Xanthogranulomatous pyelonephritis

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Learning objectives

To describe and illustrate imaging features of xanthogranulomatous pyelonephritis.

Background

Xanthogranulomatous pyelonephritis is an uncommon form of chronic pyelonephritis, characterized by destruction of renal parenchyma and replacement by lipid-laden foamy macrophages. It is believed to result from an atypical, incomplete immune response to subacute bacterial infection. The most common organisms implicated are *P. mirabilis and E. coli*, but a variety of other bacteria may be found. Some cases occur in association with a renal pelvic calculus and obstruction is believed to be a contributing factor.

Female patients are more frequently affected than male patients in a ratio of 2.1. Patients are usually in mid-life and have a long history of recurrent urinary tract infection. Other risk factors include diabetes and nephrolithiasis. Generalized symptoms include low-grade fever, malaise, chills, anorexia and weight-loss. Nonspecific urinary symptoms include flank pain and haematuria.

Although the diagnosis can often be suspected based on imaging studies, the definitive diagnosis requires histological examination. At microscopy, the caliceal mucosa is ulcerated and replaced by necrotic debris with numerous inflammatory cells, particularly polymorphonuclear leucocytes. The medullary area is replaced by lipid-laden macrophages, admixed with lymphocytes and plasma cells. In the superficial cortical areas, there is fibrous replacement of the kidney with few nephrons remaining. Extension of the xanthogranulomatous inflammation into the perinephric space is common.

Treatment usually consists of extended open nephrectomy.

Imaging findings OR Procedure details

Before the widespread of cross-sectional imaging, a pre-operative diagnosis of xanthogranulomatous pyelonephritis was possible in only a small percentage of patients. The most characteristic radiographic finding is a large staghorn calculus. Additional findings include enlargement of the renal outline and, with more advanced disease, obscuration of the ipsilateral psoas margin. Excretory urography demonstrates a pronounced decrease of the renal function of the affected side.

US typically demonstrates an enlarged kidney with loss of normal renal architecture and a central echogenicity that corresponds to a staghorn calculus (Fig. 1). It is generally associated with acoustic shadowing. Dilated calices filled with pus may appear as hypoechoic inhomogeneous areas in the renal sinus (Fig.2). In focal xanthogranulomatous pyelonephritis a focal inflammatory mass is seen. It is virtually impossible to differentiate from a renal abscess or a necrotic renal cell carcinoma.

CT is the mainstay of diagnostic imaging for xanthogranulomatous pyelonephritis as it allows a confident diagnosis and gives information about the extra-renal extent of the disease.

CT demonstrates an enlarged kidney with multiple hypodense edge shaped areas representing dilated calices and pus filled cavities (Fig.3). Negative attenuation due to lipid rich xanthogranulomatous tissue may be also observed. The walls of these cavities usually show strong enhancement after contrast administration, denoting the marked vascularity of granulation tissue and compressed normal parenchyma (Fig.3). A central obstructive calculus within a contracted or hydronephrotic renal pelvis is sometimes seen (Fig.4). The kidney is generally non-functioning with delayed or absent contrast excretion. Extrarenal extension is characterized by stranding and inflammatory changes in the perinephric fat and associated thickening of the renal fascia (Fig.5). Spread beyond the perinephric space may occur into the posterior and anterior pararenal spaces, psoas muscle, diaphragm, chest and abdominal wall (Fig. 6). Fistula formation is a possible complication.

Atypical findings include massive pelvic dilatation, absence of stones (10%), renal atrophy (13%), and presence of gas in renal parenchyma. Although gas is rarely seen, it may be confused with pyonephrosis or emphysematous pyelonephritis.

Focal xanthogranulomatous pyelonephritis is seen in approximately 10% of patients. Some of these cases are related to a duplicated collecting system and demonstrate findings similar to those seen in the diffuse form. Others closely mimic the features of a bacterial abscess or a neoplasm (Fig. 7). In this case, the diagnostic is almost always made after nephrectomy.

Diffuse xanthogranulomatous pyelonephritis can be staged based on CT findings according to the degree of extrarenal involvement. Stage 1 includes the cases with involvement limited to the kidney (Fig. 8). In stage 2 inflammatory changes extend to the perirenal fat within the renal fascia (Fig. 9). In stage 3 involvement extends beyond the renal fascia into the retroperitoneum, thorax, peritoneal cavity or abdominal wall tissues (Fig 10).

