BILATERAL FOCAL XANTHOGRANULOMATOUS PYELONEPHRITIS : A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT

Xanthogranulomatous pyelonephritis (XPN or XGP) is an uncommon entity, mostly unilateral. A case of bilateral focal xanthogranulomatous pyelonephritis is reported with literatures reviewed. Accurate preoperative diagnosis is difficult ; however ultrasound and CT may give the suggestion of this disease.

INTRODUCTION

Xanthogranulomatous pyelonephritis (XPN or XGP) is a rare atypical form of severe chronic renal parenchymal infection. Its manifestation mimics with those of neoplasm and other renal parenchymal infection,and often were misdiagnosed clinically. XPN is usually associated with stone formation (50-70%) and obstruction secondary to staghorn calculi. There is increased incidience in middle-aged female and diabetic.Pathologically, the kidney is enlarged with yellowish nodules containing Lipid-laden macrophages (Xanthoma cells). 80-90% of these kidneys have diffuse involvement. The disease is almost always unilateral and has no predilection for either side. Bilateral involvement which were previously reported in literatures, is only 8 cases.[9]

CASE REPORT

A 44-year-old female presented with on-and-off fever for 2 months.She had a feeling of fullness at right upper abdomen for 2 weeks,and lost of weight, 10 kg., within 2 months.She also had amenorrhea for 4 months.

Physical examination disclosed hepatomegaly and positive bimanual palpation at right side of abdomen.

Laboratory findings revealed anemia and leukocytosis.Urinalysis showed 1+ proteinuria and 1-2 white blood cells per high powered field.There was 2+ urine sugar. Mild elevation of BUN, Creatinine were noted and blood sugar was normal. Hepatic function test revealed chronic liver disease. Negative test for α - fetoprotein was noted.

Sonography revealed mixed echogenic mass with reniform shape (12 cm.in length) at right upper abdomen, inferior to the liver. (Fig.1) The mass had well defined border and the internal echo exhibited multiple foci of dense echoes with posterior acoustic shadowings. Upper pole of right kidney was spared. There was also 4 cm. cystic mass at upper pole of left kidney. The mass invaginated into the spleen, but not invading. Hepatomegaly with two silent gallstones are revealed. The sonographic diagnosis was renal cell carcinomas and Xanthogranulomatous pyelonephritis to be differentiated.

Liver scan with Tc^{99m}-SC showed hepatomegaly with decreased liver uptake at postero-inferior portion of right lobe liver.

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Excretory urography demonstrated a large intrarenal mass,involving nearly the entire right kidney, except the upper pole which some excretory function was preserved. The renal calyces were displaced and had distortion. There was normal size and excretory function of left kidney, with bulging renal outline at the upper pole and stretching of renal calyces. (Fig.2)

Plain CT scan revealed enlarged right kidney with hypodensity (12-16 HU.) mass, approximately 10 cm. in diameter, replacing almost the entire renal parenchyma sparing only the upper pole. (Fig.3 a) The renal outline was poorly defined, which indicated perinephric involvement of the disease. Left kidney was bulging at lateral aspect from relatively low density (16 HU.) mass, 3 cm. in diameter. (Fig. 3 b) No internal calcification or abnormal gas was noted.

Contrast enhancement CT scan showed no significant enhancement of the hypodense masses of both kidneys. The masses were more obvious due to peripheral rim enhancement of the lesions and there was also enhanced thick internal septations. The right renal mass involved ipsilateral right psoas muscle.The inferior vena cava and renal veins were normal. (Fig. 4 a,b)

Angiography revealed relatively hypovascular mass at lower pole of right kidney with neovascularization. Enlargement of renal capsular branch indicated perirenal extension was noted. (Fig. 5 a) There was cortical nephrogram defect at upper, lateral border of left kidney without any tumor vessel. (Fig. 5 b) The Inferior Vena Cava was patent. Both CT scan and angiography were suggestive of malignancy.

