Case Report

Retroperitoneal unicentric Castleman's disease: A multidisciplinary approach in a rare case

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Abstract

Retroperitoneal masses present with various clinical forms and are very difficult to diagnose before surgery. Castleman's disease occurs in the lymphoid chain and is characterized by angiofollicular hyperplasia of the lymphoid tissue with an unknown etiology. In this case report, we will share with the venerable readers a rare occurrence of retroperitoneal Castleman's disease in light of the current literature.

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Introduction

Castleman's disease (CD) is a rare lympho-proliferative disorder. It is also known as giant lymph node hyperplasia, lymph node hamartoma, angiofollicular mediastinal lymph node hyperplasia, or angiomatous lymphoid hyperplasia[1,2] and was first described in 1956 via reporting of the results of 13 multi-centered cases by Castleman B. et al. It is a rare disease determined by masses of lymph nodes, usually incidentally[3,4]. It is difficult to find any information about the incidence and prevalence of CD because it has been defined by case presentations and small case series[5]. The most common location of CD is the mediastinum and it is characterized by unicentric (localized) or multicentric large lymphoid hyperplastic masses[6,7]. It may be less frequently located at the posterior neck, abdominal and axillary areas[8]. Retroperitoneal CD in the iliak region is a rare entity. In this case report, we will present a rare occurrence of retroperitoneal CD.

Case presentation

A 56-year-old man without past medical or surgical history, presented at our hospital with abdominal pain for 4 months. There was no history of fever, night sweats, abdominal lump, decreased appetite, weight loss, or alteration of bowel habit. No pathology was found on the abdominal examination and the rectal examination was normal. Blood investigations including hematological and biochemical tests were normal. A value of the PSA was 1.4 ng/dl. A solid lesion with increased peripheral echogenicity was detected with a hypoechoic heterogeneous appearance and a size of 47 x 42 mm, extending to the obturator fossa, located in the iliac chain on the right lateral side of the bladder on ultrasound (USG). Magnetic resonance imaging (MRI) of the patient revealed a 55 x 43 x 36 mm mass lesion in the right obturator chain adjacent to the right lateral wall of the bladder. Positron emission tomography (PET-CT) imaging showed pathologically increased F-18 FDG uptake in the conglomerate lymph nodes (SUV max:7.4) in the right obturator chain and lateral pelvic structures (Figure 1 and 2). At a council with the Hematology Clinic, it was decided that two-time true-cut mass biopsy would be performed in terms of the extermination of lymphoproliferative disease diagnoses. Immunohistochemical studies on pathological material revealed no evidence of lymphoid malignancy.

Due to the suspicion of malignancy, at a second council, the decision to perform laparoscopic right obturator mass excision was made. On postoperative day 13, the patient was discharged.

On pathological gross examination, it was a capsulated mass, measuring 60 x 45 x 20 mm with a dirty white color and a multinodular cut surface. Microscopic examination showed that the lymph node was surrounded by a fibrous thick capsule. It was observed that the lymph node was large with numerous follicles throughout the cortex and medulla (Figure 3). Loss of sinuses, thick sclerotic bands, numerous cortical and medullary follicle structures were observed. Lymphoid follicle was characterized by atrophic germinal centers with surrounding concentric rings of B-lymphocytes Lymphoid follicle, bearing some resemblance to a Hassall corpuscle of the thymus, was observed (Figure 4). In some of the follicles, lymphocyte depletion, mantle-zone enlargements, hyaline deposits in the germinal centers, vascular structures perpendicular to the follicular structures, stroma-rich areas and high endothelial venules in the interfollicular areas were observed. Some of the follicles contained more than one germinal center (twinning) (Figure 5). Immunohistochemical studies revealed CD20 antibody positive and HHV-8 negative. Thus, the patient was diagnosed with a hyaline vascular variant of CD (Figure 3 and 4).

In the council after the pathology result, it was decided to follow up the patient with close-up radiologic imaging. Rituximab therapy was planned in case of the recurrence of the disease due to the absence of residual mass.

