

Unicentric Castleman's Disease Presenting With Multiple Enlarged Lymph Nodes In Mediastinum

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ABSTRACT

A case of unicentric Castleman's disease located in the mediastinum is presented. A 20 years old male suffering from cough had an image of right hilar enlargement on chest X-ray. Thorax CT revealed multiple enlarged lymph nodes both in right paratracheal and hilar region. Radionuclide accumulation was detected by ⁶⁷Ga scintigraphy. Surgical removal of lymph nodes at paratracheal region was performed by mediastinoscopic excision and reported as reactive hyperplasia. Lymph nodes located in right hilum was excised by thoracotomy and histopathologic examination confirmed Castleman's disease. Our thought about this case was a dominant infiltrative mass with an associated lymphadenopathy. ⁶⁷Ga scintigraphy imaging that we performed in our case can be a useful method in detection of Castleman's disease.

Key words: Castleman's disease, mediastinum, ⁶⁷Ga scintigraphy

Mediastende Birden Fazla Büyümüş Lenf Nodu ile Görülen Unisentrik Castleman Hastalığı

Bu yazıda mediastende yerleşim gösteren unisentrik Castleman hastalığı sunulmaktadır. Öksürük yakınması olan 20 yaşında erkek hastanın çekilen P/A akciğer grafisinde sağ hiler genişleme saptandı. Toraks BT'de sağ paratrakeal ve hiler bölgede birden fazla büyümüş lenf nodları görüldü. ⁶⁷Ga sintigrafisi ile radyonuklid birikimi saptandı. Mediastinoskopik olarak paratrakeal lenf nodu eksizyonu yapıldı ve reaktif hiperplazi olarak raporlandı. Daha sonra torakotomi ile sağ hiler bölgede bulunan lenf nodu eksize edildi ve histopatolojik inceleme sonucu Castleman hastalığı ile uyumlu bulundu. Olgunun, dominant infiltratif kitleye eşlik eden lenfadenopati olduğunu düşünmekteyiz. Olgumuzda yapılan ⁶⁷Ga sintigrafisi, hastalığın saptanmasında yararlı bir tetkik olabilir.

Anahtar kelimeler: Castleman hastalığı, mediasten, ⁶⁷Ga sintigrafisi

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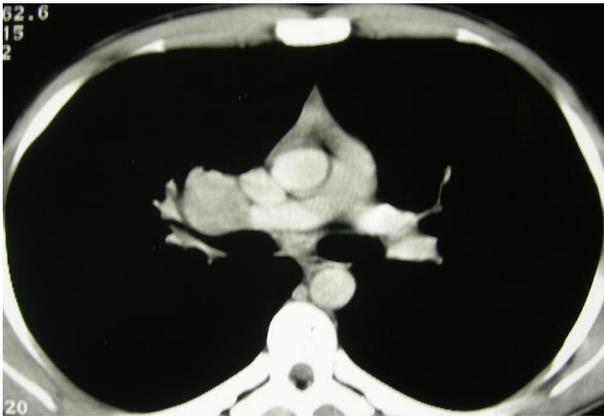


Figure 1. Contrast-enhanced Chest CT image (mediastinal window) shows 30x35 mm in diameters lobulated lymph node in right hilar field.

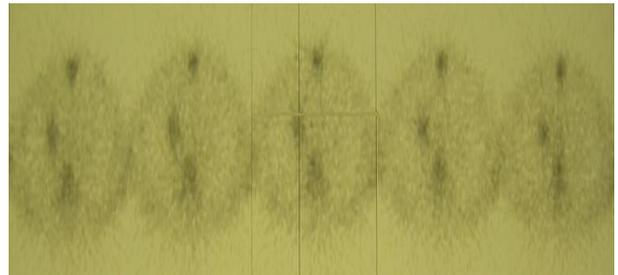


Figure 2. Galium-67 scintigraphy imaging shows an increased uptake of ⁶⁷Ga in right hilar region

INTRODUCTION

Unicentric Castleman's disease, usually seen as a mediastinal mass, is a rare lymphoproliferative disorder with an unknown etiology (1,2). This disease is named by Benjamin Castleman, a medical doctor who reported clinicopathologic features of a case with a solitary hyperplastic mediastinal lymph node first time in 1956 (3). There are two types in clinical presentation, which are localized-unicentric and multicentric disease. Hyaline-vascular and plasma cell types occurs histopathologically (2).