Right total nephrectomy demonstrated a large renal mass involving middle and lower portions of right kidney with direct extension outside renal capsule into the surrounding tissue including muscles. The mass grossly appeared firm and had yellowish nodules. Accidentally ruptured of the mass during the operation revealed yellowish pus, without any foul smell. Pus culture was negative, but hemoculture was positive for Klebsiella Pneumoniae.

Post-operative ultrasonographic guidance for percutaneous aspiration of left renal mass was failed.

Histologic study revealed thin renal parenchyma, hyalinized glomeruli,and fibrotic changes of renal pelvis, calyces and interstitium. Wall of the mass was composed of solid sheets of foamy histiocytes. (Fig. 6) There was also submucosal chronic inflammation of the ureter.

The conclusive diagnosis was bilateral XPN, focal form ,stage III (paranephric).

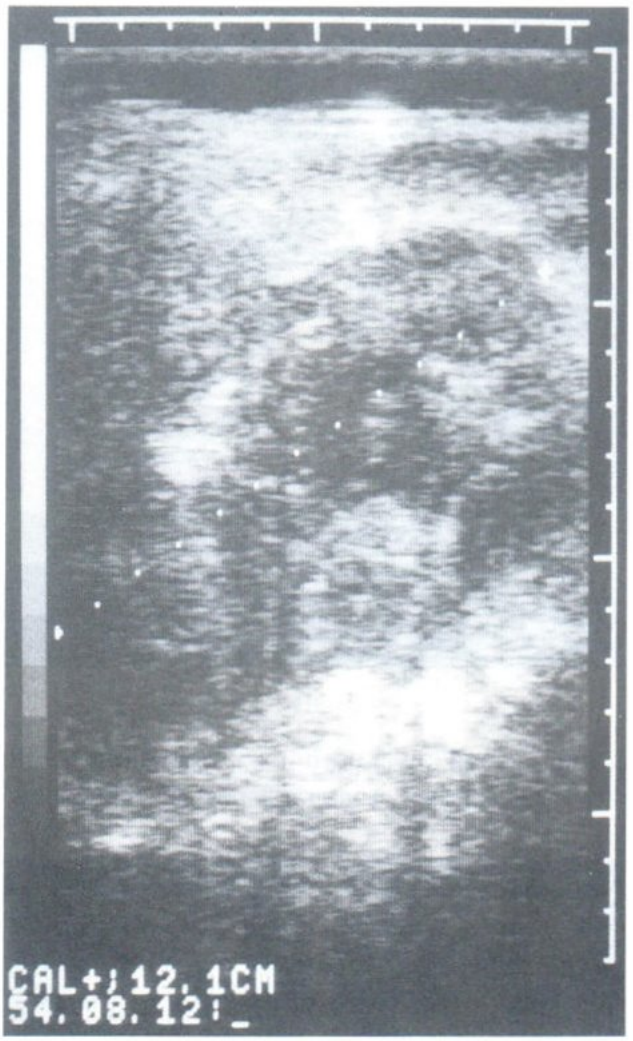


Fig. 1 Renal ultrasonography of right kidney shows a large mixed echogenic mass with dense internal echoes involving mid and lower portions with upper pole sparing.

