Castleman Disease Mimicking Ovarian Tumour

Alper KARALÖK[1], Tolga TAŞÇI[1], Işın ÜREYEN[1], Sevgi KOÇ[1], Nurettin BORAN[1], Deniz ÇAVUŞOĞLU[2], Taner TURAN[1], Gökhan TULUNAY[1]

Ankara, Turkey

ABSTRACT

Castleman disease is a lymphoid disease characterized by herpes virus infection associated hyperplasia of lymphatic tissue. Castleman disease is generally localized in the mediastinum (70%) and the regions that it may be seen outside of the thorax are neck, axilla, pelvis and retroperitoneum. Castleman disease may present unicentrically or multicentrically.

Fifty-six year old postmenopausal woman was detected to have a right adnexial mass in her routine gynecological examination. This adnexial mass was also observed in the pelvic ultrasonography and pelvic magnetic resonance imaging (MRI). A retroperitoneal mass was detected in the right hemipelvis. Pathological evaluation revealed Castleman disease, hyaline vascular type. Any lymphadenopathy other than this wasn't observed in the systemic imaging of the patient. Therefore, she was considered to have unicentric disease and was told to come to follow-up visits.

Castleman disease is a rare condition. Since symptoms and imaging findings aren't specific to the disease, preoperative diagnosis is quite difficult. Castleman disease located in the pelvic retroperitoneum may mimic adnexial masses. It is genearlly related to pelvic walls and iliac vessels. Surgical removal of unicentric Castleman disease is curative.

While Castleman disease is observed rarely in gynecological practice, it should be kept in mind in the differential diagnosis of adnexial masses.

Keywords: Castleman disease, Adnexial mass, Ovarian tumour

Gynecol Obstet Reprod Med 2016;22(2):105-107 DOI: 10.21613/GORM.2016.481

Introduction

Castleman disease (CD) is associated with herpes virus infection and is characterized by lymphoid hyperplasia (1). It was first described in 1956 and it's also called angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, hyperplasia of lymph follicle (1,2). It can be encountered anywhere that contains lymphoid tissue and it is classified into three categories based on histopathologic characteristics and into two based on clinical features (3).

In 1972, Keller et al. pathologically identified two major forms, hyalinized vascular and plasma cell, and a mixed variant that contains two types together (3).

[1] *Etlik Zubeyde Hanim Women's Health Teaching and Research Hospital Gynecologic Oncology Surgery Clinic, Ankara*
[2] *Etlik Zubeyde Hanim Women's Health Teaching and Research Hospital Pathology Division, Ankara*

Address of Correspondence: *Alper Karalök*
Etlik Zubeyde Hanim Women's Health Teaching and Research Hospital, Gynecologic Oncology Division, Etlik Ankara Turkey
alperkaralok@yahoo.com

Submitted for Publication: *04. 02. 2015*
Accepted for Publication: *22. 05. 2015*

Based on clinical features there is two types; unicentric and multicentric. Unicentric form is found in only one location and is usually asymptomatic. Multicentric form is associated with generalized lymphadenopathy, systemic symptoms, hepatosplenomegaly and malign transformation and more aggressive prognosis (4). Frequently, the disease is confined to mediastinum (70%) but extrathorasic lesions can occur in neck, axilla and retroperitoneum (3).

We present here a case of pelvic hyaline vascular type unicentric CD that mimics ovarian tumour.

Case Report

A 56 years old gravida 5, parity 3 patient applied to our hospital for urge incontinence. Gynecologic examination revealed grade 2 cystosele. A 5 cm solid mass was encountered at the right adnexa on bimanual examination. Transvaginal sonogram revealed a right sided 55x30 mm para-ovarian, complicated, cystic-solid mass. A 55x44 mm hyperintense mass showing dense pattern of contrast enhancement, located in the right ovary was encountered in magnetic resonance imaging of abdomen. Tumour markers (Ca125:10, Ca19.9: 7.4, Ca15-3:21), complete blood count and sedimentation rate (16 mm/h) were in normal range. Chest X-ray was normal. Laparatomic exploration revealed a 5 cm sized lym-

phadenopathy which settled on right external iliac and obturator artery and vein, fixed to adjacent tissue. Inner genital organs and other intraabdominal structures were considered normal. Obturator nerve was passing through the mass. The mass was dissected from obturator artery, vein and nerve. Internal iliac vein was teared during dissection and repaired primarily so vascular continuity was achieved. Intraoperative histopathologic evaluation demonstrated a lymphoid hyperplasia but malign-benign discrimination could not be made. Operation was ended due to frozen section result. There was cervical, axillar and inguinal lymphadenopathy in the postoperative systemic examination. Final pathology was suspicious for CD. Immunohystochemistry revealed hyaline vascular type CD. Computerized tomography of thorax, abdomen and neck which was ordered by hematology clinic, was normal except the minimal fluid in abdomen secondary to operation. Bone narrow biopsy was normal. Flow Cytometry showed a pattern of polyclonal growth. Finally, diagnosis was made as hyaline vascular unicentric CD. Follow-up and no adjuvant treatment were recommended.