MRI is also a valuable tool for diagnosing xanthogranulomatous pyelonephritis, as it is extremely sensitive for identifying the accumulation of the lipid-laden foamy macrophages as high-intensity signal on T1-weighted images. Chemical shift and short-tau inversion recovery imaging usually increase the sensitivity of fat detection. However,

angiomyolipoma, renal cell carcinoma and retroperitoneal sarcoma occasionally contain fat tissue, and therefore those entities may be extremely difficult to differentiate from xanthogranulomatous pyelonephritis.

Images for this section:

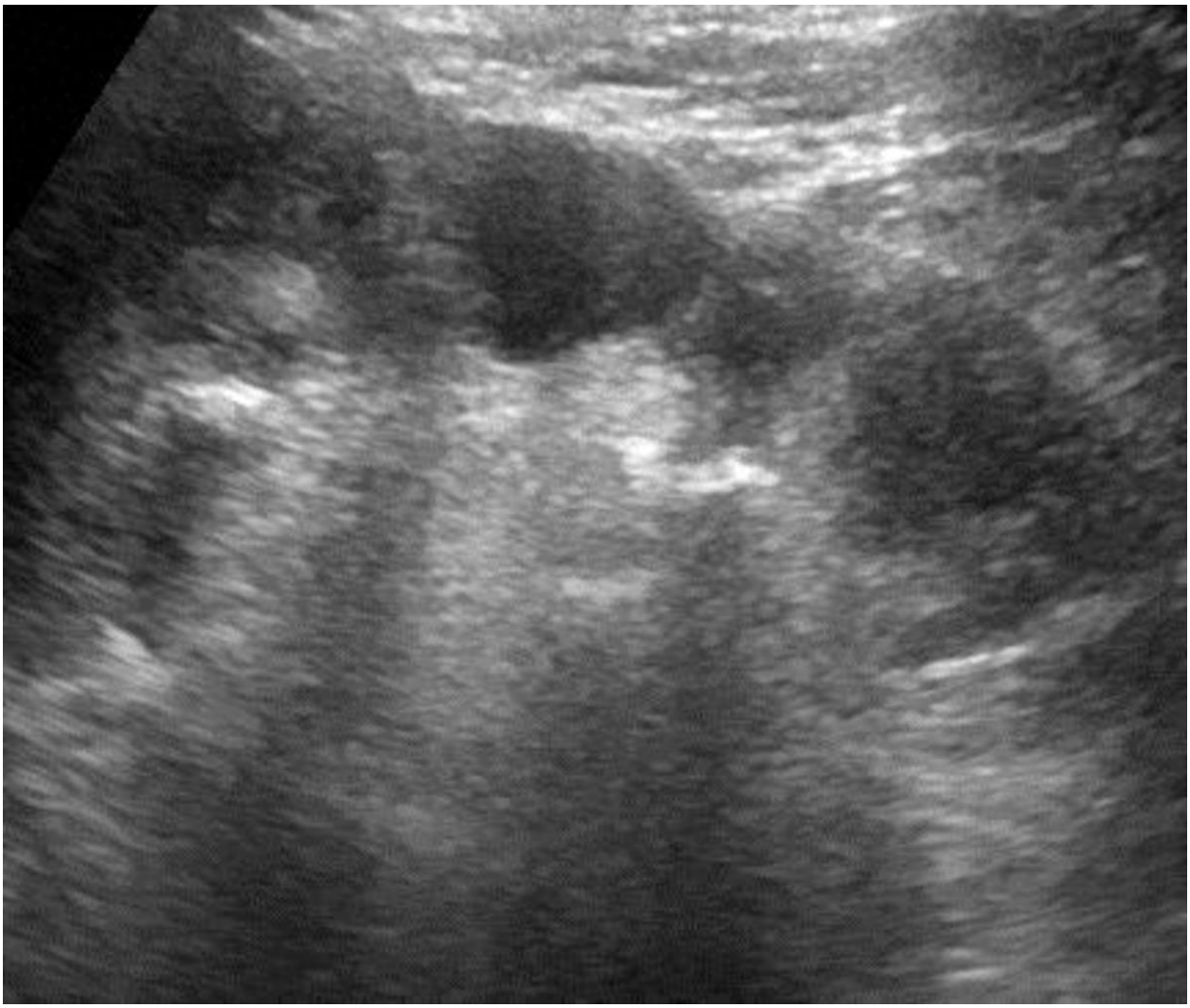

Fig. 1: Renal utrassound of a female patient with xanthogranulomatous pyelonephritis. The image demonstrates a staghorn calculus occuping most of the renal sinus and producing posterior acustic shadowing.