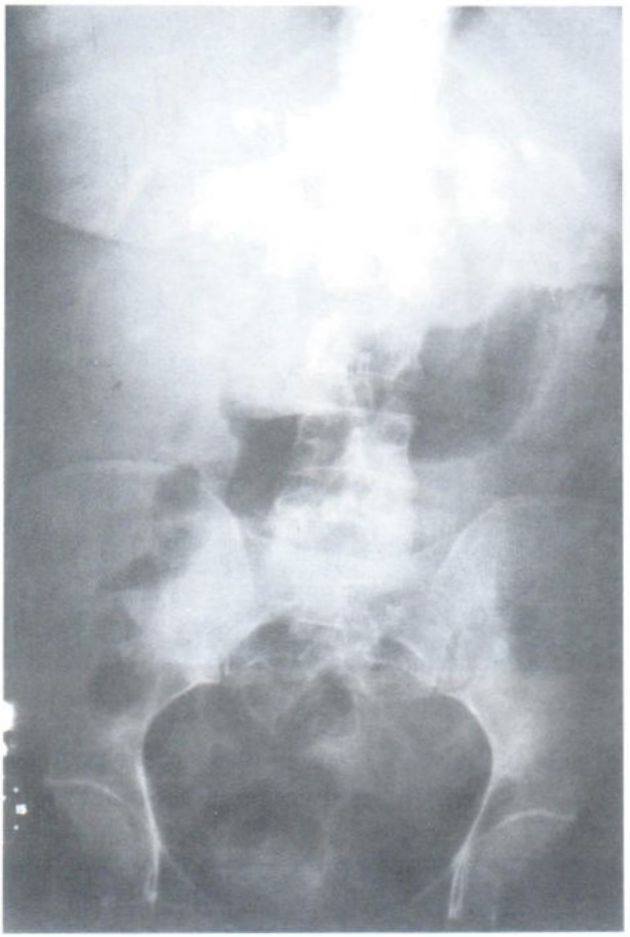
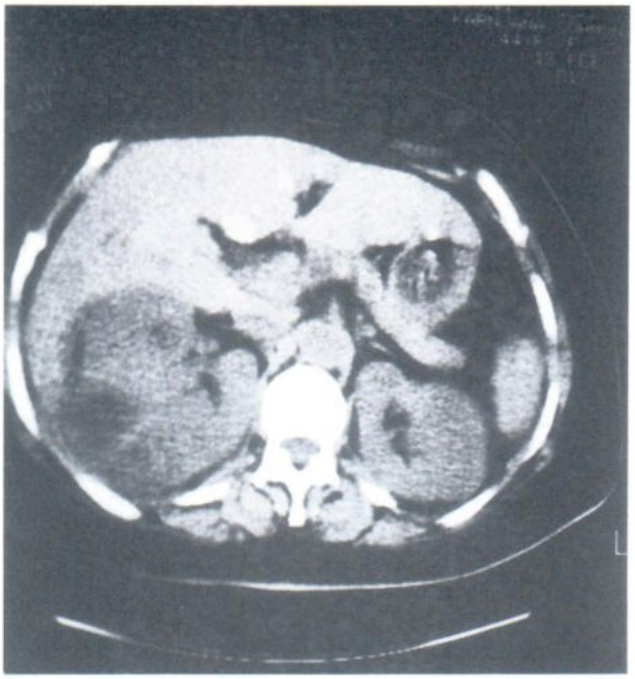
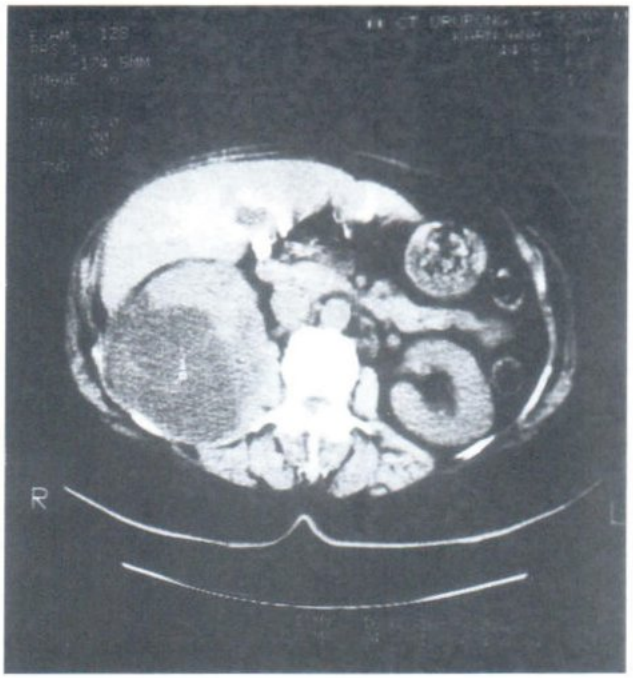


Fig. 2 Film IVP reveals a large intrarenal mass involving nearly entire right kidney with distortion of upper pole calyces and bulging renal outline of upper pole of left kidney.