Discussion

Castleman disease is usually a disease of young age and usually it is seen under age of 30 6 (4). There are two clinical forms. Unicentric (localized) form is found at solely one lymph node or lymph node region. Multicentric (systemic) form is found at multiple lymph node groups at the same time. There are three histologic type; hyaline vascular form, plasma cell form and mixed type which contains both types and is seen rarely (3,5-9)

Unicentric form comprises 90% of all cases and most of all are pathologically hyaline vascular type. Prognosis is quite well and although it is generally asymptomatic it can be encountered with pressure symptoms. Nevertheless, systemic symptoms can be seen rarely (3,10). Unicentric form which is located in the pelvis is associated with pelvic lateral wall and frequently it settled on iliac vessels. It can be associated with pressure symptoms such as pelvic pain and pollakiuria (11-13).

Multicentric or systemic form is associated with plasma cell form pathologically. This comprises 10% of cases and generally it is encountered with systemic symptoms (anemia, fever, fatigue), generalized lymphadenopathy and hepatosplenomegaly. This form can cause hepatic and renal failure via infection or malignity (14). In contrast to unicentric form its prognosis is poor and median overall survival is 30 months (3,5-8,14-16).

Castleman disease is located in mediastinum in 70%, superficial lymph nodes in 20% and retroperitoneum in 7% of the cases. In 21% of retroperitoneal cases, lesion is located in pelvic region (17). Lung, pancreas, breast, adrenal gland and muscles can rarely be involved without nodal involvement (7).

In Japanese literature, 22 pelvic retroperitoneal CD cases were presented. Nineteen of them were hyaline vascular type and asymptomatic and 3 of them were solitary and plasma cell type. Two of three plasma cell type cases were associated with fever, high CRP and IL-6 and polyclonal hypergammaglobulinemia (17).

Etiopathogenesis is not clear. It is thought that it is associated with increased production of IL-6 and human herpes virus-8. HSV-8 was demonstrated in lymphoid cells in the systemic form of disease (18). The physiopathology is slow progression of inflammatory process. This inflammatory process causes hyperplasia of lymphoid tissue (9,19,20). As distinct from certain types of lymphomas it is characterized by polyclonal proliferation (9,16). Multicentric disease can rarely be associated with amyloidosis and POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, M Protein, Skin Changes) (7).

Laboratory abnormalities such as anemia, hypoalbuminemia, polyclonal gammopathy, high erythrocyte sedimentation rate and CRP level and proteinuria are seen more frequently in multicentric form (21). Tubo-ovarian abscess, endometrioma and teratoma should be considered in differential diagnosis of CD located in the pelvis (5). There is no specific diagnostic tool for preoperative diagnosis of CD (17,22). No specific finding on computerized tomography and magnetic resonance imaging was identified. These techniques are useful for identification of localization of lesions (10). Frequently, a hyperechoic and homogeneous mass, sometimes with a sharp acoustic shadowing caused by central calcification, is seen in sonography (5,23).

The management of unicentric form differs from multicentric form. The standard in management of unicentric form is en-bloc resection of the mass. Because of the dense adhesions between the mass and adjacent tissues, possibility of complication, especially bleeding, is notably high (17). No recurrence was reported after total resection of the mass (24). Five year survival rate is nearly 100% after resection in unicentric form (16). Recurrence is rare even after incomplete surgical resection (25). Radiotherapy is an effective treatment option for patients who can't be operated or those with incomplete surgical resection (6,26).

Surgery is not curative for multicentric disease and the role of radiotherapy is not clear (6,26). Steroids are used to treat multicentric form alone or concurrent with systemic chemotherapy. Adding splenectomy to systemic treatment is shown to prolong survival (27).

In conclusion, CD is a rare condition. Because symptoms and imaging findings are not specific for the disease, preoperative diagnosis is difficult. CD that is located in the retroperitoneal space in the pelvis and mimics adnexal mass is generally unicentric type and surgery can be performed with curative intent. However, complications, especially laceration of major vessels, can developed.

