ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 02 | February - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A CASE SERIES OF XANTHOGRANULOMATOUS PYELONEPHRITIS WITH UNUSUAL CLINICOPATHOLOGICAL SPECTRUM

Pathology

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ABSTRACT

Xanthogranulomatous pyelonephritis is a rare entity which constitutes 0.6% to 1% of all pyelonephritis cases and 1 % of all kidney infections. It is an unusual condition of the kidney resulting in parenchymal destruction and chronic inflammatory reaction. It closely resembles malignancy and differentiating it is extremely significant in therapeutic and prognostic aspects. Here we report three cases of xanthogranulomatous pyelonephritis due to its rarity, diagnostic dilemma and unusual clinical presentation.

KEYWORDS

Xanthogranulomatous pyelonephritis, foamy histiocytes, touton giant cells, granuloma, great imitator.

INTRODUCTION:

Xanthogranulomatous pyelonephritis is a rare entity which constitutes 0.6% to 1% of all pyelonephritis cases and 1 % of all kidney infections.[1,2] It is an unusual chronic inflammatory condition of the kidney resulting in destruction and replacement of renal parenchyma with chronic inflammatory cells including foamy histiocytes, hemosiderin-laden macrophages and occasional giant cells. This condition is common among middle-aged females, especially in patients with diabetes.[2] It may result in abscess formation after spreading to the perinephric tissue resulting in fistula formation.[3] Surgery with nephrectomy is the only definitive management. Here we report three cases of xanthogranulomatous pyelonephritis due to its rarity, diagnostic dilemma and unusual clinical presentation.

Case Report:

Case 1: A 53 year old female presented with complaints of left loin pain radiating to back for two months, with history of vomiting for one day. She was a known case of diabetes for two months, not on medication. She also had treatment for ureteric stones with right ureteroscopy and laser stone fragmentation (URSL) with DJ stenting before two months. Vitals were normal and baseline investigations were done which showed anemia with hemoglobin of 9g/dl. Urine culture showed presence of E.coli infection. CT Abdomen showed a peripheral enhancing thick walled, lobulated cystic lesion in upper/mid pole cortex with few internal enhancing septations and suspicious solid component with calcifications described as left renal complex Bosniak cyst III/IV with bilateral renal calculi and renocolic fistula [Fig 1a]. PETCT-MRI also done which showed an exophytic complex lesion with peripheral metabolic activity. Surgery was performed with left radical nephrectomy and segmental descending colon resection with fistula repair was done as upper pole of kidney was found adherent to descending colon and the specimen was sent for frozen section to the histopathology department. Under microscopy in the frozen section, it was very difficult to rule out malignancy as there were sheets of spindle cells with xanthogranulomatous inflammation which looked like tumor cells[Fig1b]. Also since the adjacent bowel structures were involved, it raised the suspect of invading high grade tumor. In routine histopathology sections, it was known that those spindle cells were fibroblasts. Macroscopically left kidney with perinephric fat measured 10x5.5x5 cm which had ill-defined tan to yellowish area measuring 2.7x2.5x2.5 cm located in the midpole with a segment of descending colon [Fig1c]. Microscopy revealed renal parenchyma with sheets of foamy macrophages, mutinucleated giant cells (touton giant cells), granulomas, chronic inflammatory infiltrate along with numerous microabscesses and extensive areas of fibrosis [Fig1d, 2a, 2b]. Immunohistochemistry was done which showed negative for Pan-cytokeratin so that the possibility of carcinoma was ruled out [Fig2c]. Section from the colonic serosa also showed xanthogranulomatous reaction [Fig2d]. Final report of xanthogranulomatous pyelonephritis with xanthogranulomatous reaction of descending colon was made.

Case 2 and Case 3: The other two cases presented with similar features. Both of them were females, diagnosed clinically as non-functioning kidney with nephrolithiasis. Radical nephrectomy was done in both cases and histopathological examination showed features suggestive of xanthogranulomatous pyelonephritis [Fig3a, 3b, 3c, 3d].

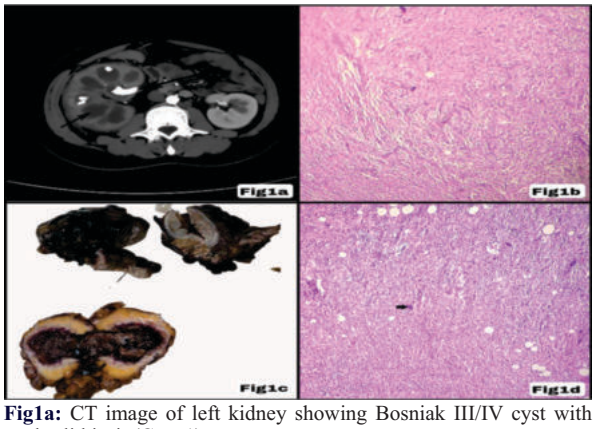

Fig1a: CT image of left kidney showing Bosniak III/IV cyst with nephrolithiasis (Case 1)
Fig1b: Sheets of spindle cells with xanthogranulomatous inflammation- 400X (Case 1)
Fig1c: Cut surface of left kidney and part of descending colon (Case1)
Fig1d: Foamy histiocytes with touton giant cells- 400X (Case 1)