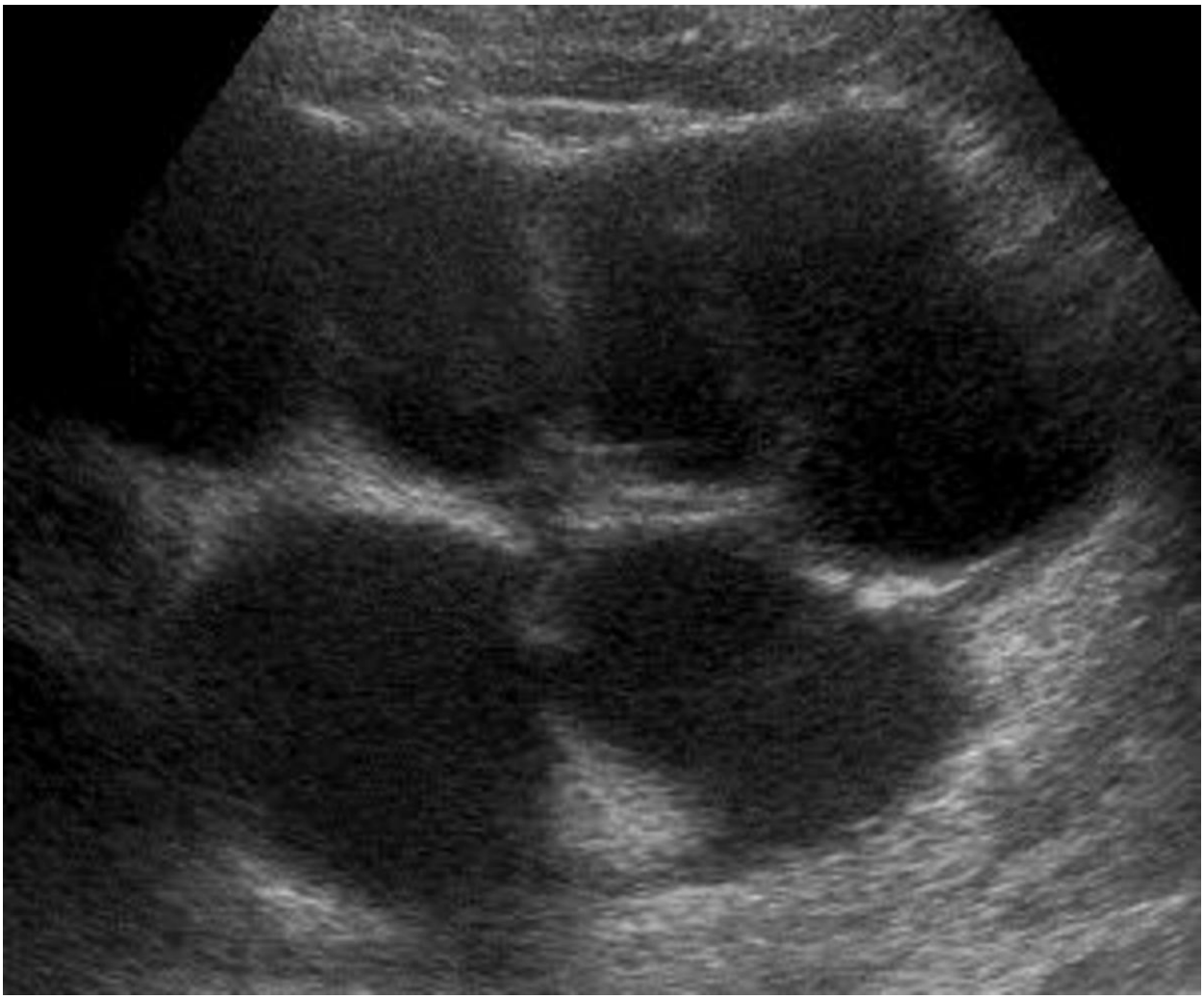

Fig. 2: Renal US of a 65 year-old woman with xanthogranulomatous pyelonephritis. The image demonstrates an enlarged left kidney (18cm in major axis) with multiple hypoechoic round areas corresponding to the pus filled dilated calicies.