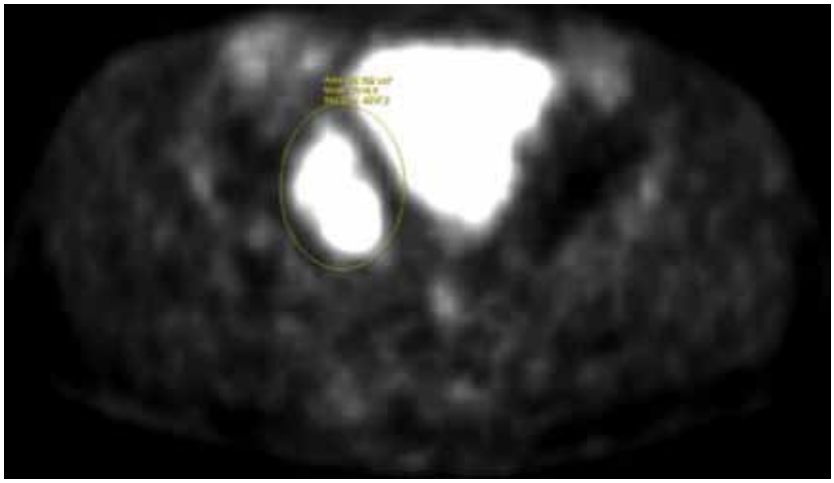

Figure 1.
PET-CT imaging showed increased F-18 FDG uptake in the conglomerate lymph nodes (SUV max:7.4)

