent follow-ups for long terms, since malignant transformation may occur (6). Since our patient was unicentric, he benefited from surgery and did not have any problems during the follow-up, so no additional treatment was recommended. In one case in the literature, it was stated that, fine needle aspiration cytology (FNAC) of a neck mass raised the suspicion of lymphoma, after excision of the mass pathology confirmed lesion as a hyaline-vascular variant of Castleman disease (11). In our case, fine needle aspiration cytology (FNAC) was reported as necrotized lymph node. During operation frozen section was performed and it was reported to be malignant, after neck dissection was performed and specimen was sent, pathology confirmed diagnosis as Castleman disease. This shows us that total excision of the mass is necessary for the accurate diagnosis of this disease, since sufficient material for pathology can be provided in only this way.

In conclusion, it is interesting that the neck mass of the follow up patient with medullary thyroid cancer is considered as metastasis and neck dissection is performed and the pathological result is Castleman disease. Although Castleman disease is a rare entity, it should be kept in mind that it may be the cause of any neck mass.

Necessary permission was obtained from the patient and relatives for the case presentation.

Informed Consent: Written consent was obtained from the participants.

Conflict of Interest: Authors declared no conflict of interest.

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