Xanthogranulomatous Pyelonephitis in a male child with renal vein thrombus extending into the inferior vena cava: A Case Report

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ABSTRACT

Background
We present a case of Xanthogranulomatous pyelonephritis (XGPN) in a male child with renal vein thrombus extending into the inferior vena cava. This is a rare presentation. XGPN is a rare type of renal infection characterised by granulomatous inflammation with giant cells and foamy histiocytes. The peak incidence is in the sixth to seventh decade with a female predominance. XGPN is rare in children. A 11 year old male child presented with a history of high grade fever and chills for 15 days, right flank pain, progressive pyuria for two months and a history of vesical calculus. In our case, a subcapsular right nephrectomy was performed.

Methods
The surgical specimens were formalin fixed and paraffin embedded. The sections were stained with routinary Hematoxylin & Eosin.

Results
Grossly, the kidney was enlarged with adherent capsule and thickening of the perinephric tissue. The pelvicalyceal system was dilated and was filled with a cast of pus. Histological evaluation revealed diffuse necrosis of the renal parenchyma and perinephric fat. Neutrophils, plasma cells, sheets of foamy macrophages and occasional multinucleate giant cells were seen. The renal vein was partially occluded by an inflammatory thrombus with fibrin, platelets and mixed inflammatory cells. The thrombus was focally adherent to the vein wall with organization.

Conclusions
The clinical presentation and the macroscopic aspect, together with the histological pattern, the cytological characteristics addressed the diagnosis towards XGPN with a vena caval thrombus. Our case illustrates that the diagnosis of XGPN should be considered even in paediatric age group when renal vein and vena caval thrombi are present.

Key words: Xanthogranulomatous pyelonephritis, renal vein thrombus, vena caval thrombus
XGPN in a male child with a renal vein thrombus extending into the inferior vena cava is an extremely rare presentation. XGPN is a chronic renal infection characterised by destruction and replacement of the renal parenchyma with sheets of lipid laden macrophages, admixed acute and chronic inflammatory cells and frequent abscess formation\(^1\). The peak incidence is in the sixth to seventh decade with a female predominance. XGPN is rare in children\(^2\). We report a case of XGPN in a male child presenting with a vena caval thrombus.

**CASE PRESENTATION**

A 11 year old male child presented with a history of high grade fever with chills for 15 days, right flank pain and progressive pyuria for two months. He had a prior history of surgery for vesical calculus and anterior urethral calculus for which he underwent open cystolithotomy and urethrolithotomy four years back.

On examination, the child was emaciated, febrile with pallor. There was diffuse tenderness in the right lumbar region with renal angle tenderness. X-ray showed radio-opaque shadow in the right lumbar region measuring 2 x 2cm. Abdominal computerised tomography (CT) revealed a dilated pelvicalyceal system with necrotic debris at the lower pole of right kidney and a thrombus in the renal vein extending into the infrahepatic vena cava. Pyonephrosis, parenchymal calcification along with marked retroperitoneal and porta hepatis lymphadenopathy were also seen on the CT. Blood urea and serum creatinine were within normal limits. Gram negative bacilli was isolated from thick pus aspirated from the right renal pelvis.

Open nephrostomy was done with only pus and no urinary drainage. Following a renal biopsy diagnosed as chronic pyelonephritis (end-stage changes), the patient underwent subcapsular right nephrectomy.
METHODS

The surgical specimens were formalin fixed and paraffin embedded. The sections were stained with routine Hematoxylin & Eosin.

RESULTS

Gross appearance

Grossly, the subcapsular nephrectomy specimen weighed 155gms and measured 10 x 6.5 x 5cm. The cut surface showed a dilated pelvicalyceal system filled with a cast of pus. Adjacent to the pelvis and calyces yellow areas were seen. The renal cortex was thinned out. The capsule and perinephric tissue were thickened with adhesions.

