XANTHOGRANULOMATOUS PYELONEPHRITIS [XGP]

DR VILESH VALSALAN

CONSULTANT NEPHROLOGIST AND TRANSPLANT PHYSICIAN

ACADEMIC CORDINATOR – EXTRACORPOREAL NEPHROLOGY GROUP

INTRODUCTION

1. XGP is a chronic granulomatous inflammatory disorder of the kidney in which rena l parenchyma is extensively replaced by foamy lipid laden macrophages (xanthoma cells), along with chronic inflammatory cells and f ibrosis.
2. Xanthogranulomatous pyelonephritis was first described by Schlagenhaufer in 1916 and later named xanthogranuloma by Osterlin in 1944.

RISK FACTORS :

- Middle-aged adults, females (slightly higher incidence than males)
- Patients with chronic urinary obstruction, renal calculi, or a history of recurrent uri nary tract infections.
- Individuals with immunosuppression or metabolic disorders such as diabetes mell itus.

CLINICAL PRESENTATION

Symptoms :

1. Flank pain , fever and malaise. Weight loss and anorexia in prolonged cases Urinary symptoms such as dysuria or hematuria
2. Occasionally, a palpable mass over the kidney region or signs - systemic infection may be present.

Investigations :

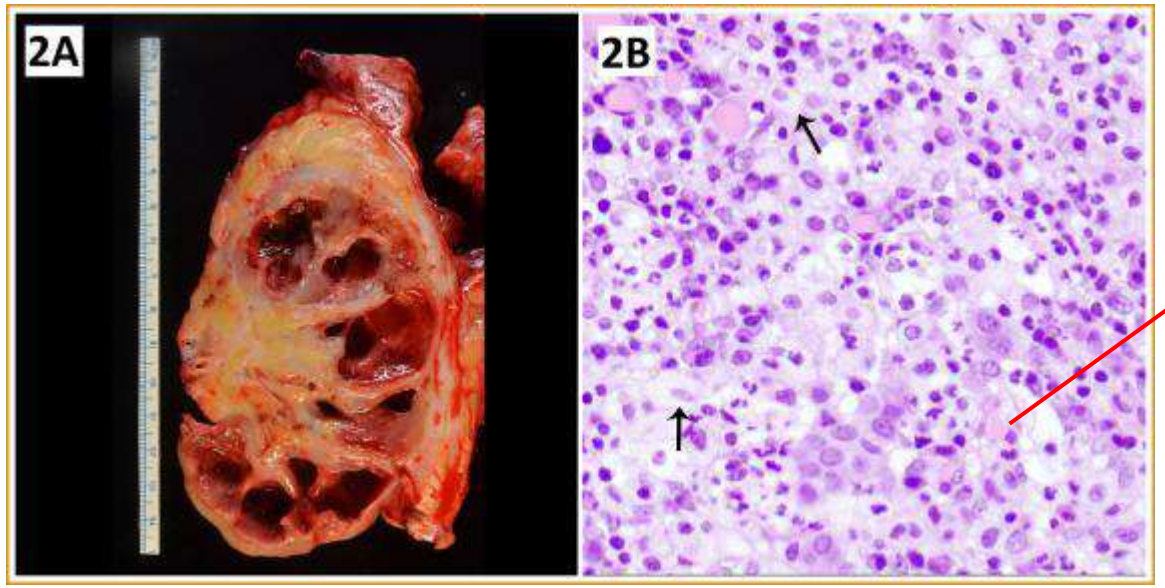
- **Laboratory tests** : elevated white blood cell count, anemia and deranged renal functions.
- **Urine culture.**
- **Imaging** : **CT scans** - enlarged kidney with areas of low density (due to lipidladen cells), calyceal dilatation, obstructing stones, and somet imes perinephric extension.
- Definitive diagnosis is confirmed histologically after nephrectomy, showing lipid-laden macrophages and chronic inflammation.

SAILIENT POINTS

- **XGP** is often confused with a true neoplasm, most commonly renal cell carcinoma, due to its similar clinical and radiographic characteristics.
- The organisms most commonly associated with **XGP** are *Escherichia coli* and *Proteus mirabilis*, followed by *Pseudomonas*, *Enterococcus faecalis*, *and Klebsiella*.
- Other abnormalities : ureteropelvic duplication, ureteropelvic junction syndrome, vesicoureteral reflux, and bladder cancer, are also associated with **XGP**.
- About 80% of patients have a staghorn stone.

1a A CT abdomen and pelvis with intravenous contrast showing severe left-sided hydronephrosis with renal cortical thinning, dilated renal calyces and non-obstructing calculi in the collecting system.

1 b BEAR PAW SIGN



2 a Gross pathology exam - an enlarged kidney with dilated calyceal system .

2 b Histopathology :
revealed chronic pyelonephritis with lipid-laden Macrophages.

TREATMENT

- XGP is generally resistant to conservative medical therapy.
- Surgical nephrectomy is the standard treatment, especially in diffuse XGP, to eradic ate infection and prevent complications.
- Preoperative or postoperative broad-spectrum antibiotics are used to manage infection.
- Partial nephrectomy nephron-sparing surgeries - when possible, especially in children.

COMPLICATIONS:

- Perinephric abscess or fistula formation.
- Sepsis or systemic infection.

Prognosis of XNG : is significantly improved. Unilateral cases have a much better prognosis.

Classification of XNG

- Xanthogranulomatous pyelonephritis is classified into focal, segmental, and diffuse forms :

- Focal: Involvement is localized within the cortex of the kidney, observed in about 20% of cases.

- Segmental: Involvement is limited to a regional or segmental area of the kidney.

- Diffuse: The most common presentation, characterized by widespread and diffuse renal involvement.

- Stage 1 (Nephric): The disease is limited to the kidney.
- Stage 2 (Perinephric): The disease involves the renal pelvis or the perinephric fat within Gerota's fascia.
- Stage 3 (Paranephric): The disease involves a wider area, including adjacent organs or the retroperitoneum.