CASTLEMAN'S DISEASE IN CHILDHOOD: A CASE REPORT

ÇOCUKLUK ÇAĞI CASTLEMAN HASTALIĞI: OLGU SUNUMU

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Abstract

Castleman's Disease (CD) is a rare disease characterized by lymph node hyperplasia described by Benjamin Castleman in 1956. CD is divided into subtypes based on its etiology, pathology, involved lymph nodes number, and regions. Mediastinal lymph nodes are often affected, but any lymph node may be involved, including the neck, axilla, and abdomen. CD is initially classified as Unicentric (UCD) and Multicentric (MCD) Castleman's Disease based on the number of regions of enlarged lymph nodes. CD is histopathologically divided into three types; hyaline vascular, plasma cell, and mixed type. There are two subtypes of MCD; HHV-8 (hMCD) associated and idiopathic (iMCD). The etiology of UCD and iMCD has not been clarified. Four possible causes (virus, acquired or inherited genetic mutation, autoimmunity) are considered. The diagnosis of CD is problematic because it mimics other neoplastic and autoimmune diseases. Excision of the mass and histopathological examination is the gold standard in diagnosis. The prognosis changes according to the localization and multicentricity of the lesion. Although surgery is the first-line treatment option, chemo-radiotherapy and rituximab are also used. Since all Castleman Disease types, reactive and neoplastic lymph node pathologies are rare in childhood, Castleman Disease in childhood should be defined in more detail and its differential diagnosis should be made carefully. This article discusses Hyalinized Vascular Type Castleman Disease in the supraclavicular lymph node in a 14-year-old female patient, which is very rare especially in children, in light of current literature

Key words: Castleman disease, childhood, hyaline vascular type, lymphoma

Özet

Castleman Hastalığı (CH), 1956 yılında Benjamin Castleman tarafından tanımlanan, lenf nodu büyümesi ile karakterize nadir bir hastalıktır. CD, etiyolojisi, patolojisi, tutulan lenf nodu sayısı ve bölgelerine göre alt tiplere ayrılır. Mediastinal lenf bezleri sıklıkla etkilenmekle birlikte boyun, koltuk altı ve karın dahil olmak üzere herhangi bir lenf nodu etkilenebilir. CD başlangıçta genişlemiş lenf nodlarının bölgelerinin sayısına göre Unisentrik (UCD) ve Multisentrik (MCD) Castleman Hastalığı olarak sınıflandırılır. CD histopatolojik olarak üçe ayrılır; hiyalin vasküler, plazma hücreli ve karışık tip. MCD'nin iki alt tipi vardır, HHV-8 (hMCD) ile ilişkili ve idiyopatik (iMCD). UCD ve iMCD'nin etiyolojisi açıklığa kavuşturulmamıştır. Dört olası neden (virüs, edinilmiş veya kalıtsal genetik mutasyon, otoimmünite) dikkate alınır. Diğer neoplastik ve otoimmün hastalıkları taklit ettiği için ÇH tanısı sorunludur. Kitlenin eksizyonu ve histopatolojik inceleme tanıda altın standarttır. Prognoz lezyonun lokalizasyonuna ve multisentrikliğine göre değişir. Cerrahi birinci basamak tedavi seçeneği olmakla birlikte kemo-radyoterapi ve rituximab da kullanılmaktadır. Çocukluk çağında tüm Castleman Hastalığı tipler, reaktif ve neoplastik lenf nodu patolojileri nadir görüldüğünden, çocukluk çağındaki Castleman Hastalığı daha ayrıntılı tanımlanmalı ve ayırıcı tanısı dikkatli yapılmalıdır. Bu yazıda özellikle çocuklarda oldukça nadir görülen 14 yaşındaki kadın hastada supraklaviküler lenf nodunda görülen Hyalinize Vasküler Tip Castleman Hastalığı güncel literatür ışığında tartışılmaktadır.

Anahtar Kelimler: Castleman hastalığı, çocukluk çağı, hiyalin vasküler tip, lenfoma

1. INTRODUCTION

Castleman's Disease (CD) is a rare disease with unclear etiology, characterized by focal or diffuse massive enlargement of lymph nodes, described by Benjamin Castleman in 1956. CD has different subtypes based on its etiology, pathology, and clinical presentation. It can affect lymph nodes in any body region, mimicking both benign and malignant malformations, primarily including the chest, neck, abdomen, and axilla (1,2).

In our case, we aimed to present CD in the supraclavicular lymph node, which is extremely rare especially in children, in a 14-year-old female patient.

