

# Asymptomatic Isolated Retroperitoneal Castleman's Disease: A Case Report

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## Abstract

Castleman's disease, giant lymph node hyperplasia, is a kind of benign lymphoproliferative disease with gentle behavior. Its etiology and prevalence are unclear. This rare disease is usually found in mediastinal area asymptotically and incidentally. It is also rare to see this tumor in the retroperitoneum. In this study, we have introduced a 34-year-old woman who referred just with occasional abdominal pain caused by compressive symptoms. Laboratory findings only reported microcytic anemia (MCH: 18.5, MCV: 63, Hemoglobin 10.2 g/dl). Chest and abdominal X-ray imaging showed no remarkable point. In abdominal ultrasonography, a solid and firm tumor with 12.2×5.3×6.6 cm was reported in patient's retroperitoneum. Patient's surgery was done and the tumor (covered by a fibrous thick capsule, with no bizarre appearance and bleeding) was completely removed. Pathologic examination indicated a Castleman's tumor, type of unicentric and hyaline-vascular. This item had been one of the rare reported items of Castleman's disease in the retroperitoneal space.

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**Keywords** • Castleman's disease • Giant lymph node Hyperplasia lymph node • Retroperitoneal space

## Introduction

Castleman's disease, giant lymph node hyperplasia, is an autoimmune, lymphoproliferative disease that shows itself with the enlargement of lymph nodes and varied clinical presentations. Castleman's disease commonly involves mediastinum and hence it is thoracic in most of the reported cases. The etiology of this disease has remained unknown. It is a rare disease that presents itself by hyperplasia of lymph nodes with no malignant origin.<sup>1</sup> This disease was introduced to the world of science in 1950s by Dr. Benjamin Castleman with presentation of 13 patients with unicentric hyaline vascular.<sup>2</sup>

This disease is known by other names such as angiofollicular lymph node hyperplasia or giant lymph node hyperplasia. Simply, the pathology of this disease can be attributed to the hypervascular of lymph nodes and hyalinization of vessels.<sup>2-4</sup>

From a pathological perspective, three variants of this type of tumor are recognized: hyaline vascular CD type, plasma cell type, and mixed type.<sup>2</sup>

From a clinical standpoint, this tumor is found in two types of unicentric and multi-centric, in which the unicentric type is much

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more common.<sup>5</sup> Often, the disease involves the mediastinum space and abdominal involvement is rare.<sup>6</sup> Previous studies have presented it as an unusual finding in retroperitoneal CT imaging.<sup>7</sup>

In this study, we aim at introducing one patient with Castleman's disease who had referred with clinical picture of occasional abdominal pain.

### Case Presentation

A 34-year-old woman complaining of occasional abdominal pain referred to the surgery clinic. Acute pains involved area below the navel, with preference left lower quadrant. There was no remarkable point in physical examination and the patient did not have any other clinical symptoms.

Laboratory findings only reported microcytic anemia (MCH: 18.5, MCV: 63, Hemoglobin 10.2 g/dl). Imaging results were chest and abdominal X-ray without any remarkable point. In abdominal ultrasonography, a solid and firm tumor with 12.2×5.3×6.6 cm was reported in patient's retroperitoneum. For more evaluation, abdominal CT scan was carried out. The tumor was not attached to the walls of the intestines and it did not cause a blockage. With coordinates obtained by CT scan and ultrasonography (US), surgery was performed in order to remove the tumor.

A big tumor was found in the retroperitoneum that was solid and firm but was not attached to the walls of the intestine or to the lymph nodes of that area. In the operation area, lymph nodes were slightly larger than usual. The tumor was completely removed in order to treat patient and more reviews. From the macroscopic point of view, the tumor was shaped like an egg, covered by a fibrous thick capsule, with no bizarre appearance and without visible bleeding on its surface (Figure 1). Its size was 121×52×67 mm before cutting. In cutting points, its surface was homogeneous, chocolaty and in terms of consistency, hard. Although it was similar to lipoma in early studies, but the initial pathologic study reported its lymphoid origin that rejected the possibility of malignancy.

More histopathological investigation of tumor was an extensive lymphatic tissue containing hyperplastic follicles that was placed in the frame of a lymphoid tumor.

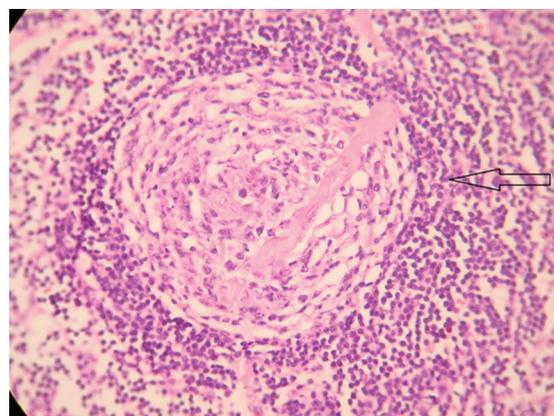
Mantle zones of follicles had spread and small germinal centers were seen (Figure 2).