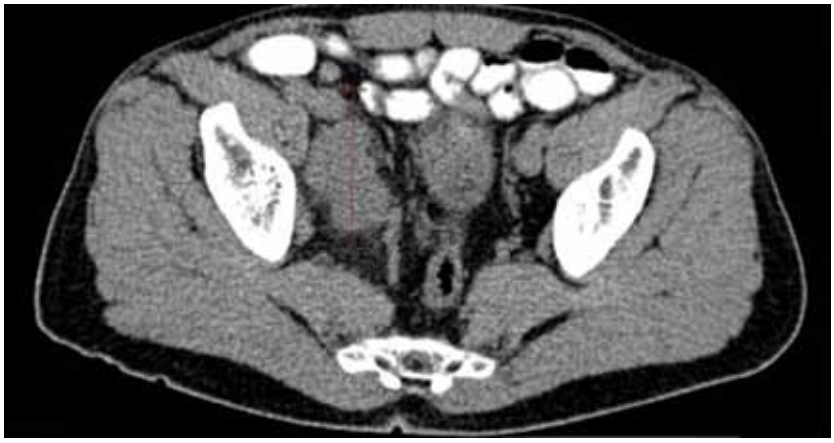

Figure 2.
CT imaging showed conglomerate lymph nodes

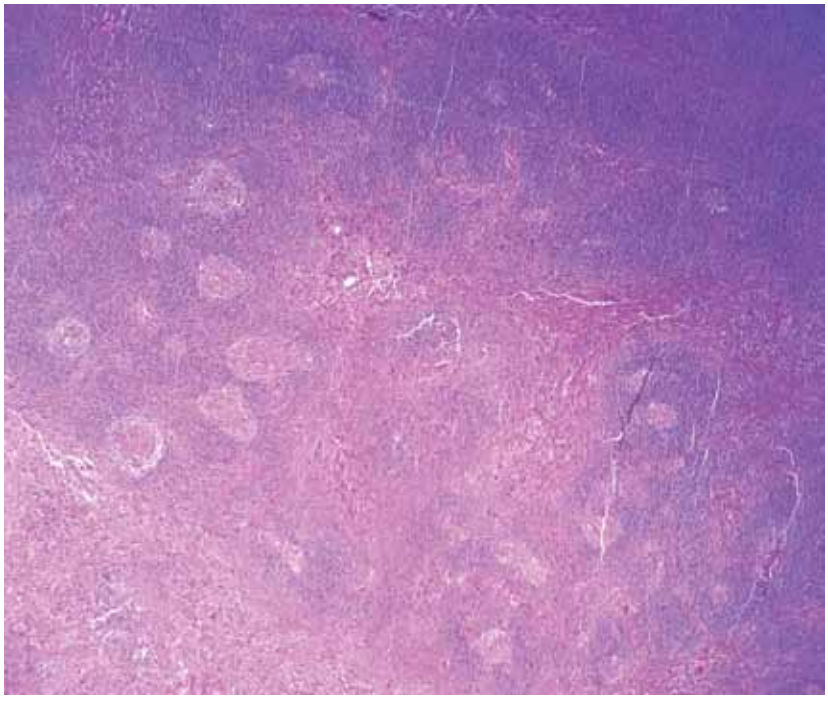

Figure 3.
H&E, X40: The lymph node observed large with numerous follicles throughout the cortex and medulla.

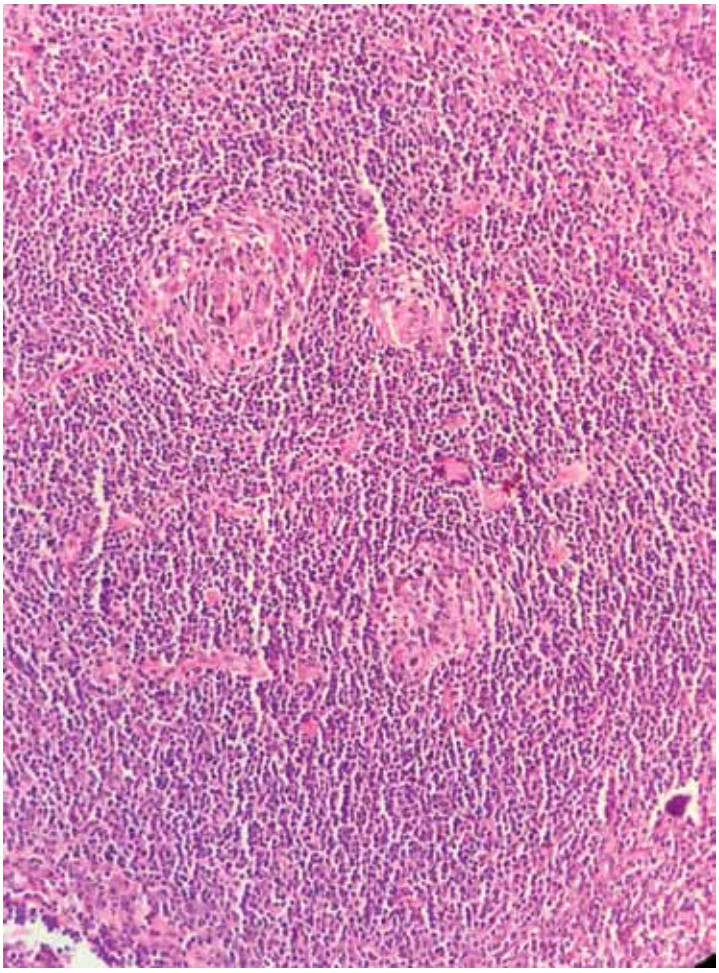

Figure 4.
H&E, X100: Lymphoid follicle characterized by atrophic germinal centers, bearing some resemblance to a Hassall corpuscle of the the thymus, was observed

