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Images in Nephrology
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Can your hunch get this punch?

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A 34 year-old Hispanic female with a history of nephrolithiasis presented with right flank pain and fever. A non-contrast computer tomography (CT) showed a 4.1 × 3.9 cm cystic lesion in the right kidney lower pole and a 6 mm non-obstructive renal calculus. She was treated with cephalexin for *Escherichia coli* (*E. coli*) urinary tract infection. One year later she returned to the emergency room with recurrent flank pain, fever, positive right renal punch and a bulge on her right flank (Figure 1 white arrow). A new contrast CT of the abdomen showed several fluid collections (Figure 2 black arrow) with septations located in the right kidney forming a 'bear paw sign' (Figure 2 white arrow inset). The largest collection (6.7 × 8.6 cm) had a sinus tract to the posterior lateral abdominal wall. The collections were drained by interventional radiology and grew *Lactobacillus* species. She subsequently underwent a right total nephrectomy. Pathology of the kidney showed a small tan-yellow, well circumscribed, pseudo-encapsulated, nodular mass

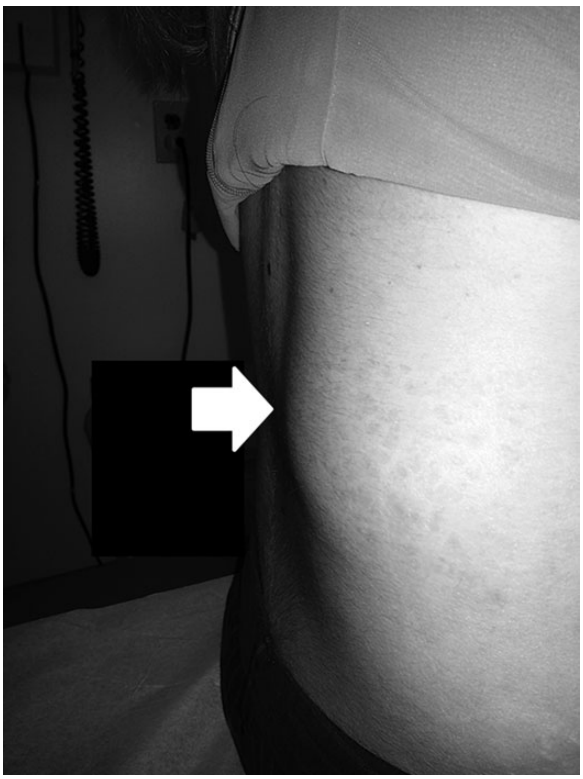

Fig. 1. Patient's back showing the right bulge noted on physical examination.

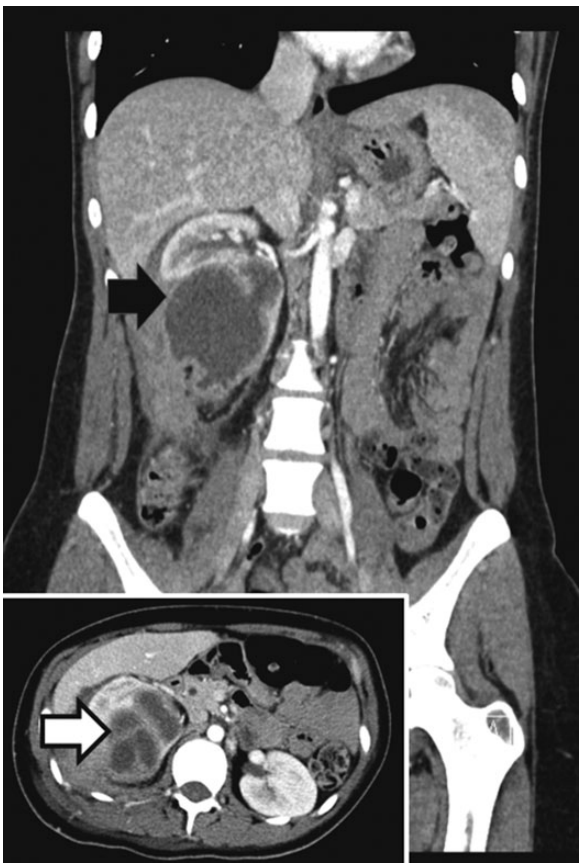

Fig. 2. Coronal Section showing the right kidney with septations illustrated by black arrow and transverse section inset showing a 'bear-paw' appearance illustrated by the white arrow.

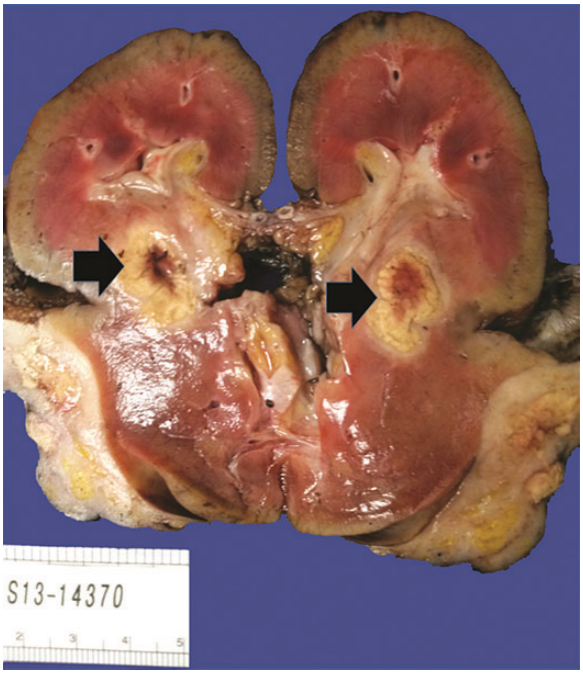

Fig. 3. Small tan-yellow, well circumscribed, pseudo-encapsulated, nodular mass located near the renal hilum extending from the medulla to part of the renal cortex illustrated by black arrows.

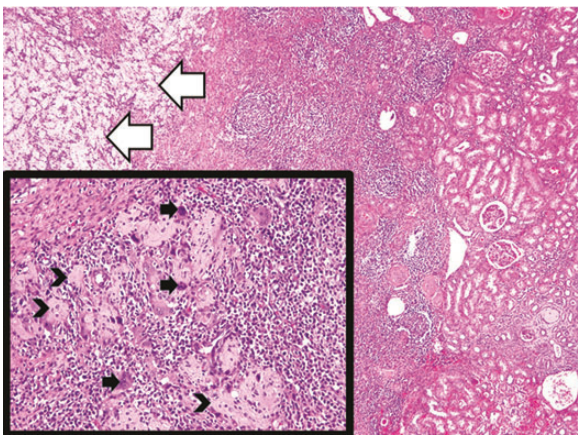

Fig. 4. White arrows and black arrowheads illustrate clusters of large lipid-laden macrophages with foamy cytoplasm (xanthoma cell) and black arrows illustrate multinucleated giant cells.

located near the renal hilum extending from the medulla to part of the renal cortex (Figure 3, black arrows). This gross appearance is seen in xanthogranulomatous pyelonephritis, renal oncocytoma or chromophobe-type renal cell carcinoma. Histology of the mass revealed two hallmarks: (i) clusters of large lipid-laden macrophages with foamy cytoplasm (xanthoma cell seen in Figure 4, white arrow and black arrowheads) and (ii) multinucleated giant cells (Figure 4 inset black arrows).

Xanthogranulomatous pyelonephritis has a female preponderance compared with men with a ratio of 7:3. It occurs as a result of chronic destructive granulomatous inflammation of the renal parenchyma as a consequence of chronic infection [1]. The common organisms isolated include *E. Coli*, *Proteus* species, *Staphylococcus aureus*, group B Streptococcus, Candida, Klebsiella, and Bacteroides [1]. Nephrolithiasis is the primary etiology in up to 83% of cases of described, with occasional staghorn calculus formation. On CT scan, the classic 'bear paw sign' can be seen with the triad of unilateral renal enlargement, non-functional kidney, and pelvic or ureteric obstructing calculus [2]. A differential diagnosis would include transitional-cell carcinoma of the renal pelvis, lymphoma, tuberculosis (unlikely without pulmonary involvement), primary retroperitoneal tumors and primary or secondary retroperitoneal fibrosis [3]. The pathogenesis is thought to occur as a consequence of chronic obstructive nephropathy with bacterial infection leading to systemic chemokine release and stimulation of monocytes and macrophages with subsequent adipose tissue proliferation. Subsequent defective macrophage bacterial removal produces the foam cells [4]. The gross pathological features mimic benign to malignant diseases such as renal oncocytoma or renal cell carcinoma, respectively [5]. Both of these tumors are usually confined to the kidney and can be found in a rare autosomal dominant syndrome called Birt–Hogg–Dubé syndrome, although our patient lacked the cutaneous and pulmonary features of this syndrome.

Treatment is nephrectomy with an excellent postoperative prognosis provided good residual contralateral renal function exists.

In conclusion, the present case re-emphasizes the importance of appropriate follow-up of patients with recurrent urinary tract infections and nephrolithiasis. Chronic infections can lead to granulomatous changes within the kidney and rarely complicate into xanthogranulomatous pyelonephritis with subsequent renal loss.

Conflict of interest statement. None declared.

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