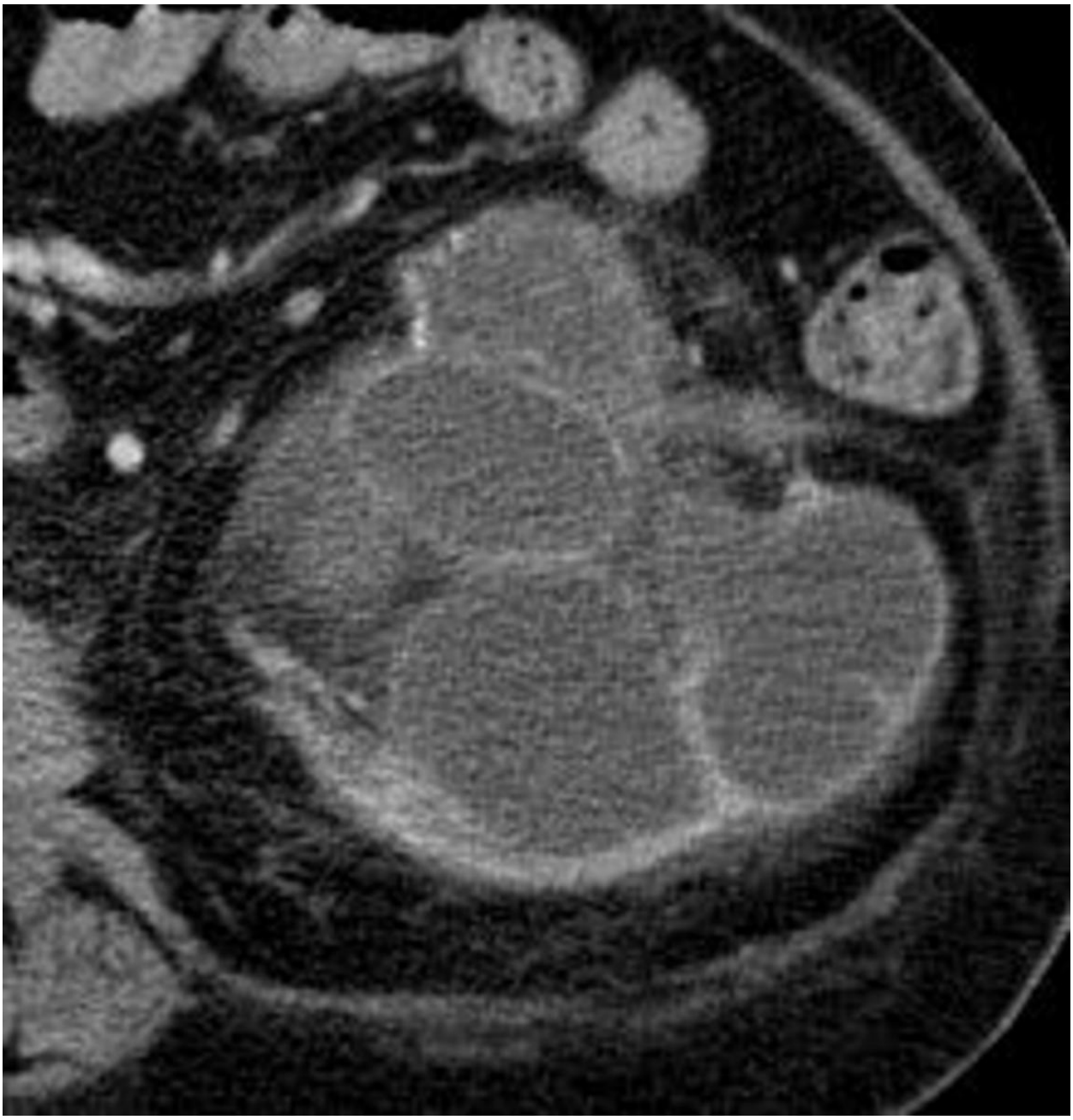

Fig. 3: Contrast enhanced abdominal CT of a female patient with xanthogranulomatous pyelonephritis. The kidney is enlarged and have multiple hypodense edge shaped areas with hyperenhancing walls.

