

Xanthogranulomatous pyelonephritis: A rare entity

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Abstract

Context: Xanthogranulomatous pyelonephritis is an extremely rare but known entity resulting from prolonged suppuration of the kidney. The disease may be focal or diffuse and may pose considerable diagnostic dilemma at times. Surgery is curable and remains optimal treatment of choice. Owing to its rarity and clinical curiosity we report such a rare case encountered in our clinical practice. **Case Report:** We report a case of non-functioning kidney suggestive of either Tuberculosis or xanthogranulomatous pyelonephritis in the pre-operative period depending upon the clinical aqumen and investigations available and nephrectomy was done. It was confirmed as xanthogranulomatous pyelonephritis histopathologically. **Conclusion:** Preoperative diagnosis of xanthogranulomatous pyelonephritis may pose difficulty and nephrectomy remains the treatment of choice especially in diffuse cases of xanthogranulomatous pyelonephritis.

Keywords: Xanthogranulomatous pyelonephritis, tuberculosis, non-functioning kidney, nephrectomy.

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Introduction

Xanthogranulomatous pyelonephritis (XPN) is a variant of chronic pyelonephritis which is frequently associated with urinary tract obstruction or nephrolithiasis. Pre-operatively it may mimic renal tuberculosis or renal carcinoma due to its vague clinical presentation, equivocal laboratory and radiological investigations [1]. The confirmatory diagnosis of this entity is based on histopathological examination and surgery remains the treatment in almost all the cases. We report an unusual case of XGP occurring in a patient of nephrolithiasis with non functioning kidney.

Case Report

MS 40 yrs male presented with pain in left flank, fever off and on and malaise for 3 months. Abdominal examination revealed a tender vague mass in the left lumbar region. Laboratory investigations showed increased white cell counts with normal renal function tests. Routine urinary evaluation showed increased number of pus cells but urine culture was sterile. USG abdomen revealed enlarged left kidney with multiple echogenic foci with no corticomedullary differentiation. Intravenous

pyelography was showing non functioning left kidney with multiple renal and ureteric calculi and extensive renal parenchymal calcification. All these features were suggestive of either tuberculosis or XPN (Figure 1). Nephrectomy was done and specimen was sent for histopathology examination (Figure 2). Cut section of the specimen revealed distorted architecture of the kidney, corticomedullary junction was not identifiable, the pelvi-calyceal system was dilated with presence of staghorn calculus in the pelvis. Microscopically there were polymorph leucocytes, foamy histiocytes, lymphocytes, plasma cells and foreign body giant cells. Hence a diagnosis of XPN was made. The postoperative period was uneventful and the patient was doing well in the follow up period of 6 months.

Discussion

Xanthogranulomatous pyelonephritis (XPN) is a rare entity and constitutes less than 1% of chronic pyelonephritis. It mainly occurs in adults with preponderance in females of younger age group. It usually involves single kidney and is associated with renal stones in 2/3 of cases and concomitant infection with *E.*

coli, *Proteus mirabilis*, *Klebsiella spp*, *Staphylococcus aureus*, *Enterococcus spp*, *Pseudomonas spp*, *Streptococcus spp*, including anaerobic organisms [2].



Fig. 1 Intravenous pyelography showing non-functioning left kidney with multiple renal calculi with extensive renal parenchymal calcification.



Fig. 2 Intraoperative picture showing xanthogranulomatous left sided kidney.

Symptoms are frequently nonspecific and include fever, flank pain, weight loss, malaise, anorexia, and constipation. Other uncommon presentations include urinary symptoms like dysuria, frequent urination and hematuria or rarely a discharging cutaneous fistula originating at the level of involved kidney. Physical examination usually reveals an

ill-defined, palpable and tender flank mass. However, less than one-half of patients may present with hypertension or hepatomegaly [3].

Although none of the laboratory or radiological investigations are confirmatory. Diseases like renal tuberculosis may mimic XPN. However the definitive diagnosis of this entity is achieved only by histopathological examination which shows the presence of lipidladen macrophages (xanthoma cells), as well as other inflammatory cells, including plasma cells, leukocytes, and histiocyte [4, 5]. Antibiotics usually do not resolve the problem and nephrectomy either partial or total remains the treatment of choice [6].

Conclusion

XPN is a rare cause of chronic pyelonephritis resulting in non-functioning kidneys and poses a preoperative diagnostic dilemma which may mimic other diseases but whenever encountered it should be treated with optimal surgical removal.

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