An Unusual Case of Retroperitoneal Xanthogranulomatous Pseudotumor

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Xanthogranulomatous pyelonephritis is a rarely described chronic inflammatory process characterized by partial or total destruction of the kidney due to parenchymal replacement by granulomatous tissue containing foamy, lipid-laden macrophages. The disease is associated with obstructive uropathy, most frequently secondary to the stone, resulting in a nonfunctioning enlarged kidney in most cases. We present an unusual case of xanthogranulomatous pseudotumor originated retroperitoneally without history of calculus or recurrent infections of urinary tract. (Korean J Urol Oncol 2013;11:102-104)

Key Words: Xanthogranuloma, Pyelonephritis, Retroperitoneal space

CASE REPORT

A 67-year old woman visited our hospital because of intermittent flank discomfort of several months’ duration. The patient had no history of urinary stone, symptomatic UTI or diabetes mellitus. Physical examination was unremarkable. Laboratory findings indicated a white blood cell count of 11,500/mm³, hemoglobin of 11.7 g/dl, and platelet of 339,000/mm³. Urinalysis was unremarkable. Contrast-enhanced computed tomography (CT) demonstrated an irregular soft tissue mass measuring 5 cm in maximum dimension with heterogeneous enhancement between adrenal gland and upper pole of left kidney. It was attached to the perinephric tissue with formation of abscesses and even fistulas. We report an unusual case of xanthogranulomatous pseudotumor originated from retroperitoneal space, which needed to be differentiated from mimicking a sarcoma radiologically.

Xanthogranulomatous pyelonephritis (XGP) is a chronic variant of pyelonephritis, in which normal renal tissue is replaced with chronic inflammatory cell infiltrate containing lipid-laden macrophages, resulting in an enlarged tumor-like kidney with decreased function. This condition is commonly seen in middle-aged female patients, especially diabetics. A higher incidence of about 18% has also been reported in the pediatric age range. Unlike chronic pyelonephritis, it may spread to the perinephric tissue with formation of abscesses and even fistulas. We report an unusual case of xanthogranulomatous pseudotumor originated from retroperitoneal space, which needed to be differentiated from mimicking a sarcoma radiologically.
Fig. 1. Contrast-enhanced computed tomography (CT) demonstrated a 5cm sized irregular soft tissue mass with heterogeneous enhancement between adrenal gland and upper pole of the left kidney. It was attached to the upper medial capsule of left kidney showing undefined boundary with psoas muscle, suggestive of sarcoma originated in the area.

Fig. 2. Positron emission tomography (PET)/CT showed an irregular hypermetabolic mass in medioposterior side of left perirenal space, suggestive of FDG-avid malignancy.

Fig. 3. Gross specimen showed an irregular yellowish fatty mass clearly demarcated from renal tissue.

Fig. 4. Microscopic examination reveals extensive accumulation of foamy histiocytes that represents for xanthogranulomatous inflammation in the perirenal soft tissue (haematoxylin and eosin stain, x200).

DISCUSSION

XGP is a chronic inflammatory lesion typically resulting in diffuse renal destruction with infiltration by sheets of lipid laden macrophages, admixed with acute and chronic inflammatory cells and frequent abscess formation. The disease is associated with obstructive uropathy secondary to the stone, resulting in a nonfunctioning, enlarged kidney in most cases.5,6 The cause of XGP remains undetermined, but is presumed to be related to calculus or non calculus urinary tract obstruction, ineffectively treated urosepsis, chronic renal ischemia causing localized alteration in renal metabolism, lymphatic obstruction, alteration in lipid metabolism and an altered immune response.5,7 Obstruction of the kidney with concomitant ipsilateral UTI is the most common etiology. The occurrence of XGP has rarely been reported in the absence of urinary tract obstruction or stone.

XGP is generally a disease limited to the affected kidney, but spread to adjacent tissues has also been seen. According to the extent of involvement of the adjacent tissue, Malek and Elder have classified this disease into three stages. Stage I:
Nephric. Disease confined to renal parenchyma only. Stage II: Nephric and perinephric. Disease process involves renal parenchyma along with perinephric fat. Stage III: Nephric and perinephric. Disease extending into adjacent structure or diffuse retroperitoneum.8

Reports of complications such as renocolic fistula and psoas/paranephric abscess have been described.9 The varied clinical presentation and paucity of definite diagnostic tests make the diagnosis difficult to confirm. However, studies have shown that clinical laboratory findings when combined with imaging modalities such as computed tomography scanning and magnetic resonance imaging scan can help to identify this fulminate kidney infection preoperatively.10

In this case, our patient did not have the typical clinical characteristics of XGP such as recurrent UTI, stones, and massive destruction of renal parenchyma replaced by granulomatous tissue. CT scan demonstrated that the xanthogranulomatous mass might be of extrarenal origin and the presence of sarcoma was strongly suspected. It was attached to the upper medial capsule of left kidney showing undefined boundary with psoas muscle, suggestive of sarcoma originated in the area. At laparotomy, a retroperitoneal mass, well demarcated from kidney but infiltrating into the psoas muscle, was found at the posterosuperior portion of left kidney.

REFERENCES