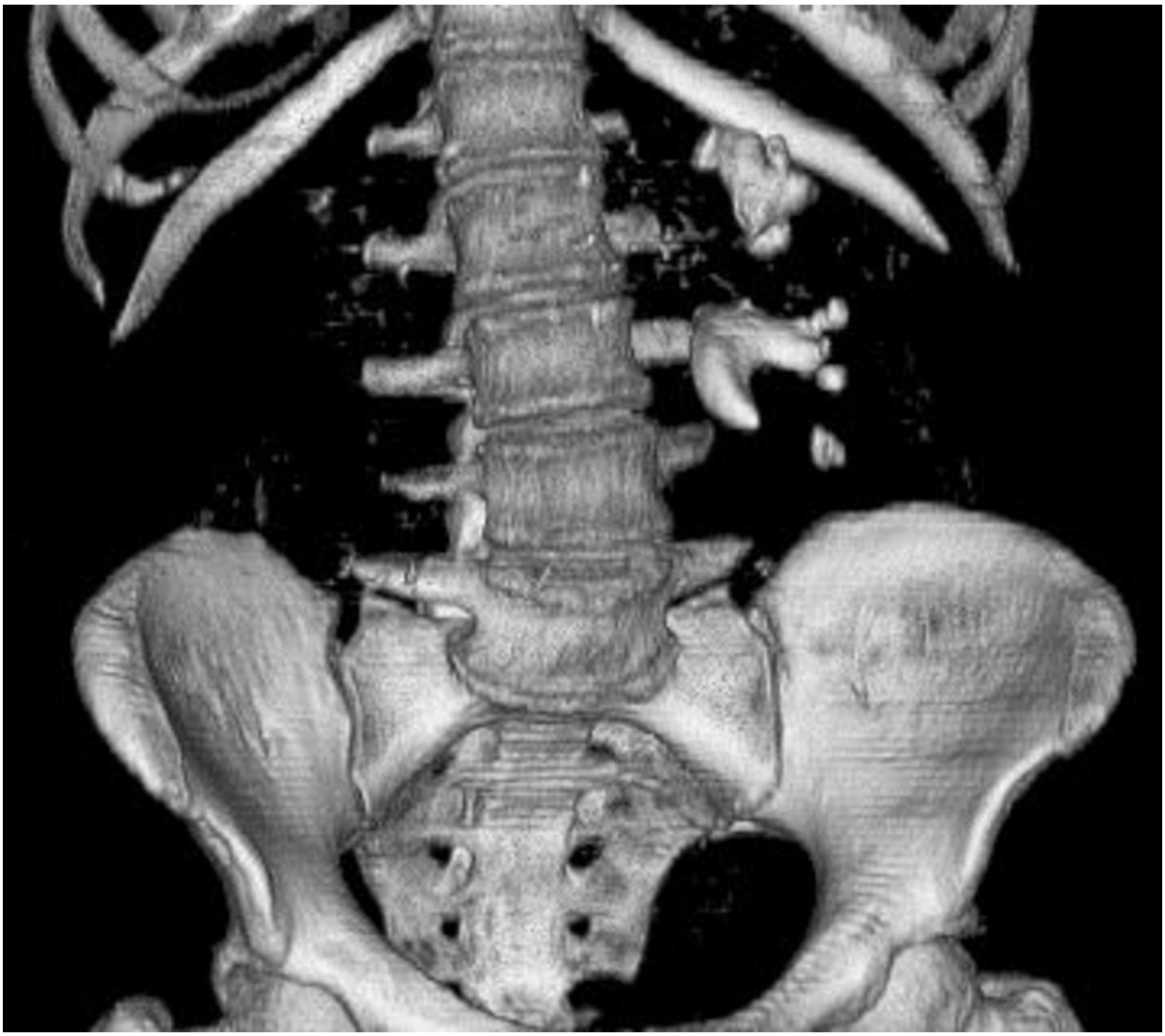

Fig. 4: Volume rendering reconstruction of an abdominal CT demonstrates the three-dimensional appearance of a staghorn calculus.

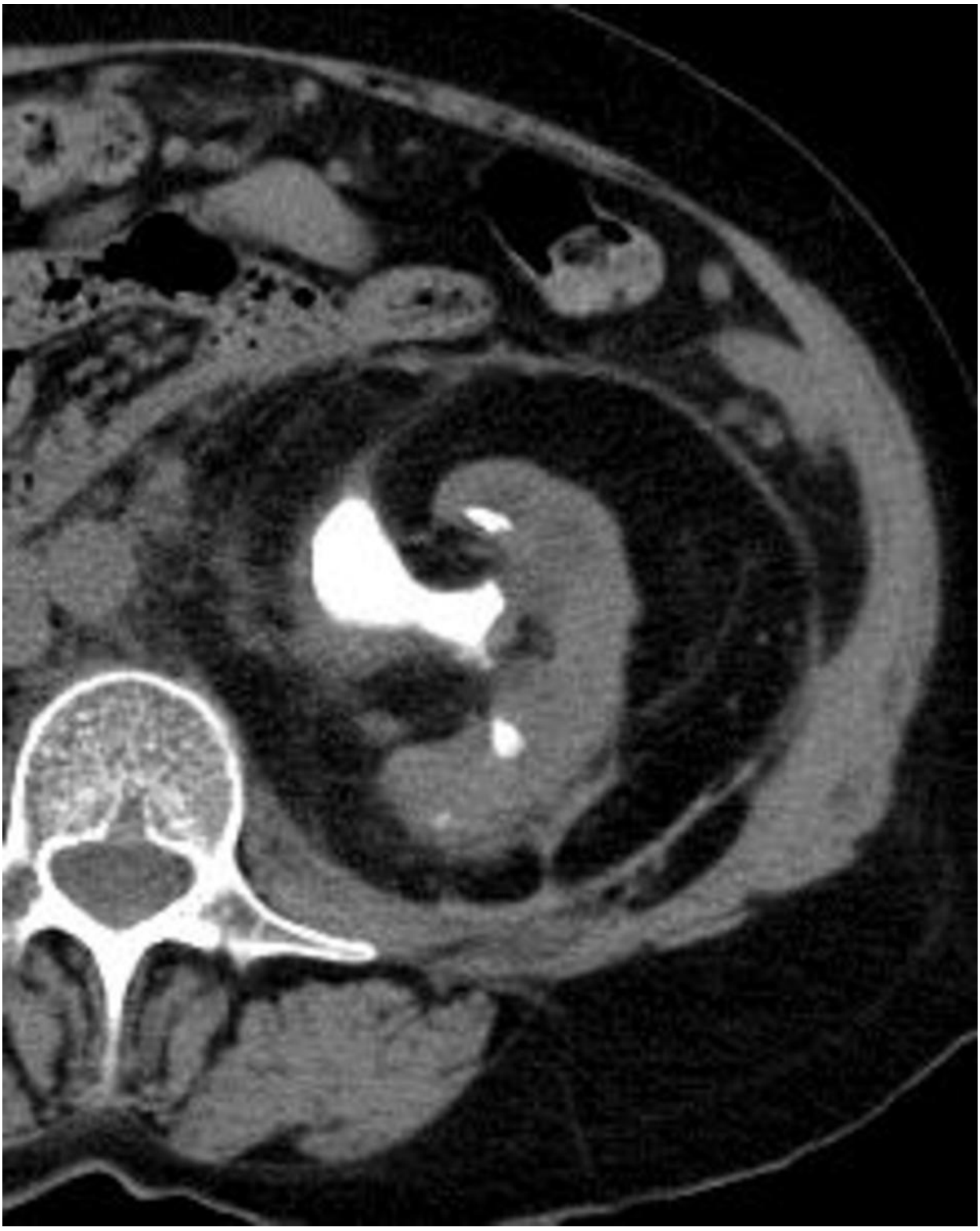

Fig. 5: Contrast enhanced abdominal CT of a female patient with xanthogranulomatous pyelonephritis. The kidney contains multiple hypodense cavities and a central obstructing calculus. The renal facia is thickened and hyperenhancing. Pronounced stranding and inflammation of the perirenal fat.

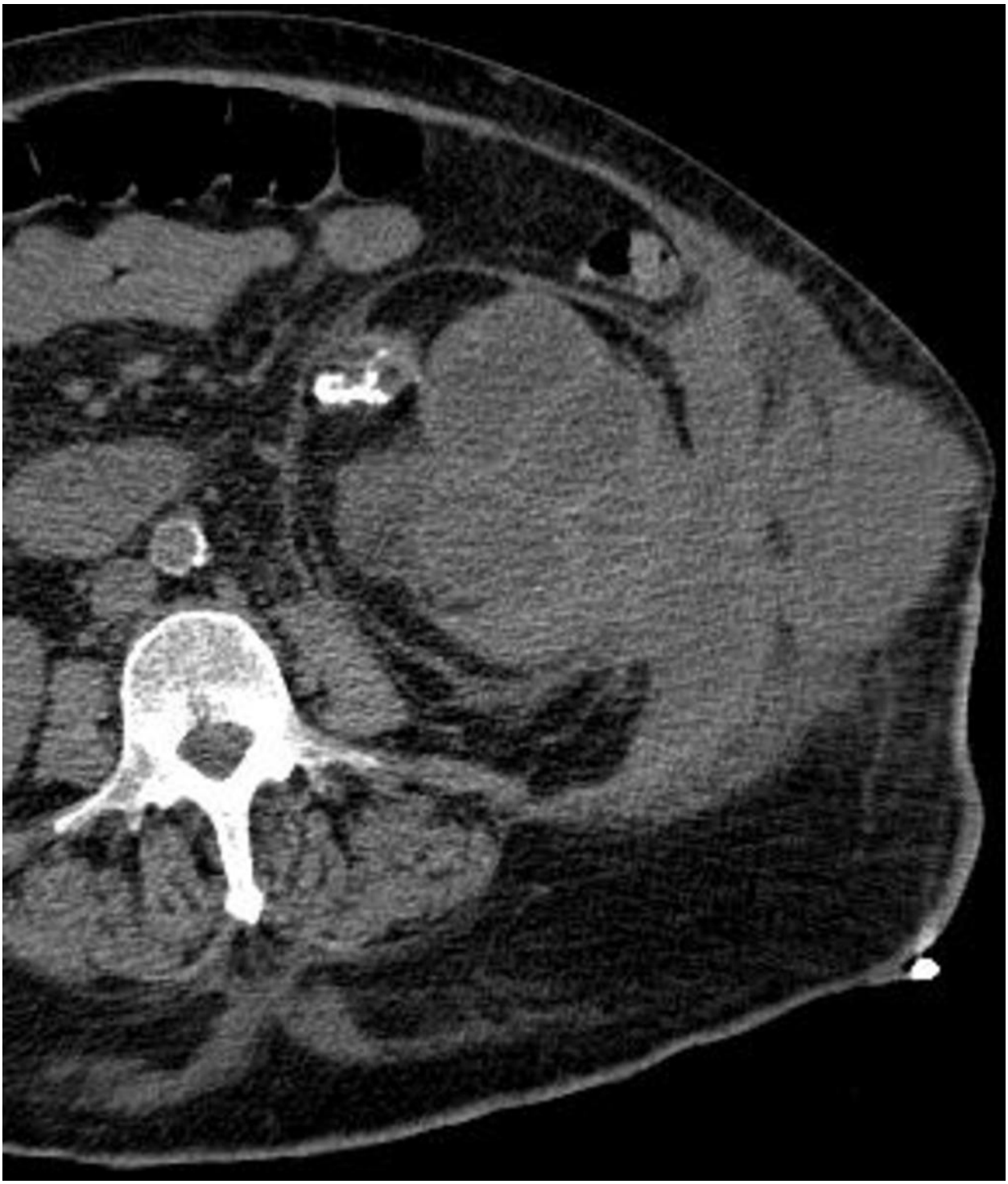

Fig. 6: Abdominal CT of a 59 year-old woman with xanthogranulomatous pyelonephritis and extra-renal extension. The affected kidney is enlarged with multiple hypodense areas. The inflammation spread beyond the perirenal space into the abdominal wall, forming an abscess.