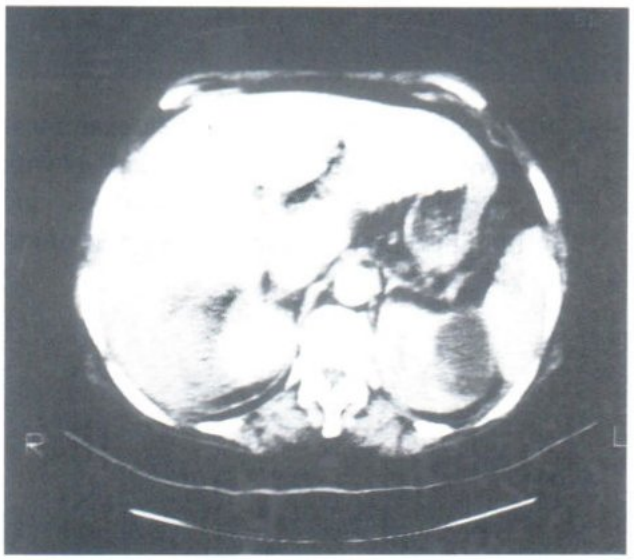


3A



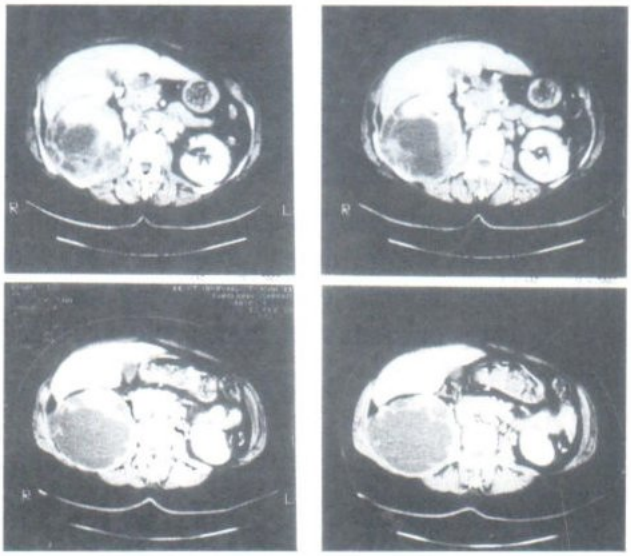
3B

Fig. 3a,b Plain CT scans show a large hypodensity mass at mid and lower portions of right kidney, with perinephric involvement. Smaller low density mass is noted at lateral aspect of left kidney.



