Case Report: Xanthogranulomatous Pyelonephritis - An Unusual Variety

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INTRODUCTION:

Xanthogranulomatous pyelonephritis is an uncommon inflammatory process characterized by focal or diffuse replacement of the renal parenchyma by yellow lipid contained macrophages [1].

Here, we would like to report a case of xanthogranulomatous pyelonephritis in which amorphous amount of fat was present surrounding the staghorn calculus in left renal sinus.

CASE REPORT:

A 30 years old woman patient came to hospital with complains of left sided flank pain, low grade fever and weight loss during last three months.

On clinical examination, lump was palpable in left lumber region.

Radiograph KUB was taken, showed large staghorn calculus in left renal area and other small multiple calculi noted inferolateral to the stag horn calculus. (Fig. 1)

Sonography performed with 3.5 MHz curvilinear probe showed huge pyonephrotic left kidney occupying whole left hypochondrium and lumber region with gross dilatation of upper and mid calyces with thinned out cortex with internal echoes in it. Large staghorn calculus noted in left renal sinus with marked echogenic area noted surrounding the calculus and renal sinus was not dilated. Other multiple small calculi were noted in left lower calyx. (fig.2)

FIGURE 1. Radiograph KUB showing staghorn calculus with other small calculi noted in left renal area.

FIGURE 2. USG Abdomen showing pyonephrosis with echogenic area surrounding calculus in left renal sinus.

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IVU was done, showed nonfunctioning left kidney with staghorn and other small calculi in left renal area.

Plain CT Scan of abdomen was performed. CT Scan showed hydronephrotic huge left kidney with gross dilatation of upper and middle calyces. Large dense staghorn calculus was present in left renal sinus with large areas of low attenuation (negative attenuation value -85 to -100 HU) noted surrounding it. Left renal sinus was contracted. Multiple small calculi noted in left lower calyx with fat density noted at corticomedullary junction in lower part. Posterior perirenal fat was involved by infection. (fig.3)

From above findings diagnosis of Xanthogranulomatous pyelonephritis was considered.

Subsequently MRI of abdomen was performed, which confirmed the findings of CT Scan. MRI showed upper pole of left kidney was not separately seen from left dome of diaphragm. (fig.4, 5, 6)

FIGURE 3. Plain CT Scan Abdomen showing area of fat density present is surrounding the dense calculus in left renal sinus.

FIGURE 4. T2 weighted coronal image of abdomen showing huge hydronephrotic left kidney occupying left abdomen with calculi in left renal sinus and left lower calyx.

FIGURE 5. T1 weighted axial image of abdomen showing fat surrounding calculus in left renal sinus.

FIGURE 6. T2 weighted STIR sequence of abdomen, axial image showing suppression of fat surrounding the calculus, appears hypointense.

FIGURE 7. Gross specimen of left kidney showing fat surrounding left renal sinus and calculi.
During operation, left kidney was found to be adherent to left dome of diaphragm, lateral part of transverse colon and left upper psoas muscle. Left kidney was removed except adherent portion. Calculi were removed and large amount of fat was present surrounding the staghorn calculus. (fig.7)

Histopathological report was xanthogranulomatosus pyelonephritis. Microscopically appearance was typical of xanthogranulomatosus pyelonephritis, showed abundance of lipid-laden macrophages, lymphocytes and plasma cells. (fig.8)

FIGURE 8. Microscopic appearance of specimen showing abundance of lipid-laden macrophages, lymphocytes and plasma cells.

DISCUSSION:

Xanthogranulomatous pyelonephritis is a chronic renal infection that in its most common form leads to scarred contracted renal pelvis, dilated calyces and diffuse infiltration of the renal parenchyma by plasma cells and lipid laden macrophages, which may form multiple yellow-colored masses. The term xanthogranulomatous describes the yellow color imparted to the renal parenchyma and inflammatory masses by the high lipid content of macrophages. Calculus in the pelvis of xanthogranulomatous kidneys occur in over 75 percent of cases. Calculus usually assumes staghorn shape and is composed of struvite. The dilated calyces, whose walls are thickened by inflammatory are filled with pus. Extension of the inflammatory process into psoas muscle and perirenal and pararenal spaces occurs frequently. Parenchymal calcification is uncommon[2].

There is marked female preponderance and 10 percent patients are diabetic[3]. There is accumulation of lipid laden macrophages (xanthoma cells) and a granulomatous infiltrate because of failure of local immunity[4]. Signs and symptoms include abdominal and flank pain, low grade fever, weight loss, and malaise. Many patients have no lower urinary tract symptoms. A renal mass may be palpable. Anemia, leukocytosis, pyuria and albuminuria may be present. Bacteria are cultured from the urine in approximately two third of patients. Proteus mirabilis and Escherichia coli are the organisms most likely to be found[2]. A history of urolithiasis is present in about 35 percent of the patients and hepatic dysfunction may be present in 50 percent of the patients[5].

In 85 percent the entire kidney is involved but disease may be focal. Computed tomography is very useful, because the findings on sonography and urography are nonspecific. On CT, xanthogranulomatosus pyelonephritis is associated with: (a) a large central calculus, often a staghorn; (b) enlargement of the kidney (or of a segment); (c) poor or no excretion of contrast into the collecting system; and (d) multiple focal low-attenuation(-10 to +30 HU) masses scattered throughout the involved portions of the kidney. The low attenuation collection represents dilated, debris-filled calyces and xanthoma collections. The collection themselves do not enhance, and there is no excretion of contrast, but there is bright enhancement of the rims of the collections, because of inflammatory hypervascularity. Perinephric extension occurs in about 14 percent and is well shown on CT, fistulae may develop and gas may rarely seen. Some variation occur: the kidney may be small, and calculi may be absent, making it difficult to distinguish xanthogranulomatosus pyelonephritis from other infections or neoplasm[6]. CT is particularly valuable in that it not only demonstrates characteristic renal findings, but also shows the extent of inflammation and extent into adjacent tissues. This will aid in surgical planning in choosing an approach that will provide adequate exposure and facilitate patient care[7].

In diffuse form of xanthogranulomatosus pyelonephritis, USG demonstrates multifocal enlargement of the kidney. The dominant findings reflects the markedly dilated calyces that are filled with products of inflammation and are seen as multiple, uniformly aligned hypo echoic structures with an internal pattern of fine echoes. The frequently present pelvic calculus causes a highly reflective image with acoustic shadowing. Parenchymal xanthogranulomatosus masses cause focal zones of echogenicity[2].

REFERENCES: