Double Bear Paw : A Case of Complicated Xanthogranulomatous Pyelonephritis

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Citation

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Abstract

Xantholomatous Granulomatous Pyelonephritis (XGP) is a rare variant of chronic pyelonephritis, initially described in 1916 by Schlagenhaufer as staphylomycosis due to the histological findings of staphylococci, foam cells, leucocytes and fibroblasts. It is an acute on chronic inflammatory process characterised by replacement of the renal parenchyma with yellowish looking lipid laden macrophages[1]. It has been known to mimic other conditions such as renal neoplasms and renal tuberculosis[2,3]. The classic radiological finding of XGP is of a 'Bear Paw' due to the dilated renal calyces and atrophied and infected renal parenchyma.
We present the case of a patient with a unilateral XGP with an additional complication which resulted in the appearance of a Double Bear Paw.

BACKGROUND

Xantholomatous Granulomatous Pyelonephritis (XGP) is a rare variant of chronic pyelonephritis, initially described in 1916 by Schlagenhaufer as staphylomycosis due to the histological findings of staphylococci, foam cells, leucocytes and fibroblasts. It is an acute on chronic inflammatory process characterised by replacement of the renal parenchyma with yellowish looking lipid laden macrophages[1]. It has been known to mimic other conditions such as renal neoplasms and renal tuberculosis[2,3]. The classic radiological finding of XGP is of a 'Bear Paw' due to the dilated renal calyces and atrophied and infected renal parenchyma.
We present the case of a patient with a unilateral XGP with an additional complication which resulted in the appearance of a Double Bear Paw.

CASE REPORT

The patient was a 46 year old male, who is a long term nursing home resident due to a ruptured cerebral aneurysm 10 years prior. He is cognitively intact but is quadriplegic and has a long term urinary catheter. He initially presented with pyrexia of unknown origin and weight loss of 20kg over 6 months. His white cell count was 20.6 x 109 /L with 83% neutrophils. A CT scan performed by the treating medical unit (see images) demonstrated a markedly hydronephrotic kidney with a large loculated retroperitoneal collection traversing the posterior abdominal wall musculature. He was subsequently referred to the Urology Unit at the Repatriation General Hospital.
Initial management was incision and drainage of the subcutaneous abscess, which yielded over 1 litre of purulent material and multiple calculi, which on culture grew Proteus mirabilis. The cavity was packed with ribbon gauze for a period of 4 weeks prior to open nephrectomy through a flank incision. Histology was consistent with XGP and multiple calculi ranging from 10 to 35mm in size.
Post operative course was uncomplicated and the patient returned to hospital 4 months later for a planned cystolitholapaxy at which time he reported that he had regained 15kg of weight.

Figure 1

CT Scan Images – Transverse and Coronal Slices demonstrating a Double Bear Paw sign, renal calculi and an incidental bladder stone.

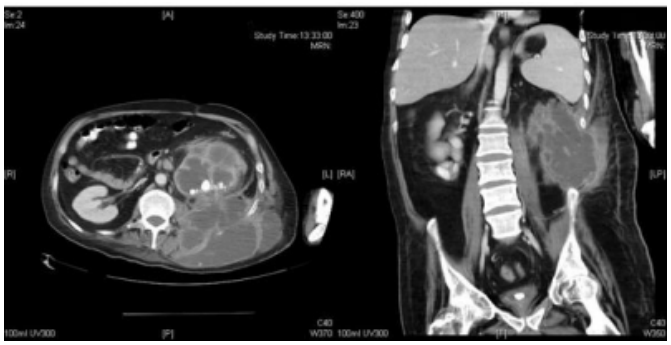
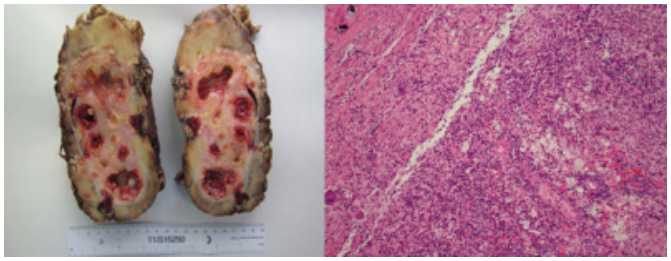


Figure 2

Histopathological images- Macroscopic bivalve section and Light Microscopy



DISCUSSION

XGP predominantly affects adults, but has been reported in children mainly below 8 years old. It has a female preponderance 3:1 in adults, with a male preponderance in children[5]. It can be focal or diffuse with focal XGP more common in paediatric cases. It is usually unilateral but rare cases of bilateral XGP have been described[3,5]. About 80% of patients are generally unwell for about a month or more, with non specific symptoms such as weight loss, fever and anorexia. Preceeding lower urinary tract symptoms such as dysuria, frequency, urgency and haematuria are often absent, hence the possible misdiagnosis with other conditions such as renal neoplasms[6-8].

Obstruction and infection of the collecting system are the 2 main features predisposing to XGP. Calculi causing obstruction has been described in > 50% of cases, with other forms of obstruction,eg.pelviureteric junction(PUJ)obstruction less commonly observed[3,6,7]. Positive urine cultures with E. coli and Proteus sp. are the commonly observed organisms in XGP in about 70% of cases. Raised inflammatory markers (leucocytes, C-reactive protein, erythrocyte sedimentation rate) are the main abnormality in laboratory findings[8,9]. Radiological findings with plain film and USS are usually inconclusive with USS demonstrating an enlarged and distorted renal outline, with loss of the normal renal architecture. CT scan demonstrates the most accurate and helpful finding of “bear paw sign” on cross sectional images.The renal pelvis is contracted whereas the calyces are dilated, resembling the toe-pads of the bear paw[10,11].

Tissue biopsy for histology provides confirmation of the diagnosis. Macroscopically there is gross distortion of architecture of renal parenchyma with or without the collecting system. There is replacement of the renal parenchyma by areas of yellowish nodules with or without central areas of necrosis, likely extending into perirenal fat. Microscopically as described above, there is a heavy infiltrate of foam cells - yellowish looking macrophages with lipid filled vacuoles[6,8,12].

A staging system by Malek and Elder has been in existence since 1978 ; stage 1(Nephric XGP) is classified as disease confined to kidney(20-46%), stage 2 involves kidney and perirenal fat(14-70%) and stage 3 is widespread retroperitoneal involvement(10-36%)[6,7]. In this case of XGP, extension into subcutaneuous paravertebral tissue and musculature is beyond what Malek and Elder described as stage 3 XGP, and a modification of the staging system to include Stage 4 could be considered.

XGP can be complicated by risk of hypertension, persistent bacteriuria, amyloidosis and postoperatively by duodeno/colocutaneous fistulae[6].

Renal unit salvage is generally not possible, however there have been rare cases of nonsurgical management with antibiotics and anti-inflammatory agents, mainly in children with focal XGP. Nephrectomy is treatment of choice, usually total, but nephron sparing(partial nephrectomy) surgery has been described in certain cases of focal XGP[8].

In this case removal of the easier collection (subcutaneous) was undertaken prior to nephrectomy. Once the infective process was removed, the patient demonstrated a good recovery with return to his baseline level of function, indicating that surgical management with nephrectomy or partial nephrectomy if appropriate, are the main stay of treatment of XGP.

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