Retroperitoneal Castleman's Disease Incidentally Detected due to Urinary Calculus

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Castleman's disease (CD) is a rare benign tumor characterized by hyperplasia of lymphoid tissue. It can occur in almost any l ocation, with 70% in the mediastinum, 7% in the retroperitoneum, and only 2% in the perirenal area. Approximately 80% of primary retroperitoneal tumors are malignant, so tumor resection and differential diagnosis with other retroperitoneal tumors is important. We present a case of a hyaline-vascular type CD in the pararenal area, detected incidentally during evaluation of a urinary calculus, with a review of the literature. (**Korean J Urol 2008; 49:186-189)**

Key Words: Castleman's disease, Retroperitoneum, Neoplasms

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Castleman's disease (CD) is a fairly rare and unusual condition of lymph node hyperplasia with unknown etiology.[1] Most CD is seen in the mediastinum, but may be located in cervical, axillary, inguinal, shoulder and vulvar regions. 7% of reported cases may be in the retroperitoneum, with only 2% in the perirenal area in particular.[2]

In the literature, there were 122 cases of retroperitoneal CD found in a research conducted in the year 2005.[3] In Korea, retroperitoneal CD has been reported occasionally, since reported in 1992.[4]

We present a case of CD, incidentally detected due to urinary calculus, with review of literature.

CASE REPORT

A 29-year-old man was hospitalized with intermittent painless gross hematuria and right flank pain from one month before admission. The patient had been healthy previously and had no history of disease. Physical examination revealed no significant findings such as costovertebral angle tenderness, palpable mass and peripheral lymphadenopathy.

Urinalysis revealed microscopic hematuria. Examination of complete blood count, routine blood biochemistry and HIV test showed no abnormal findings.

With the clinical impression of urinary calculus in right side, intravenous pyelography (IVP) was performed. IVP demonstrated a 0.6x0.2 cm sized urinary calculus of ureterovesical junction on the right side and lateral tilting of right upper ureter just like a mass effect (Fig. 1).

Abdominal computed tomography (CT) scan demonstrated a well-defined enhancing mass (8x5 cm) on the back of the vena cava at the level of renal hilum of a right kidney. The vena cava was displaced forward by the retroperitoneal mass. The mass had inhomogeneous, lower density portions, but not any calcifications. There was no invasion into adjacent organs (Fig. 2). There was a small sized urinary calculus of ureterovesical junction, measured about 0.6x0.2 cm, on the right side.

Considering his young age, there was a possibility of lympadenopathy caused by testicular tumor. Therefore, palpation of scrotum was conducted and both testis were found to be normal. The same result was found on scrotal ultrasonography.

After spontaneous passage of urinary calculus with conservative treatment, the patient did not complain of gross hema-

turia or right flank pain.

Tumor markers, consisting of αFP, β-hCG, LDH, CEA and CA 19-9, showed normal findings. Surgical exploration of the retroperitoneal mass was performed with an impression of retroperitoneal malignant tumor.

Operative findings were the following: the retroperitoneal mass was yellowish gray, large and elastic. It adhered to surrounding structures, including inferior vena cava, vertebrae and the aorta. The mass had three feeding vessels from the aorta.

Grossly, it was round, well-demarcated yellowish gray soft mass, measuring 8x6.5x5 cm in dimensions and weight was 155 g. On section, the cut surface showed a well demarcated yellowish white homogenous rubbery parenchyma (Fig. 3A). Microscopically, the sections showed large follicles scattered in a mass of lymphoid tissue. The follicles showed marked vascular proliferation and small, relatively inconspicuous germinal centers (Fig. 3B). The interfollicular stroma was prominent with numerous hyperplastic vessels of the postcapillary venule type and showed an admixture of plasma cells, eosinophils, immunoblasts, and kp-1 positive plasmacytoid monocytes. The diagno-