Microscopic appearance

Histological evaluation revealed diffuse necrosis of the renal parenchyma and perinephric fat. Neutrophils, plasma cells, sheets of foamy macrophages and occasional multinucleate giant cells were seen. The renal vein was partially occluded by an inflammatory thrombus with fibrin, platelets and mixed inflammatory cells. The thrombus was focally adherent to the vein wall with thrombus. Adjacent areas showed histologic features of chronic pyelonephritis.

The clinical presentation and the macroscopic aspect, together with the histological pattern, the cytological characteristics addressed the diagnosis towards XGPN with a vena caval thrombus.

DISCUSSION

Xanthogranulomatous pyelonephritis (XGPN) is a severe, atypical chronic renal parenchymal infection accounting for 6/1000 surgically proved cases of chronic pyelonephritis[2]. Women are affected more frequently than men with a peak incidence in the sixth to seventh decades[3]. XGPN is rare in children. It has been suggested that the pediatric variant of xanthogranulomatous
pyelonephritis differs from that of adults and, furthermore, sexual and racial differences in pediatric disease expression may exist\[^2\]. Changes of XGPN have been described in kidneys destroyed as a result of pyonephrosis, renal cell carcinoma, transitional cell carcinoma and rarely a renal cyst\[^4\]. Several interrelated etiological factors are thought to be responsible for the pathogenesis of XGPN.

They include calculus or non-calculus urinary obstruction, ineffectively treated urosepsis, chronic renal ischemia causing localized alteration in renal metabolism, lymphatic obstruction, alteration in lipid metabolism and an altered immune response. The extent of the pathologic process within the affected kidney varies. Two forms of XGPN have been described, a diffuse or global form (83-90%) as in our case, and a focal form (10-17%)\[^3\]. In the rare localized form, the lesion can be confined to one or other pole. More commonly it is a diffuse process involving whole kidney leading to reduced renal function. XGPN has been termed the great imitator because it may be misdiagnosed as a renal neoplasm especially if the lesion is focal. The most common presenting features are intermittent high grade fever, flank pain, loin pain, hematuria and vague gastrointestinal symptoms. Proteinuria and pyuria are frequent. The organisms isolated on urine culture are Proteus mirabilis, Escherichia coli, Staphylococcus aureus, Klebsiella, Pseudomonas and enterobacter species. Urine culture may show negative results in 30-39% of patients despite positive results from kidney\[^4\]. Mostly one kidney is affected. In a few cases contralateral kidney is enlarged due to compensatory hypertrophy. XGPN has been described in three stages. The lesion is confined to kidney in stage I as in our case, extends to Gerota’s space in stage II, to the paranephric space and other retroperitoneal structures in stage III\[^3A\]. Treatment of XGPN is partial or total nephrectomy\[^5\].

**CONCLUSION**

Xanthogranulomatous pyelonephritis is a common histological variant of surgically managed end stage pyelonephritis. In paediatric patients with this process clinical and pathological findings are similar to those in adults. Pathogenesis and, thus, time to establishment of this process have not been clearly defined and may be of a shorter interval than currently believed. Given an appropriate clinical and radiographic presentation, and without regard to patient age or duration of symptoms, the diagnosis of xanthogranulomatous pyelonephritis should be made prospectively. Our case illustrates
that the diagnosis of XGPN should be considered even in paediatric age group when renal vein and vena caval thrombi are present.

COMPETING INTERESTS

None declared

AUTHORS’ CONTRIBUTION

Reecha Singh conceived the idea of the study, helped to draft the manuscript and prepared the illustrations.

Geetanjali Gupta drafted the manuscript and carried out the literature search.

Pradeep Kumar helped to draft the manuscript and acquire histological images for illustration.

Dhananjay S. Kotasthane was responsible for overall coordination and final proofreading of the manuscript.

All the above mentioned authors read and approved the final manuscript.

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LEGENDS

**Figure 1-** Gross photograph
Specimen showing dilated calyces filled with purulent material and adjacent yellow areas.

**Figure 2-** Hematoxylin and Eosin staining (200x)
Showing sheets of fat laden macrophages and occasional multinucleate giant cells. Thick arrow indicates sheets of fat laden macrophages and Thin arrow indicates dense chronic inflammatory cells.