The most common type is unicentric hyaline vascular type which consists 70% of all cases. Although it is commonly seen in mediastinum, it can be found in lymphoid nodes of cervical, axillary or abdominal chain. Lymphoid nodes can be involved unique or through the chain way. We presented a rarely seen case with lymphadenopathy in right hilum and mediastinum diagnosed with unicentric Castleman's disease. Additionally, there are some contradictory findings about Galium 67 (⁶⁷Ga) uptake in Castleman's disease in literature. In discussion session we report our thought about our case.

CASE

A 20 years old male with a symptom of cough was admit-

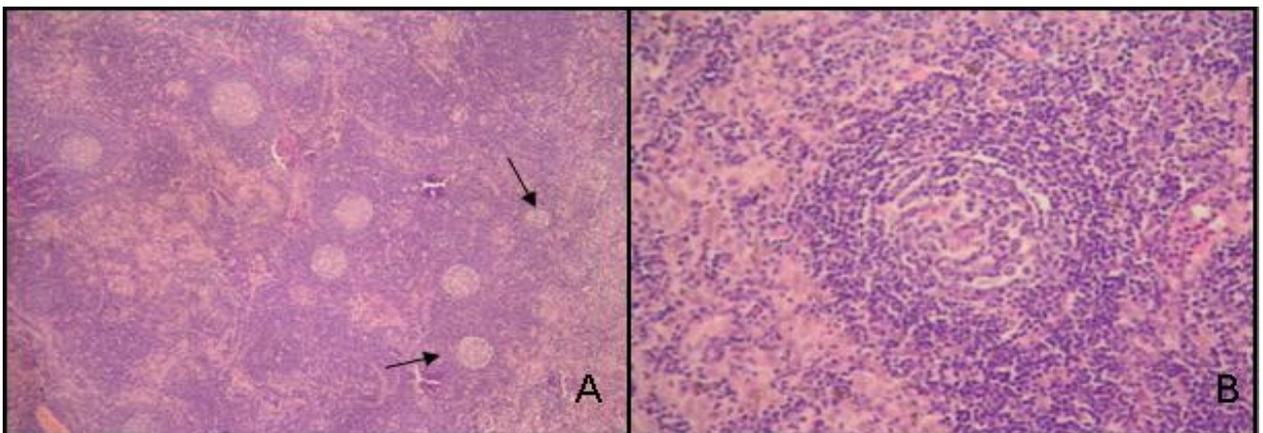


Figure 3. Histopathologic appearance of lymph node in Castleman's disease. (A) Decreased lymphoid follicles (Hematoxyline-Eosin, x40). (B). Involution in the germinal centre of lymphoid follicle and concentric distribution of lymphocytes around. "Onion skinning" appearance, lymphoid follicle with the hyaline vascularized center. (Hematoxyline-Eosin, x100).