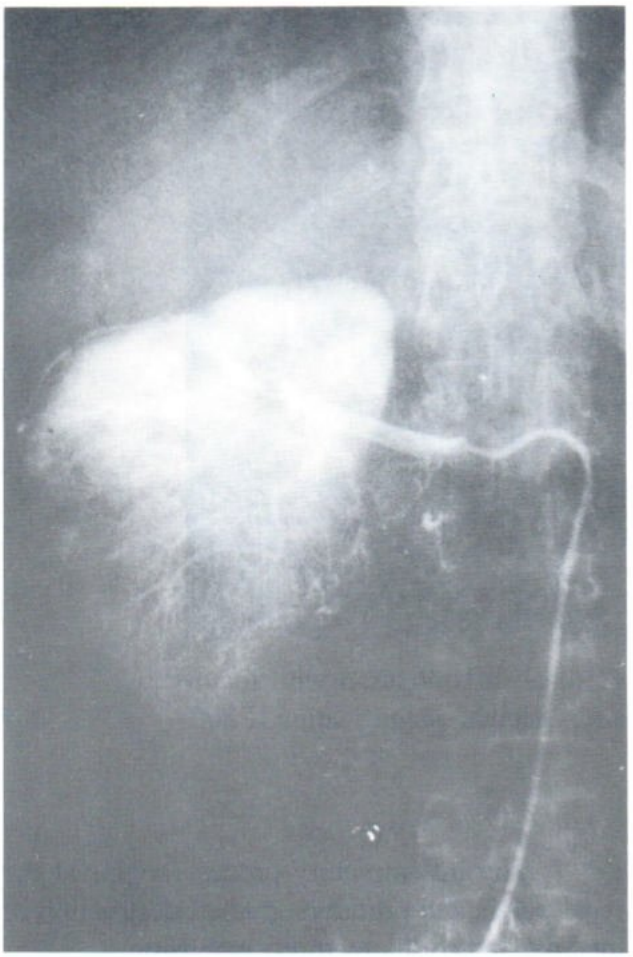
4A

Fig. 4a Contrast enhanced CT scan at level of left kidney shows non-enhancing hypodense mass at the lateral portion.



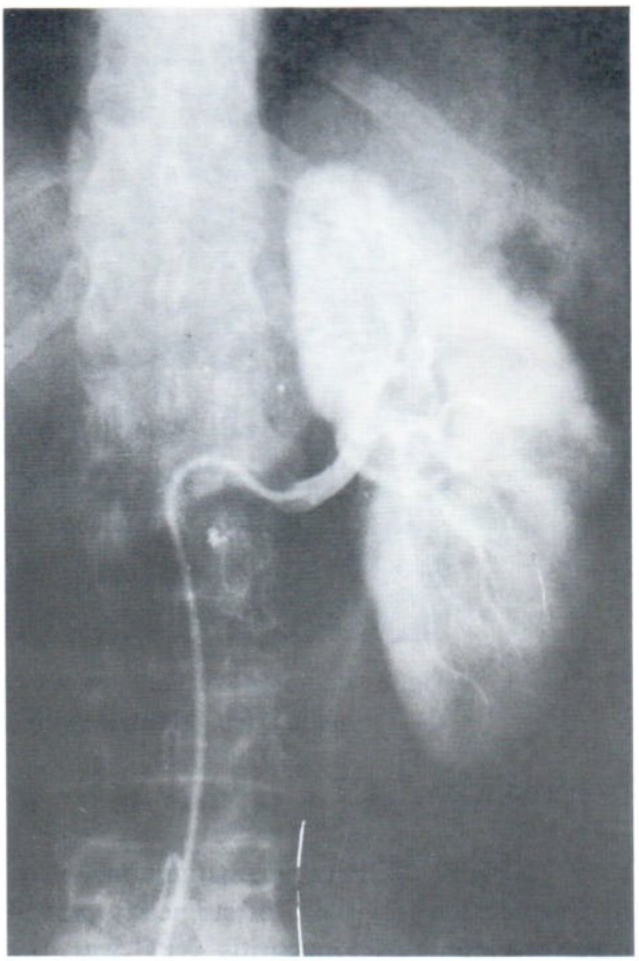
4B

Fig. 4b Contrast enhanced CT scans at multiple levels of right kidney reveal peripheral rim enhancement of non-enhancing hypodensity mass which has enhanced thick internal septations and also involves right psoas muscle.



5A

Fig. 5a Right renal angiography demonstrates a large hypovascular mass with neovascularization, at lower half of right kidney and also reveals enlargement of renal capsular artery.



5B

Fig. 5b Left renal angiography shows a cortical nephrogram defect at upper lateral aspect of left kidney.

