Case Report

Castleman Disease Mimicking Ovarian Tumor

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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 generally 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

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 intra-abdominal 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 tearing during dissection and repaired primarily so vascular continuity was achieved. Intraoperative histopathologic evaluation demonstrated a lymphoid hyperplasia but malignant-benign discrimination could not be made. Operation was ended due to frozen section result. There was cervical, axillary and inguinal lymphadenopathy in the postoperative systemic examination. Final pathology was suspicious for CD. Immunohistochemistry 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 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


