XANTHOGRANULOMATOUS PYELONEPHRITIS- A CASE REPORT

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ABSTRACT

Xanthogranulomatous pyelonephritis (XGP) is a rare entity and constitutes less than 1% of chronic pyelonephritis. It is a serious, chronic inflammatory disorder of the kidney characterized by a destructive lesion that invades the renal parenchyma. A 28 year old female who is a known case of right pyonephrosis underwent right nephrostomy and drainage of pus one month back. Urinalysis showed combined hematuria and pyuria. Intravenous pyelogram revealed no excretion of contrast in both kidneys. U.S.G abdomen showed mild right Hydronephrosis with drain tube insitu. The patient was admitted to our Hospital for right nephrectomy and the specimen was sent for histopathological examination. On microscopy, it was diagnosed as Xanthogranulomatous pyelonephritis.

KEYWORDS: Xanthogranulomatous pyelonephritis, nephrectomy, Chronic pyelonephritis.

INTRODUCTION

Xanthogranulomatous pyelonephritis (XGP) is a variant of chronic pyelonephritis which is frequently associated with urinary tract obstruction and nephrolithiasis.[2] It is four times more common in women than in men and is usually noted in the fifth and sixth decades of life. XGP affects both kidneys with equal frequency.[3,4] XGP is characterized by diffuse or focal,
chronic, and severe renal parenchymal infection leading to destruction and replacement of the renal parenchyma by lipid-laden macrophages (xanthoma cells) which impart yellowish tan to the tissue. We report one such case in a 28 year old patient.

**CASE REPORT**

A 28 year old female presented with recurrent fever and right flank pain. She is a known case of left nephrolithiasis and right pyonephrosis underwent right nephrostomy and drainage of pus one month back. Complete blood count revealed neutrophilia. Urinalysis showed hematuria and pyuria. Intravenous pyelogram revealed no excretion of contrast in both kidneys (FIG-1). U.S.G abdomen showed mild right hydronephrosis with drain tube in situ (FIG-2). The patient underwent right nephrectomy for a non functioning kidney and the specimen was sent for histopathological examination.

Grossly, the specimen was weighing 255 g, measuring 11x7x5 cm in size. External surface was nodular with multiple scars (FIG-3). Cut section- Cortex was tan yellowish with coarse, discrete, corticomedullary scars and necrotic debris in the overlying dilated, blunted calyces (FIG-4).

Histopathological examination showed renal parenchyma largely replaced by sheets of foamy histiocytes, lymphoplasmacytic infiltration and foreign body giant cell reaction (FIG-6). Viable renal parenchyma showed contraction of tubules with atrophied lining epithelium. Few dilated tubules contained eosinophilic casts. Glomeruli were atrophied and few showed sclerosis. Interstitial tissue was fibrosed with dense lymphocytic infiltration. Blood vessel walls are thickened and hyalinised. Few areas showed micro abscesses and necrosis (FIG-5). There is no evidence of malignancy in the stromal or epithelial components. Gross morphology and microscopic features are suggestive of XanthoGranulomatous Pyelonephritis.

![Figure 1: Intravenous Pyelogram showing no excretion in both kidneys](image-url)
Figure 2: Ultrasonogram abdomen showing right hydronephrosis with drain tube in situ.

Figure 3: External surface showing nodularity with multiple scars.

Figure 4: Cut section showing corticomedullary scars and tan yellowish areas.
DISCUSSION

XGP is a serious, chronic inflammatory disorder of the kidney associated with indolent bacterial infection. Schlangenaufer first described the disease in 1916.[5] Xanthogranulomatous pyelonephritis (XGP) is a rare entity and constitutes less than 1% of chronic pyelonephritis. [1] XGP is seen in approximately 1% of all renal infections, is four times more common in women than men and is usually noted in the fifth and sixth decades of life, although most cases of XGP are unilateral, bilateral disease has been reported and is generally fatal.[11,9] The disease process is characterized by an infectious phlegmon which begins in the renal pelvis and extends into the medulla and cortex which are gradually destroyed and replaced by lipid laden macrophages (Xanthoma Cells) which impart yellowish
tan to the tissue; the infective process then can extend into the extra-renal tissues, either focally or diffusely.\textsuperscript{[7,9,10]}

Xanthogranulomatous pyelonephritis (XGP) is a variant of chronic pyelonephritis which is frequently associated with urinary tract obstruction and nephrolithiasis.\textsuperscript{[2]} Examination of the urine confirms the presence of urinary tract infection. The urinalysis reveals pyuria and bacteriuria.\textsuperscript{[3,4]} Urine culture typically demonstrates enterobacteriaceae. The most common organisms associated with XGP are E. coli, Proteus mirabilis, Pseudomonas, Enterococcus faecalis and Klebsiella species.\textsuperscript{[13]} The urine cultures are sterile in 25 percent of cases.\textsuperscript{[14]} Urine culture was sterile in our case.

Xanthogranulomatous pyelonephritis is most frequently misdiagnosed as renal cell carcinoma due to its clinical presentation and radiographic appearance.\textsuperscript{[8]} Evidence of chronic urinary tract infection and CT scan findings usually make it easier to differentiate these disorders. Although rarely, the two disorders can occur together.

CT scan is now the best tool to diagnose these infections, and it is also important to establish the presence and extension of extra-renal involvement.\textsuperscript{[4,6]}

The lipid-laden xanthomatous cells may mimic the clear cells of clear cell RCC. Foamy histiocytes may also present extensively in papillary RCC. The xanthomatous cells have a foamy cytoplasm compared with the More clearer cytoplasm of clear cells. Adequate sampling should reveal granulomatous inflammation of XGP and the papillary structures of papillary RCC. Other differential diagnoses include other types of RCC, Malakoplakia, tuberculosis, and renal abscess. Malakoplakia of the kidney is primarily a disease of the renal pelvis with involvement of the renal parenchyma, and Michaelis- Gutmann bodies are characteristic of the lesion\textsuperscript{[16]} Renal tuberculosis is diagnosed by characteristic epithelioid granulomas with central caseous necrosis. Abscess can be differentiated by necrosis surrounded by neutrophils and absence of granulomas.

The gold standard therapy for XGP is nephrectomy. Nephrostomy before nephrectomy can be considered a method that facilitates surgery, because it allows a reduction in renal mass and favours the access to the kidney at the time nephrectomy is done. Antibiotics alone are not effective for these infections, but should be initiated before surgical procedure in order to
control the infectious process and avoid systemic involvement. In our case, nephrostomy followed by nephrectomy was done

CONCLUSIONS

Xanthogranulomatous pyelonephritis is an unusual variant of chronic pyelonephritis. Most cases occur in the setting of obstruction due to infected renal stones. Affected patients usually have massive destruction of the kidney due to xanthogranulomatous reaction. The radiological and gross appearance may be confused with renal malignancy. The confirmatory diagnosis of this entity is based on histopathological examination. Nephrectomy remains the treatment of choice in almost all the cases.

REFERENCES