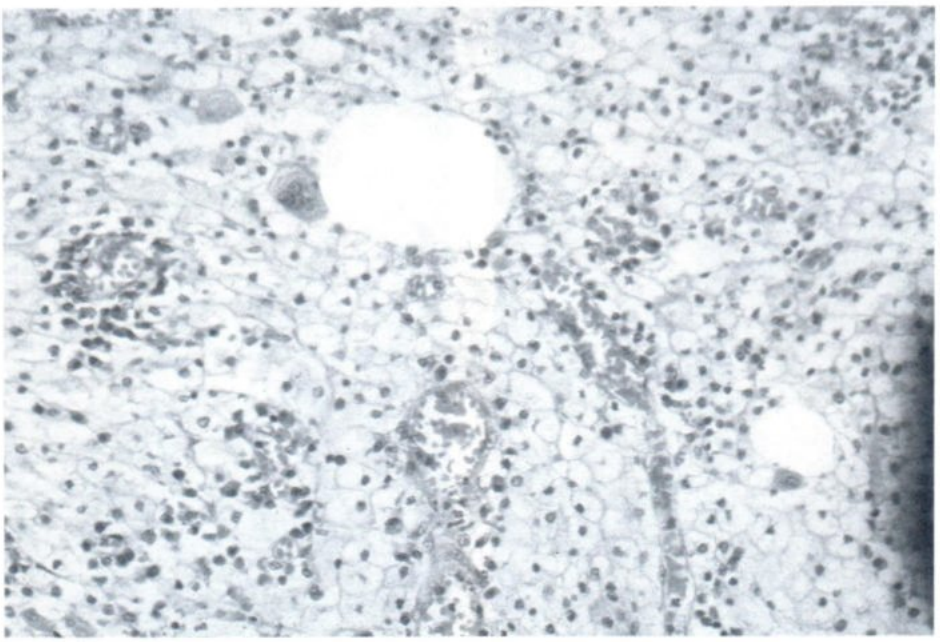


Fig. 6 Pathological slide shows chronic renal inflammation with foamy histiocytes. (Hematoxylin & eosin ; Original magnification X 200)

DISCUSSION

XPN or XGP is an uncommon entity, responsible for only 0.6% of surgically proved cases of chronic pyelonephritis. XPN was first described in 1916 by Schlagenhaufer. He used the term " Staphylomykosen " because of the macroscopic resemblance to Actinomycosis granuloma and the microscopic appearance of Staphylococci. Since 1916 to 1987, nearly 500 cases have been described in the literatures. It has apparently increased in frequency in more recent years.

ETIOLOGY

The etiology of XPN is unknown and usually associate with urinary tract obstruction and calculous disease (50-70% of cases). Abnormal lipid metabolism, lymphatic blockage, chronic renal ischemia with resultant localized alteration in renal metabolism, altered immune response, DM and finally, calculous or non-calculous urinary tract obstruction with prolonged antibiotic therapy have been suggested.

The inflammatory process begins at the renal pelvis and produces gradual destruction of medulla and cortex by direct extension.

XPN is almost always unilateral involvement, and equal incidence on each side. Bilateral involvement have been found only in 8 cases.[9] XPN is predominantly found in female (58-78%), with a peak of age incidence at 4^{th}-6^{th} decade of life.

TYPE AND STAGING

Type of involvement is divided into three forms.

1. Diffuse : This is the most common form (80-90%) and it is associated with staghorn calculi. It represents an entire renal involvement.

2. Segmental : The condition is confined to one pole of duplex kidney or the parenchyma which drains into an obstructed calyx.

3. Focal : It is also refered to as a Xantho-

granuloma. The inflammation usually originates within the renal parenchyma and does not communicate with the collecting system. In this case there will be a nonspecific solid or cystic renal mass.

There are three stages of XPN :

Stage I (nephric) : Inflammatory process was confined to the kidney.

Stage II (perinephric) : Involving kidney and gerota fascia

Stage III (paranephric) : Involving affected kidney and its surrounding fat with widespread retroperitoneal involvement

CLINICAL DATA

The presenting symptoms are generalized and composed of flank pain, flank mass, weight loss, malaise, fever, mild lower urinary tract symptoms and draining flank sinus.