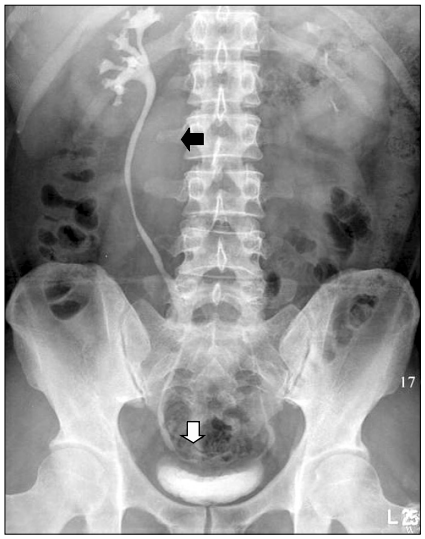

Fig. 1. Intravenous pyelography (IVP) shows a small urinary calculus of the right ureterovesical junction (white arrow). The right ureter was tilted laterally because of the mass effect (black arrow).

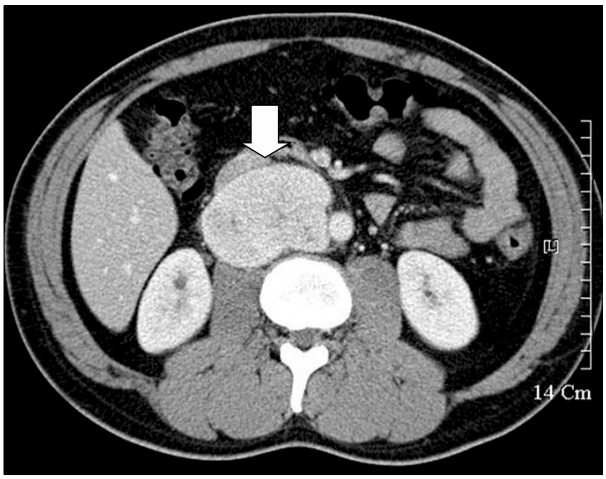

Fig. 2. CT scan of the abdomen shows a well-defined enhancing mass (8x5 cm) with a non-homogenous, low density portion on the back of the vena cava at the level of renal hilum. The vena cava is pushed forward by the mass (white arrow).

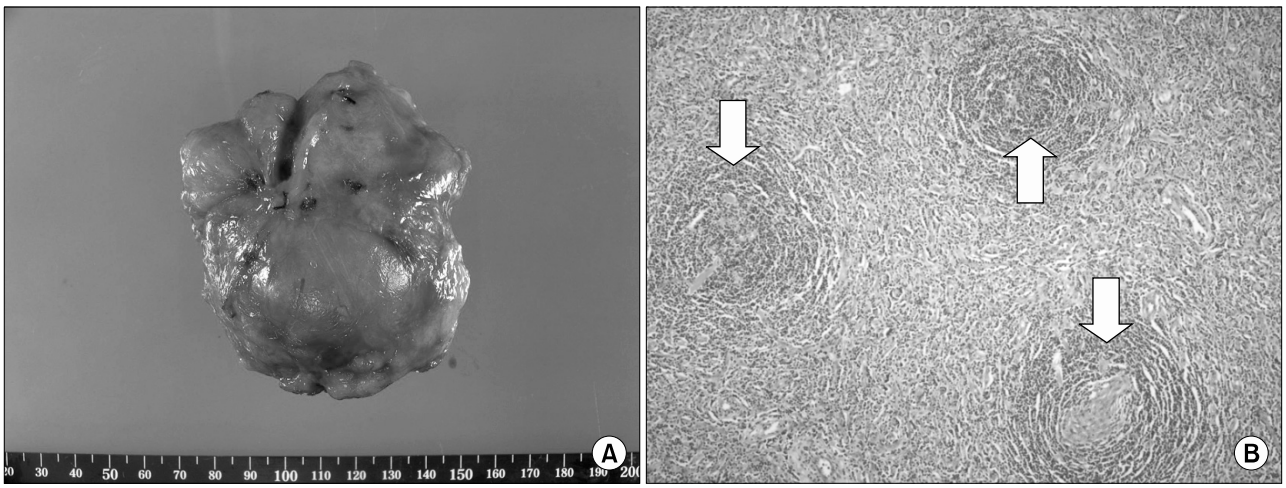

Fig. 3. (A) Grossly, the excised mass is well-circumscribed, with a solid gray cut surface, measured 8x6.5x5 cm and weighed 155 g. (B) Microscopically, there are prominent germinal centers showing well-developed vascular hyaline changes (H&E, x100) (white arrows).

sis of mass was CD of the hyaline vascular type.