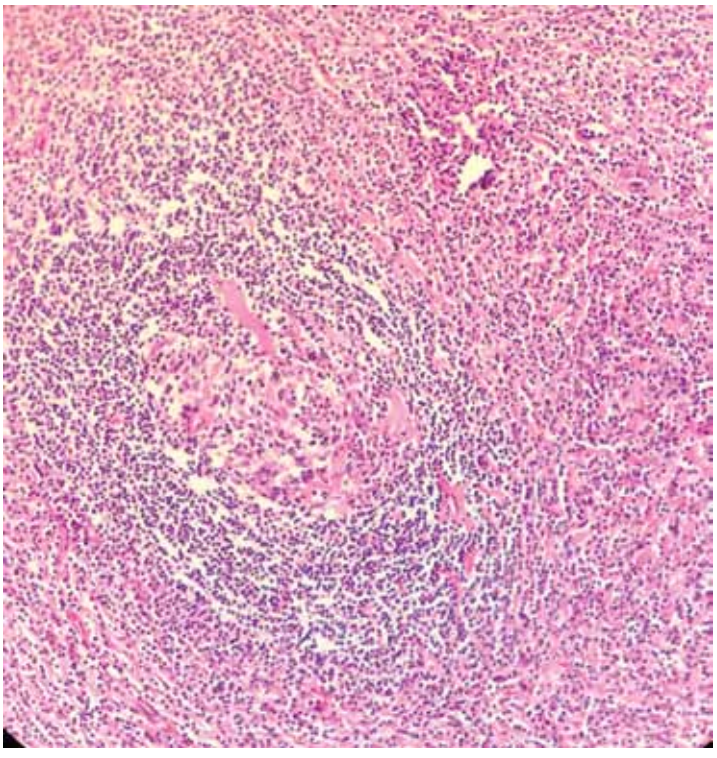

Figure 5
H&E, X200: Two follicles are present in this field. The follicle contains more than one germinal center (twinning).

Discussion

In retroperitoneal mass cases approach and management are difficult. CD is a rare cause of retroperitoneal masses and they present in various clinical forms. Viral agents, angiogenesis, and interleukin-6 play a role in the pathophysiology of this disease[9,10]. Epstein-Barr Virus, Human Herpes Virus-8 and Human Immunodeficiency Virus as viral agents have various mechanisms of CD formation[9,11]. In angiogenesis, Vascular Endothelial Growth Factor (VEGF) leads to CD formation[9]. In CD cases, VEGF supernatant in pathologic material was found to be 100 times higher than VEGF supernatant in normal lymphoid hyperplasia[9,12]. There was not any viral infection history in our patient. CD is observed in 3 clinical variants. Seventy-two percent of patients with CD are clinically unicentric hyaline vascular variant, 18% unicentric plasma cell variant and 10% multicentric plasma cell variant[9]. Unicentric variant CD may be mediastinal, cervical or abdominal. It is usually incidentally detected in the 3rd decade and its 5-year survival is 100%[7]. Multicentric variant CD is clinically characterized with systemic symptoms in the 6th decade, presents with AIDS or lymphoma, and its median survival is 30 months[7]. In our case, the unisentric mass was located in the right obturator fossa. After the complete excision of the mass, on the pathological examination the patient was diagnosed with a hyaline vascular variant of CD.

Surgical excision, chemotherapy, radiotherapy, immunomodulators (thalidomide, interferon) and monoclonal antibodies are used in the treatment[9]. In unicentric variant CD treatment, surgical excision can be curative, but it is often difficult due to its close proximity to the major vascular stricture[9]. We planned a close follow-up with imaging studies, and in the case of the recurrence of the disease, rituximab treatment was planned.

Therefore, careful dissection should be performed during mass excision. As in our case, one of the major problems in lymphoid mass excisions is the prolonged post-operative lymphatic leakage. We could not discharge our patient until the 13th day postoperatively because of lymphatic leakage. Octreotide treatment was needed to prevent lymphatic leakage in our patient.

In the current literature, anti-IL-6 monoclonal anti-body (atlizumab) and anti-CD20 monoclonal antibody (rituximab) are used in multicentric variant CD patients[9,13]. Because of the CD-20 positivity in our case, we planned rituximab treatment in the case of the recurrence during the follow-up period.

Conclusion

Castleman's disease is a rare cause of retroperitoneal masses, which have indeterminate behavior and can be treated with numerous treatment options.

References

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