The physical examinations of XPN are non-specific, such as flank mass and tenderness, fever, anemia, hepatomegaly, pyuria and hypertension.

Duration of the symptom is usually six months to one year. Several associated conditions have been held responsible for XPN, including renal stones (50-70%). hypernephroma, DM, multiple pregnancy and previous Urinary Tract Infection. No significant association with any systemic disease is noted.

BACTERIOLOGY

Bacteriology finding on urine or resected specimens mostly yielded Escherichia coli and Proteus mirabilis.

LABORATORY FINDINGS

No laboratory tests are specific for the condition ; anemia, leukocytosis, elevated ESR, pyuria, hematuria, proteinuria, rising BUN, Creatinine, and abnormal liver function test (nephrogenic and hepatic dysfunction).

ROENTGENOLOGIC FINDINGS

Plain film : The affected kidney is often enlarged and has staghorn renal calculi (or uretero-pelvic junction stone, or ureteric stone). The renal outline and psoas shadow are sometime ill-defined because of perinephric edema or infiltration (perirenal extension).

Intravenous pyelography : The stone bearing (70%) and functionless (80%) kidney have been the most frequent findings. The other findings are space occupying lesions, or intrarenal masses (focal form) with distortion of renal collecting system, or renal outline. Diffuse renal enlargement with transient rim nephrogram, indicating hydronephrosis, may be presented. Perinephric extension of the disease process produces ill -defined renal margin or a nondescripted renal mass.

Retrograde pyelography : The study may disclose obstruction at uretero-pelvic junction with gross dilatation of pelveocalyceal system. On occasion, irregular filling defect in a contracted renal pelvis with marked renal caliectasis may be noted.

Renal scintigraphy : The study generally reveals non-uptake of the affected kidney and no excretion.

Ultrasonography : In diffuse form;the affected kidney is enlarged and contains multiple cystic areas, with internal fine echoes. The renal calyces may be surrounded by thin zone of increased echogenicity. There may be central strong echogenic foci, with acoustic shadowings, correspond to staghorn renal calculi.The other findings

are hydro-or pyonephrosis,and echogenic areas due to the solid foci of granulomatous tissue;as well as debris and calculi. Sometime,the inflammation may extend into perirenal space or psoas muscle

Focal form reveals hypoechoic mass with irregular border and calculi in renal collecting system.

Computerized tomography : Diffuse form usually discloses a calculus in renal pelvis or collecting system with absence of contrast excretion in the involved kidney or focal area of i nvolvement. The other findings include (1) multiple non-enhancing rounded areas within medullary space (resemble hydronephrotic pattern) (2) discrete solid masses (3) area of fat density (4) diffuse renal enlargement with preserve reniform outline (5) the lesion may extend beyond the expected confines of the kidney into the perirenal space, pararenal space, psoas muscle, back muscle, diaphragm, colon or spleen.

Focal form demonstrates localized water-density mass adjacent to calyx, that contains calculus.

Angiography : Diffuse form reveals sparse, narrowed, and stretched intrarenal arteries around foci of granulomatous tissue. The intrarenal arterial pattern may resemble the neovascularity of hypernephroma. Renal capsular arteries are often enlarged, reflecting extrarenal extension of the inflammatory process.

Angiographic findings of focal form XPN usually resemble the diffuse form,with additional finding of irregular defect in nephrogram.

Four cases of renal vein thrombosis were previously reported.

PATHOLOGICAL CHARACTERISTIC

Macroscopic findings : - The kidney is usually enlarged with yellowish nodules and dilated collecting system containing purulent material. Perinephric inflammation and fibrosis may be present.

Microscopic findings : - The disease is characterized by Lipid-laden macrophages (Xanthoma cell) and granulomatous reaction. The abscess, focal calcification and cholesterol slits may be revealed.

The histology of XPN may resemble clear cell type of renal cell carcinoma.