The patient was discharged after 7 days from the operation without any complication. The patient is attending a follow up and shows no signs of abnormality.

DISCUSSION

Primary retroperitoneal tumors are rare, and more than 80% of them are malignant. Localized CD is radiologically nearly indistinguishable from malignant neoplasm, before histopathologic examination. Therefore if retroperitoneal mass is suspected then a biopsy should confirmed this after operation. Retroperitoneal CD should be added to the extensive list of differential diagnoses for tumors in the retroperitoneum.[5]

CD has been known as angiofollicular lymphoid hyperplasia, angiomatous lymphoid hamartoma, and giant mediastinal lymph node hyperplasia. It is characterized by enlarged hyperplastic lymph nodes histologically characterized by hyperplasia of lymphoid follicles and capillary proliferation with endothelial hyperplasia.

CD is a rare disorder that is found in young and healthy person regardless of sex.[6] Recently there has been a speculation of immunological mechanisms, including overproduction of interleukin-6 and human herpes virus type 8 infection.[7] A retroperitoneal malignant tumor was suspected in this index case, and therefore no test for IL-6 and herpes virus type 8 infection was conducted. Although the test is not accurate in the case of urology retroperitoneal mass, it is necessary for differential diagnosis.

The most common locations of CD are in the mediastinum (70%), neck, abdomen, axilla, shoulder, orbit, pelvis, pancreas, leptomeninges, vulvar, and retroperitoneum have also been reported as locations of CD.[3]

Keller et al[6] described two pathologic types of CD in 1972; hyaline-vascular type and plasma cell type. This may be further divided into localized form and systemic form according to the localization of the region. Hyaline-vascular type generally tends to be asymptomatic, but can be associated with mass effect symptoms, such as post-prandial discomfort, abdominal pain and weight loss. However, the clinical manifestations of plasma cell type CD are systemic symptoms, associated with fever, anemia, and fatigue. The hyaline vascular (HV) type is usually asymptomatic and radiologic chatacteristics of CD are not typical but it has some characteristics that are quite specific. This variant appears as a hypervascular mass with a strong enhancement and star-shaped microcalcifications on CT scan.[8] Grossly it appears as a well encapsulated homeogenous mass with a generally yellowish color. Microscopically, it is characterized by giant lymph follicles centered on a central vessel with marked hyalinization. The plasma cell (PC) type has the similar gross findings as HV type, but contains much more mature polyclonal plasma cells with a less marked hyalinization and vascularization.[3] The histology of PC type is not a specific systemic CD and can be found in autoimmune disease, AIDS and in lymph nodes draining carcinoma, so it is imperative to exclude this condition before diagnosing CD of PC type.[9]

Retrospectively, this patient showed negative HIV diagnosis, showed no sign of autoimmune disease and therefore a systemic type of CD could be ruled out. One peculiar point was that the large size of CD had severely tilted vena cave and hence problems of venous drainage was preliminarily suspected. However, no sign of such problem was shown. Also, the CD was detected through Rt flank pain and gross hematuria. However the dramatic decrease of symptoms after the natural discharge of urinary stone, shows that the CD was incidentally found along with urinary stone.

In the case of absence of preliminary diagnosis, open biopsy can be undertaken to assess the malignancy of the tumor. This can help to avoid extensive resection. With the exception of lymphomatous tumors, curability depends on the completeness of the surgical resection. Complete surgical excision of localized CD is curative and recurrences have been described only after incomplete resection. The prognosis is excellent with a five-year survival of nearly 100%.[3] There has been a report that radiotherapy is helpful in case a complete surgical excision has not been performed. But for the systemic CD, there is no curative optimal treatment methods yet. If surgical removal is possible, a complete excision will help.[10]

The present case has performed complete surgical excision of the CD mass and the patient has been showing no particular symptoms during the past 8 months of follow up and hence the illness is considered to be cured.

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