Castleman's Disease with an Unusual Radiological Presentation: A Case Report

Farklı Radyolojik Görünüm İle Prezente Olan Castleman Hastalığı Olgusu

Castleman Hastalığı Olgu Sunumu / Castleman's Disease: A Case Report

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Özet

Castleman hastalığı, nadir görülen bir lenfoproliferatif patolojidir. Localize ve yaygın olmak üzere iki farklı klinik tablo ile ortaya çıkabilmektedir. Histopatolojik olarak ise hyaline-vasküler tip, plazma hücreli tip ve mikst tip olmak üzere 3 grupta değerlendirilir. Olgular sıklıkla asemptomatik seyretmekte olup radyolojik bulgularla şüphe edilerek tanı konulur. Radyolojik olarak en sık prezentasyonu mediastinal alanda dev lenf nodu şeklindedir. Lenfadenomegali en sık paratrakeal lenf nodlarında izlenir. Bu nadir görülen patolojiyi vurgulamak ve farklı radyolojik görünümü ile özellik arz eden bir olgu ile bu hastalığı hatırlatmak istedik.

Anahtar Kelimeler

Castleman Hastalığı; Histopatoloji; Hiyalin Damar; Lenf Nodu

Abstract

Castleman's disease is a rare lymphoproliferative pathology which has two clinicoradiological forms; localized disease and disseminated disease. Histopathologically, Castleman's disease is evaluated in three groups; hyaline-vascular type, plasma cell type and mixed type. Patients are often asymptomatic and are diagnosed by radiological findings. The most common radiological presentation is huge lymph nodes in mediastinal area. Lymphadenopathy is most frequently observed in paratracheal lymph nodes. We wanted to emphasize this very rare entity and remind you this disease by the mean of a case with different radiological appearance.

Keywords

Castleman's Disease; Histopathology; Hyaline Vascular; Lymph Node

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Introduction

Castleman's disease (CD) is an uncommon, lymphoproliferative disorder often also called angiofollicular lymph node hyperplasia which involves a massive non-malignant proliferation of lymphoid tissues and and vascular proliferation in the interfollicular region. CD was initially described among a small series of patients in 1956 by Benjamin Castleman [1]. It typically presents as mediastinal masses with an unknown etiology [2].

It has two clinicoradiological forms; localized/unicentric disease (UCD) or disseminated/multicentric disease (MCD). The incidence of UCD form is higher than MCD. The UCD form is commonly localized to the mediastinum or pulmonary hilum but can also involve cervical, axillary, and abdominal lymph nodes. Whereas, MCD may involve lymph nodes separately or clustered and often submits systemic symptoms, autoimmune phenomena and aggressive course [3,4].

The diagnosis of CD usually depends on clinical suspicion on radiological findings and confirming the identification with histopathological evaluation. CD is histologically subdivided into hyaline-vascular variant (80–90% of cases), plasma cell variant (10–20%) and mixed variants. While hyaline-vascular variant accounts for approximately 90% of UCD, plasma cell and mixed variants are observed in both UCD and MCD, and accounts approximately 10% of UCD. Patients with the hylaline-vascular variant especially in UCD forms are generally asympthomatic or exhibit with only lymphadenopathy. On the other hand, patients with the plasma cell variant usually admit with weakness, weight loss, fever, rash and anemia [4,5]. While the main histopathological findings of hyaline-vascular variant are sclerotic blood vessels radially penetrating lymphoid follicles and imparting the characteristic "lollipop on a stick" appearance; those of plasma cell variant are sheets of polyclonal plasma cells within the interfollicular zone and more variable vascular proliferation compared to the hyaline-vascular variant [6,7].

The aim here was to describe a case of asymptomatic intrathoracic UCD with atypical radiological presentation in interlober lymph nodes.

Case Report

PATIENT: 21 years old male, with no complaint, referred to our clinic because of a lesion seen in his chest graphy in routine screening for health screening. Because of suspectious atelectasis in middle lobe on chest X-ray, Thorax CT scan taken on. Thorax CT revealed peribronchovascular densities around bilateral lower lobe bronchii and collapse in middle lobe of right lung and inferior of the lingula in left lung (Figure 1). Exact boundaries of the lesions were indistinguishable and the structure of the lesions could not be identified exactly. In order to make a clear diagnosis, the patient underwent diagnostic bronchoscopy. Multiple transbronchial biopsies were taken from the mediobasal segment bronchus of the right lower lob bronchus and transbronchial needle aspiration performed through to the medial wall of the right lower lob bronchus. The histopathological evaluations of the biopsies were not diagnostic. Examples were composed of bronchial mucosa cells with minimal inflammatory signs. Whereupon, diagnostic surgical excisional biopsy was performed to the patient in the thoracic surgery department. Pathology of the specimens obtained during surgical dissection were reported as CD, hyaline vascular type histopathologicaly.