PREOPERATIVE DIAGNOSIS

The correct preoperative diagnosis is range from 5-44% and the remaining patients were thought to have malignant tumors (hypernephroma, epidermoid carcinoma,or retroperitoneal tumor), hydronephrosis or pyonephrosis, renal tuberculosis, or renal abscess.

TREATMENT AND PROGNOSIS

The treatment of choice is nephrectomy (total or partial). Some patients were treated by percutaneous drainage before the major operation, due to extensive retroperitoneal involvement.

XPN doesnot seem to recur,metastasize, or involve contralateral kidney, and the prognosis is excellent. However, some of the conditions, associated with the disease, such as bacteremia and hypertension, may continue to be a problem.

CONCLUSION

XPN or XGP is an atypical, uncommon form of severe chronic granulomatous renal infection in chronic obstruction,and almost always

unilateral involvement. We reported one case of bilateral XPN which was one among small number of cases in worldwide literatures, and probably the first reported case of bilateral XPN in Thailand.

ABBREVIATION:

XPN, XGP = Xanthogranulomatous Pyelonephritis

REFERENCES

1. Albert Solomon, Zvi Braft, Joseph Papo, Ephraim Merimsky. Computerized tomography in Xanthogranulomatous Pyelonephritis. J Urol 1983;130:323-25
2. Bala R. Supramanyam, Alec J. Megibow, B. Nagesh Raghavendra, Morton A. Bosniak. Diffuse Xanthogranulomatous Pyelonephritis : Analysis by computed tomography and sonography. Urol Radiol 1982;4:5-9
3. Bhupendra M. Tolia, Alfredo Iloreta, Selwyn Z. Freed, Bernard Fruchtman, Boyce Bennett, Harry R. Newman. Xanthogranulomatous pyelonephritis: Detailed analysis of 29 cases and a brief discussion of atypical presentation. J Urol 1981;126: 437-42
4. F.R. Smith. Bilateral Xanthogranulomatous pyelonephritis. B J Urol 1981;53:81
5. H. Claes, R. Vereecken, R. Oyen, B. Van Damme. Xanthogranulomatous Pyelonephritis with emphysis on Computerized tomography scan. Urology 1987;29:389-93
6. I. Husain, A. Pingle, T. Kazi. Bilateral diffuse Xanthogranulomatous Pyelonephritis. Br J Urol 1979;51:162-63
7. J.C. Gingell, J. Roylance, E.R. Davies, J. B. Penry. Xanthogranulomatous Pyelonephritis. Br J Radiol 1973;46:99-109
8. Mason Goodman,Thomas Curry, Thomas Russell. Xanthogranulomatous Pyelonephritis (XGP): Local disease with systemic manifestation; Report of 23 patients and review of the literature. Medicine 1979; 58:171-81
9. Mohammed Akhtar, Wajeh Qunibi. Bilateral Xanthogranulomatous Pyelonephritis involving native kidneys in a renal transplant recipient: Association with renal cell carcinoma and amyloidosis. Am J of Kidn dis 1992;20(3):289-93
10. Owen C. Van Kirk, Raymundo T. Go, Victor J. Wedel. Sonographic features of Xanthogranulomatous Pyelonephritis. AJR 1980;134: 1035-39
11. Reza S. Malek, Jack S. Elder. Xanthogranulomatous Pyelonephritis: A critical analysis of 26 cases and of the literature. J Urol 1978;119:589-93
12. R.G. Grainger, A.J. Longstaff, M.A. Parsons. Xanthogranulomatous Pyelonephritis : A reappraisal. Lancet 1982;19: 1398-1401
13. Stanford M. Goldman, Davis S. Hartman, Elliot K. Fishman, Joseph P. Finizio, Olga M.B. Gatewood, Stanley S.Siegelman. CT of Xanthogranulomatous Pyelonephritis : Radiologic-pathologic correlation. AJR 1984;142:963-70