Among these germinal centers, there were widespread vascular assemblies with hyalinized wall.

Interfollicular stroma as hyperplasia is defined as post-capillaries venules, in which



**Figure 1:** Macroscopic view of the resected Castleman's tumor (the tumor was shaped like an egg, covered by fibrous thick capsule, with no bizarre appearance and bleeding).



**Figure 2:** Microscopic view; expanded mantle zone and the small germinal centers of the lymphoid follicles have shown apparently. Hyaline vascular type revealed Castleman's disease (H&E, X400).

combination of plasma cells and eosinophils are seen. Histopathology of these samples gave a definitive diagnosis of angiofollicular lymph node hyperplasia, which in this case was hyaline vascular type.

A chest CT scan was performed at the end of treatment and its review implied the absence of a similar tumor elsewhere. Accordingly, the patient was placed in the group of isolated Castleman's disease. The patient did not have any particular postoperative problems and was discharged on day-7 after surgery. Patient's follow up was done until 9 months after discharge for symptoms of recurrence or any kind of clinical abnormalities. The patient was treated for anemia and her blood lab results were absolutely normal.

Our study was approved by the Medical Ethics Committee, according to the Helsinki declaration. Our patient was informed and gave consent.

## Discussion

Castleman's disease is a kind of rare pathology, usually benign, with unclear etiology and prevalence. It is reported in childhood and adolescence periods in much lower numbers.<sup>6,8</sup> In most of the reported cases, the disease arises in the chest and especially in the mediastinum. From other involved areas, we can mention mesentery, armpits, neck and in very rare cases in retroperitoneum.

Despite the fact that the etiology of this disease remained unknown, we can cite autoimmune disease, and some viral diseases as predisposing factors for this disease, abnormality of test results and clinical examinations.<sup>9</sup> None of the above diseases was seen in this reported case, even until the end of the follow-up period. Therefore, these immunity factors are discussed as predisposing factors and not as a main etiological factor.

As mentioned, the Castleman's disease is clinically divided into two groups of unicentric and multi-centric. In unicentric type, that is more common, the possibility of invasion is lower and is seen with hyaline vascular type. The tumor is asymptomatic and localized, which is often found accidentally. Patients sometimes report symptoms such as fever, fatigue, or pain from pressure.<sup>10</sup> These symptoms were also observed in our reported patient. Therefore, it is difficult and sometimes impossible to diagnose only based on disease symptoms before the operation and pathology study, especially in such diagnosis.

Among clinical tests, it is possible that complete blood to be along with anemia with small changes, similar to what was observed in our patient, while this finding is not specific.<sup>11</sup>

In some case reports, the next step is imaging, initiated with the X-ray primary approach.<sup>11</sup> We also conducted chest and abdominal X-ray on this patient. Lack of result from these cannot be a reason to reject Castleman's disease.

Imaging with ultrasonography has been the main approach in some previous reports. Some studies started their diagnostic procedures with endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA).<sup>12,13</sup> In this study, we used ultrasonography after abdominal X-ray leading to tumor detection and locating its exact location by using an abdominal CT scan. Of course, since no study with a high sample size has been conducted in this field, sensitivity and specificity of none of these diagnostic methods were exactly identified. Histopathology of tumor tissue after surgery is the only way for tumor (and its type) diagnosis.

Most of the Castleman's tumor is seen as homogeneous and hypoechoic tumor in the view of the sonography. In color Doppler view, we often see capillaries with homogeneous spread. This view is also seen more in hyaline vascular type. In general, it can be concluded that the radiological view is not specific enough to detect this disease and the observed views could be mistaken with each type of benign or malignant lymphomatous tumor.<sup>14</sup>

In this study, similar to most other published case reports, the tumor was of the unicentric type while multi-centric type is less common and should be noted with a broader therapeutic approach. It should be noted that in this disease, the localized type often responds to surgical treatment alone.

Complete removal of tumor and its margin will suffice with the treatment of laparotomy and even laparoscopy. These kinds of tumors do not have invasive behavior and are completely benign.<sup>15</sup>

On the other hand, the treatment of malignant group often requires additional interventions such as chemotherapy or corticotherapy.

## Conclusion

Castleman's disease is a kind of rare vascular hyperplasia that has often benign and with noninvasive behavior. There is no reliable diagnostic method and its definitive diagnosis is based on histopathology report. It is often found in the chest, especially in the mediastinum and is asymptomatic in most cases.

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