2. CASE REPORT:

A 14-year-old female patient was admitted to the pediatric surgery department of our hospital with complaints of increasing swelling in the supraclavicular region for about 5 months, and fever and night sweats recurring every 3-4 days recently. In the ultrasound examination, two lymph nodes, 4 cm and 1,5 cm in diameter, were observed in the supraclavicular region. In this examination, no pathological lymph node was detected in a different

location. Laboratory values were normal. Lymph node excisional biopsy was performed and macroscopic examination of the material sent to our department revealed two lymph nodes, the largest measuring 4x3,5x1,5 cm and the smaller 1,5x1,5x0,5 cm, with smooth cross-sectional surface, solid appearance and gray-beige in color. In the microscopic examination, lymphoid follicles, which were not limited to the cortex and scattered throughout the entire parenchyma, were noted in the lymph nodes. It was observed that the involuted, twinning finding was observed in places, the collagenized germinal centers were radially penetrated by the capillaries and the lymphoid follicle had a typical lollipop appearance. The germinal centers are hyalinized and the concentric layers of small lymphocytes are surrounded by a "typical onion peel appearance". There is marked vascular proliferation covered by hyperplastic endothelial cells and surrounded by a collagen sheath in the interfollicular areas. HHV-8 and EBV were evaluated as negative in the immunohistochemical study. The present findings were considered compatible with Hyaline Vascular Type Castleman's Disease.

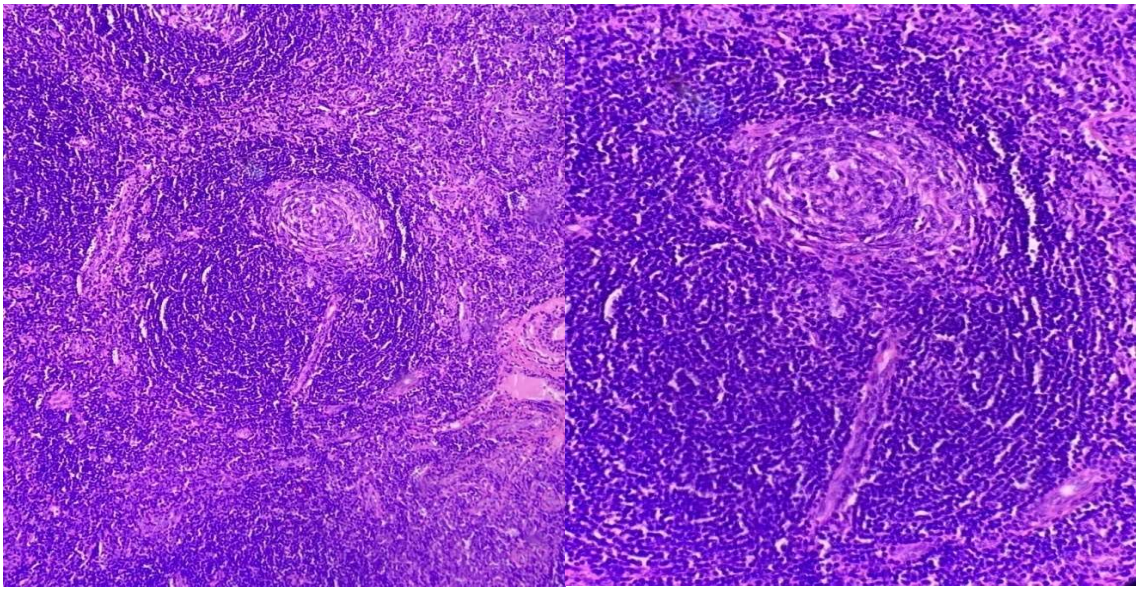

Figure 1: Typical lollipop appearance of the lymphoid follicle. (H&Ex4 and H&Ex20)