References

1. Castleman B, Iverson L, Menendez VP. Localized mediastinal lymph-node hyperplasia resembling thymoma. Cancer 1956;9:822-30.
2. Toni LM, John KMCT Features of Castleman disease of the abdomen and pelvis. AJR 2000;175:115-8
3. Keller AR, Hochholzer L, Castleman B. Hyaline-vascular and plasma-cell types of giant lymph node hyperplasia of mediastinum and other locations. Cancer 1972;29:670-83.
4. Peterson B, Frizzera G. Multicentric Castleman's disease. Semin Oncol 1993;20:636-47.
5. Hsieh CH, Changchien CC, Lan KC, Huang CC, Shen CC, Chang SY, Lin H. Pelvic Castleman's disease presenting as an adnexal tumour. Acta Obstet Gynecol Scand. 2004;83(3):311-3.
6. Bowne WB, Lewis JJ, Filippa DA, Niesvizky R, Brooks AD, Burt ME, et al. The management of unicentric and multicentric Castleman's disease: a report of 16 cases and a review of the literature. Cancer 1999;85:706-17.
7. Bucher P, Chassot G, Zufferey G, Ris F, Huber O, Morel P. Surgical management of abdominal and retroperitoneal Castleman's disease.World J Surg Oncol 2005;3:33.
8. Seco JL, Velasco F, Manuel JS, Serrano SR, Tomas L, Velasco A. Retroperitoneal Castleman's disease. Surgery 1992;112:850-5.
9. Larroche C, Cacoub P, Godeau P. La maladie de Castleman. Rev Med Interne 1996;17:1003-13.
10. McCarty MJ, Vukelija SJ, Banks PM, Weiss RB. Angiofollicular lymph node hyperplasia (Castleman's disease). Cancer Treat Rev 1995;21:291-310.
11. Ylinen K, Sarlomo-Rikala M, Laatikainen T. Pelvic Castleman disease mimicking an adnexal tumour. Obstet Gynecol 1995;85:894-7.
12. Giaretta MI, Hyun J, Gibbons JM Jr. Angiomatous lymphoid hamartoma as a pelvic mass. A case report. Obstet Gynecol 1971;38:391-4.
13. Bainbridge ET. Angiomatous lymphoid hamartoma of the pelvis. Br J Obstet Gynecol 1976;83:823-6.
14. Ziv Y, Shikiar S, Segal M, Orda O (1993) Bilateral localized Castleman disease of the retroperitoneum. Eur J Surg Oncol 19:188-91
15. Goldberg MA, Deluca SA. Castleman's disease. Am Fam Physician 1989;49:151-3.
16. Shahidi H, Myers JL, Kvale PA: Castleman's disease. Mayo Clin Proc 1995;70:969-77.
17. Sato A. Castleman's disease in the pelvic retroperitoneum: A case report and review of the Japanese literature. Int J Surg Case Rep 2013;4(1):19-22
18. Cesarman E, Knowles DM: The Role of Kaposi's sarcoma-associated herpesvirus (KHSV/HHV-8) in lymphoproliférative diseases. Semin Cancer Biol 1999;9:165-74.
19. Gaba AR, Stein RS, Sweet DL, Variakojis D. Multicentric giant lymph node hyperplasia. Am J Clin Pathol 1978; 69:86-90.
20. Kessler E. Multicentric giant lymph node hyperplasia. A report of seven cases. Cancer 1985;56:2446-51.
21. Kim TJ, Han JK, Kim YH, Kim TK, Choi BI. Castleman disease of the abdomen: imaging spectrum and clinicopathologic correlations. J Comput Assist Tomogr 2001; 25:207-14.
22. Barki Y, Shadked G, Levy I. Mesenteric Castleman disease: sonographic diagnosis. J Clin Ultrasound 1992;20: 486-8.
23. Taura T, Takashima S, Shakudo M, Kaminou T, Yamada R, Isoda K. Castleman's disease of the spleen: CT, MR imaging and angiographic findings. Eur J Radiol 2000, 36:11-15.
24. Kiguchi H, Ishii T, Ishikawa Y, Masuda S, Asuwa N, Yamafuji K, Takahashi T. Castleman's disease of the abdomen and pelvis: Report of three cases and a review of the literature. J Gastroenterol 1995;30:661-6.
25. Ebisuno S, Yamauchi T, Fukatani T, Ohkawa T: Retroperitoneal Castleman's disease: A case report and brief review of tumours of the pararenal area. Urol Int 1989;44:169-72.
26. Chronowski GM, Ha CS, Wilder RB, Cabanillas F, Manning J, Cox JD. Treatment of unicentric and multicentric Castleman disease and the role of radiotherapy. Cancer 2001;92:670-6.
27. Lerza R, Castello G, Truini M, Ballarino P, Tredici S, Cavallini D, PannacciulliI: Splenectomy induced complete remission in a patient with multicentric Castleman's disease and autoimmune haemolytic anaemia. Ann Hematol 1999;78:193-6.