ted to our department. His medical history and physical examination were unremarkable. Chest X-ray revealed right hilar enlargement. He had a mild anemia, positive PPD skin test and an elevated sedimentation of 71 mm/h at presentation. Thorax CT revealed enlarged paratracheal lymph nodes and lobulated lymph node in right hilar region measured 30x35 mm in diameters (Figure 1). Bronchoscopy was performed and no pathologic finding was seen. Sputum and bronchoalveolar lavage smear and culture for acid-fast bacilli (AFB) were both negative. Galium-67 scintigraphy imaging showed an increased uptake of ^{67}Ga in right hilar region (Figure 2) and interpreted as a benign disorder with a prediagnosis of sarcoidosis or tuberculosis. Surgical removal of right paratracheal lymph nodes was performed by mediastinoscopy. Histopathologic examination of specimen was reported as a reactive hyperplasia of lymph nodes. Based on available clinical data the patient was diagnosed as tuberculosis lymphadenitis initially and treated with antituberculosis drugs for two months. Chest CT was performed after the initial therapy but there was no change in size of mediastinal lymph nodes. The case was discussed in medical council and underwent to thoracotomy procedure. Conglomerated lymph nodes in right hilar region were excised and histopathologic examination revealed Castleman's disease of hyaline-vascular (HV) type (Figure 3). In mediastinopic examination, the excision biopsy samples that are taken from paratracheal lymph nodes have been re-examined after the patient was diagnosed with Castleman's disease. However, besides nonspecific lymphadenitis no other diagnostic findings were reached.

DISCUSSION

Unicentric Castleman's disease usually appears as a single sharply marginated smooth or lobulated mass in mediastinum (2, 4-7). McAdams and colleagues published series of 30 case with Castleman's disease in 1998 (8). Several cases were reported in which the mass was invading contiguous structures or associated with multiple enlarged lymph nodes. They observed three morphologic patterns of unicentric-localized Castleman's disease by CT or MR imaging: (a) solitary mass, (b) a dominant infiltrative mass with associated lymphadenopathy, (c) multiple lymph nodes of similar size. In our case, there were multiple enlarged lymph nodes in mediastinum where the biggest one was 30x35 mm in diameters in

right hilar region. We think our case had dominant infiltrative mass with associated lymphadenopathy. We have screened the published data about Castleman's disease in our country and we have found that most of the cases were unicentric and hyaline vascular type Castleman's disease and the most often location was in mediastinum. These findings are in accordance with the literature. However, the prevalence of the illness in our country is still unknown.

^{67}Ga scintigraphy is still used in the search of lymphoproliferative diseases because it is a noninvasive and practical test (9). The test is used in the discrimination of malign and benign type lymphoproliferative diseases and at the same time to narrow the diagnostic possibilities. In our case there were lymphadenomegalies in mediastinal and hilar regions. In order to evaluate the ^{67}Ga accumulation in the mediastinal and hilar regions of our patient, a ^{67}Ga scintigraphy was performed. The result of the test revealed a ^{67}Ga accumulation in the hilar region. The findings were benign by scintigraphy. Therefore, with the consideration that tuberculosis lymphadenitis is common in our country anti-tuberculosis treatment was started to the patient. However, the patient did not benefit from the treatment, therefore an invasive surgical procedure was carried out. Even though the diagnostic range was narrowed by scintigraphy, the importance of histopathological diagnosis was once again revealed.

Okamoto et al. performed ^{67}Ga scintigraphy in two patients with the diagnosis of unicentric Castleman's disease, but there was no uptake (10). One of these cases had mediastinal mass of HV and the other had mesenteric mass of plasma cell type. The reason for negative ^{67}Ga uptake was indicated as presence of low activated lymphocyte rate. Kinoshita et al. found enlarged lymph nodes in anterior cervical region and uptake of ^{67}Ga in a patient with unicentric Castleman's disease. Finally they mentioned ^{67}Ga scintigraphy as a sensitive test in the diagnosis of Castleman's disease (10). Our case also had ^{67}Ga uptake in right hilar region as reported in these series. We had considered these lymph nodes as a benign disorder and treated as tuberculosis disease initially. Later the cases is rediagnosed as Castleman's disease after the surgical removal of mediastinal lymph nodes by thoracotomy. In cases of benign mediastinal lesions that are confirmed with ^{67}Ga scintigraphy, Castleman's disease is also kept in mind in clinical diagnosis.

In conclusion; we think our case had dominant infiltrative mass with associated lymphadenopathy and ⁶⁷Ga scintigraphy is a useful method to detect this disease in addition to radiologic and histopathologic examinations in unicentric Castleman's diseases.

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