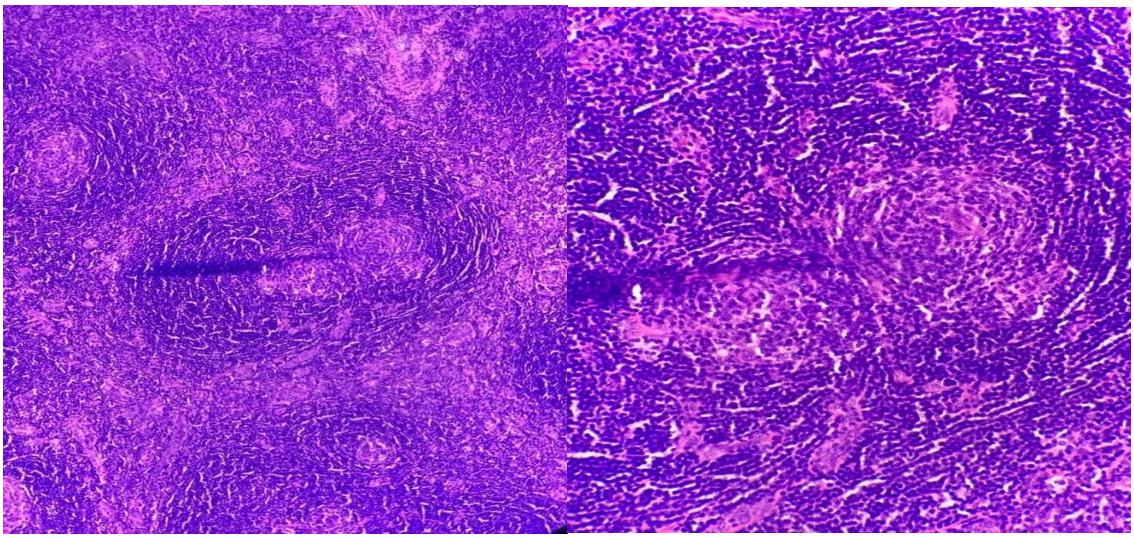

Figure 2: Finding of twinning in germinal centers (H&Ex4 and H&Ex20

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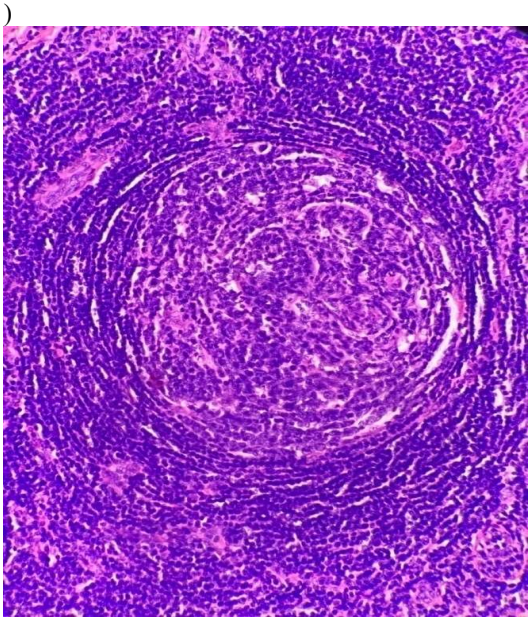

Figure 3: Mantle zone thickening and typical onion skin appearance (H&Ex20)

3. DISCUSSION

CD, first described in 1956 in a patient with a localized mediastinal mass, is also known as angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, lymph node hamartoma, and benign giant lymphoma. CD is first classified according to the number of regions of enlarged lymph nodes (often the mediastinum) (3). Unicentric Castleman's Disease (UCD) includes a pathological single lymph node or a single site, while Multicentric Castleman's Disease (MCD) includes multiple pathological lymph node sites. UCD can affect all age groups and is most common in the 4th decade. There is no gender predilection. MCD, on the other hand, can be seen at any age, but is most common in the 5th and 7th decades. There are two subtypes of MCH as HHV-8 (hMCD) associated and idiopathic (iMCD) (4).

CD is examined histopathologically under three headings; hyaline vascular, plasma cell and mixed type. Although the hyaline vascular type, which we described in our case, is the most common, it is usually asymptomatic and the focal lymph node is affected. With the enlargement of the lymph node, it gives symptoms because of compression. The plasma cell type is a systemic lymphoproliferative disorder associated with immunological abnormalities that appear mostly multifocal. Its prognosis is worse (5). In childhood, as in our case, UCH and Hyalinized Vascular Type are seen more frequently (6).

The etiology of UCD and iMCD has not been clearly elucidated. Recently, four possible causes have been postulated: viruses, an acquired genetic mutation, an inherited genetic mutation, or autoimmunity. HHV-8 is the best-known cause of hMCD, accounting for approximately 25-50% of all MCD cases. Approximately 50-75% of MCD cases are HHV-8 negative and thus "idiopathic" (4,7,8).

Although the gold standard diagnostic method in CD is histopathological evaluation (incisional/excisional biopsy), radiological findings and laboratory tests are helpful.

Surgery is considered the first-line treatment option. If surgical excision is not possible, rituximab, IL-6, chemotherapy, radiotherapy, and stem cell transplantation are preferred (9).

4. CONCLUSION

CD is a mostly benign proliferation of lymphocytes and vascular structures, also known as giant lymph node hyperplasia, lymph node hamartoma, angiofollicular lymph node hyperplasia (10). CD is easily confused with lymphoma or other solid tumors. Therefore, it is essential to accurately diagnose the complete type of CD and differentiate it from other diseases with clinical history and laboratory diagnostic measures, along with additional imaging techniques for rapid treatment and management procedures. Although CD has been described as a benign lymphoproliferative disease, systemic forms are particularly associated with related neoplasms and autoimmune disorders such as Kaposi's sarcoma and Follicular dendritic cell (FDC) tumors (8). Because of the rarity of all types of CD, which can give the impression of malignancy such as lymphoma etc. Clinically and radiologically and diagnosed by histopathological examination in children, and its wide variety in treatment, there is a need for continuous definition of diagnosis, treatment, and outcomes in this population (6,11).

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