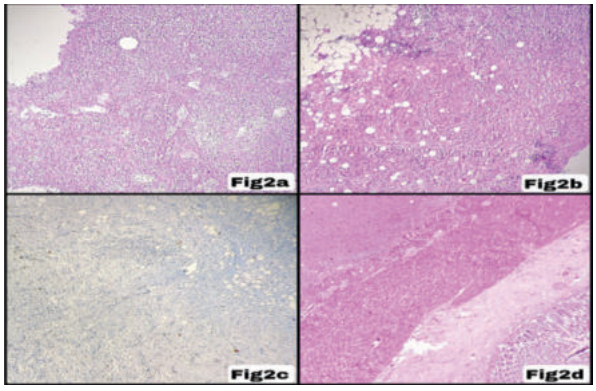

Fig2a: Nidus of microabscesses- 200X (Case 1)
Fig2b: Granulomas and touton giant cells with adjacent perinephric fat- 200X (Case1)
Fig2c: IHC showing Cytokeratin negativity- 200X (Case1)
Fig2d: Xanthogranulomatous reaction of colonic serosa- 200X (Case1)

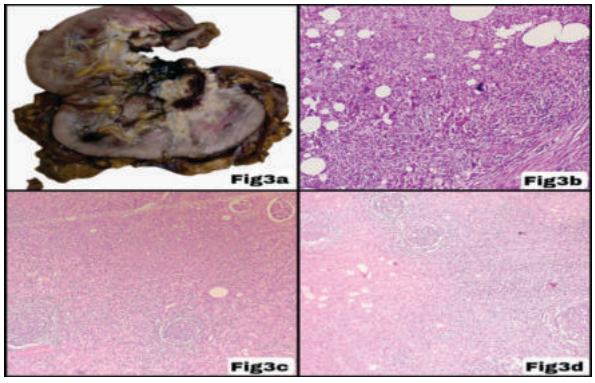

Fig3a: Cut surface of left kidney (Case2)
Fig3b: Foamy histiocytes with touton giant cells- 400X (Case 2)
Fig3c: Multiple lymphoid follicles with normal and sclerosed glomeruli- 200X (Case3)
Fig3d: Focal xanthogranulomatous reaction with lymphoid follicular hyperplasia- 400X (Case3)

DISCUSSION:

Xanthogranulomatous pyelonephritis (XGP) is also known as the great imitator or the great masquerader as it closely resembles the malignancy in clinical, radiological and histopathological manifestations.[4] Though the pathophysiology of XGP remains unclear, most important causes of XGP are presumed to be infection and chronic obstruction.[1] XGP is most commonly associated with calyceal stones and staghorn calculi, recurrent urinary tract infections, chronic interstitial nephritis and also obstructive conditions such as pyeloureteric junction obstruction, ureteropelvic duplication, ureteral schistosomiasis and tumors including renal and transitional cell carcinomas.[5] The most common organisms encountered in urinary tract infection are Escherichia coli and Proteus mirabilis, rarely Pseudomonas and Klebsiella also seen. XGP can occur in three forms-focal, segmental and diffuse. Diffuse XGP is the most common form and shows three stages radiologically. In stage I, the lesion is confined to the renal parenchyma (Nephric XGP). In stage II, the lesion involves perirenal space with extension into the Gerota's fascia (Perinephric XGP). In stage III, the lesion extends into pararenal space and retroperitoneal structures (Paranephric XGP).[6] In our first case, nidus of microabscesses and pan-cytokeratin negativity strongly excludes the possibility of carcinoma. In the third case, there were multiple lymphoid follicles and focal xanthogranulomatous reaction with lymphoid follicular hyperplasia which are usually not seen in the XGP. Eventhough there were simulating and perplexing pathology, with the evidence of some key features, XGP can be differentiated from other conditions.[4] The most common differential diagnosis are malakoplakia which has Michaelis-Gutmann bodies and renal clear cell carcinoma. It can be treated medically mainly in children, but surgery with nephrectomy either by open or laparoscopic remains the mainstay of the treatment. Nephron-sparing partial nephrectomy is done in focal XGP, whereas total nephrectomy should be considered in case of focal XGP.[6]

CONCLUSION:

Xanthogranulomatous pyelonephritis is a great mimicker which simulates malignancy and differentiating it is extremely significant in therapeutic and prognostic aspects. It had an unusual presentation with associated involvement of the colonic serosal region (case 1) which rendered the diagnosis difficult. The histologic heterogenity of the three cases also adds to the diagnostic dilemma. Early diagnosis and treatment can prevent the complications of this condition. Surgery with nephrectomy and excision of the surrounding affected tissue proved to be curative and sufficient with an excellent prognosis.

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