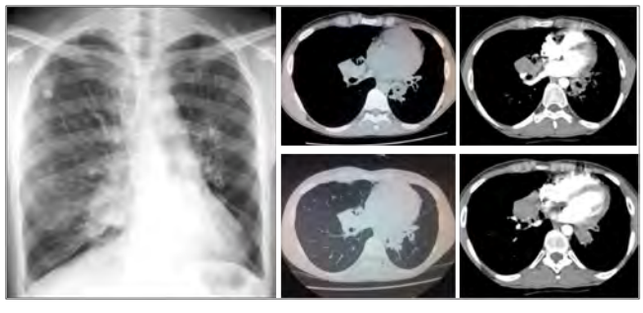
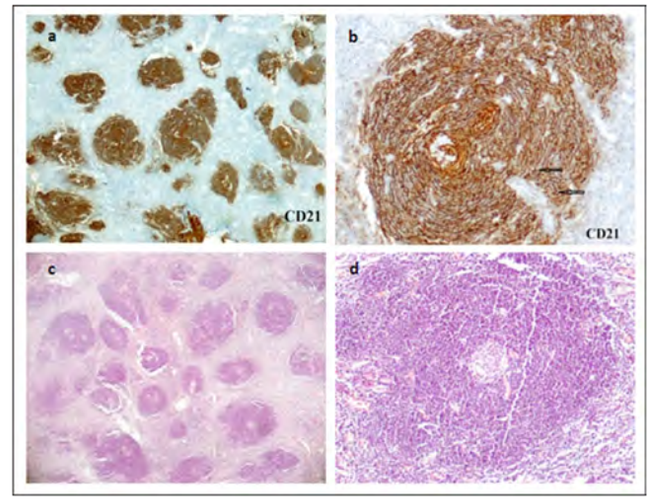


Figure 1. Radiological appearance of peribronchovascular densities around bilateral lower lobe bronchii

HISTOMORPHOLOGİCAL FINDINGS: The histological sections revealed that the nodal architecture of the lymph node was generally preserved, but it was remarkable that the germinal centers were contracted and they had hyalinized vascular structures at the center of them. There were increased number of dilated vessels, small lymphocytes and scattered plasma cells in the interfollicular area (Figure 2A). The mantle zones surrounding the germinal centers were markedly expanded and had increased follicular dendritic cell meshwork surrounded by concentric rings of small lymphocytes that lead to characteristic onion-skin appearance (Figure 2B). Some germinal centers were penetrated by a small hyalinized blood vessel (Figure 2D).

IMMUNOHISTOCHEMICAL FINDINGS: Immunohistochemical study demonstrated increased follicular dendritic cell meshwork by using CD21 antibody which is a dendritic cell marker (Figure 2C and 2D). There was no evidence of clonality within the Mantle zone cells by using Kappa and Lambda light chain antibodies. And also there was no immunoreactivity with the Cyclin D1 antibody.



Figue 2. lymphoid follicles with atretic germinal centers dispersed in a vascular-rich stroma. Note prominently dilated blood vessels and plasma cells in interfollicular area (20xH&E)(A), at higher magnification, expanded mantle zone containing follicular dendritic cells surrounding concentrically the germinal center. This appearance may resemble an onion-skin (100xH&E)(B), immunohistochemical study with CD21 antibody illustrated prominent FDC mesh work (20xIHC-CD21) (C), at higher magnification, the same staining concretized the onion-skin pattern. Note the blood vessel penetrating through the follicle (arrows) (100xIHC-CD21) (D).

Discussion

CD, firstly described by Dr. Benjamin Castleman in 1956, is a rare clinical entity also referred as lymph node hyperplasia, or angiofollicular lymph node hyperplasia. Although the majority

of lesions occur within the chest, less commonly other sites including neck, pelvis, retroperitoneum and axilla may be involved. The UCD, which is more common than MCD, generally affects young patients less than 30 years of age and complete excision allows full recovery although some cases may need a combination of chemoradiotherapy and surgery. The disease has two distinct histological types; hyaline-vascular and plasma cell variants. The former, most common type is characterized by histologically distinctive follicles with expanded mantle zones of small lymphocytes forming concentric rings surrounding one or more atretic germinal centers. There is prominent vascularity of the germinal centers, often with a single prominent penetrating vessel. Another important feature is vascular proliferation between the follicles, often with perivascular hyalinization [1,2,6]. The case which has been presented in this paper is corresponding to this type of CD with its similar features.

The main differential diagnosis should be made between hyaline-vascular type of CD and Mantle cell lymphoma (MCL) which also has the similar histological features with expansion of mantle zone areas. The latter is usually devoid of dilated blood vessels in the interfollicular area. And also, in MCL proliferating mantle zone cells are neoplastic and they show a clonal proliferation which can be illustrated by using kappa and lambda light chain antibodies by immunohistochemically [6]. More importantly, the neoplastic cells show nuclear positivity of Cyclin D1 (or Bcl-1) which is a product of a proto-oncogene generated by a t (11; 14) (q13; q32) translocation in MCL [7]. In our case no clonality was detected by using kappa/lambda light chain antibodies and mantle zone cells were negative for Cyclin D1 by immunohistochemically.

Mediastinal localization is the most common manifestation of UCD. But, the disease usually occurs in the form of a giant mediastinal lymph node. In this case exact boundaries of the lesions were indistinguishable and the structure of the lesions could not be identified exactly. Therefore, it could not be clearly identified that the lesion belongs to a lymphatic structure firstly. In detailed evaluation of the radiological findings, lesions also partially resembled lymphadenomegalies in sarcoidosis. Frankly, CD was not one of our pre-diagnosis preliminarily. Viewed from this aspect, we think that this case is meaningful to remind the atypical variations of the CD and importance of a good histopathological evaluation in such cases.

Competing interests

The authors declare that they have no competing interests.

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