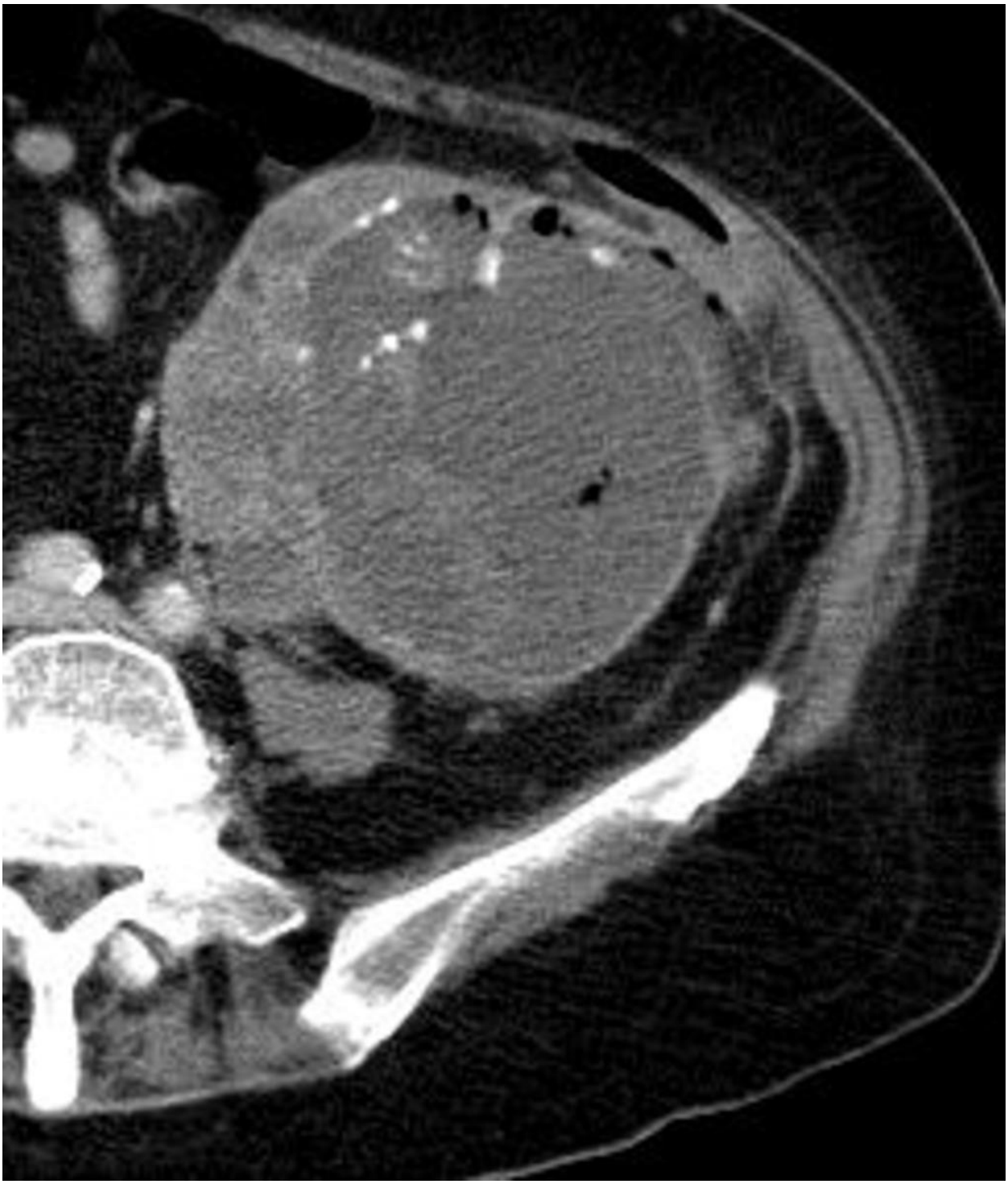

Fig. 7: Focal xanthogranulomatous pyelonephritis in a 61 year-old woman. Contrast enhanced abdominal CT demonstrates a large heterogeneous masse in the lower pole of the left kidney. It contains some focus of calcification and air. Imaging methods cannot distinguish this lesion from a renal abscess or a neoplasm. Histological examination proved a focal xanthogranulomatous pyelonephritis.

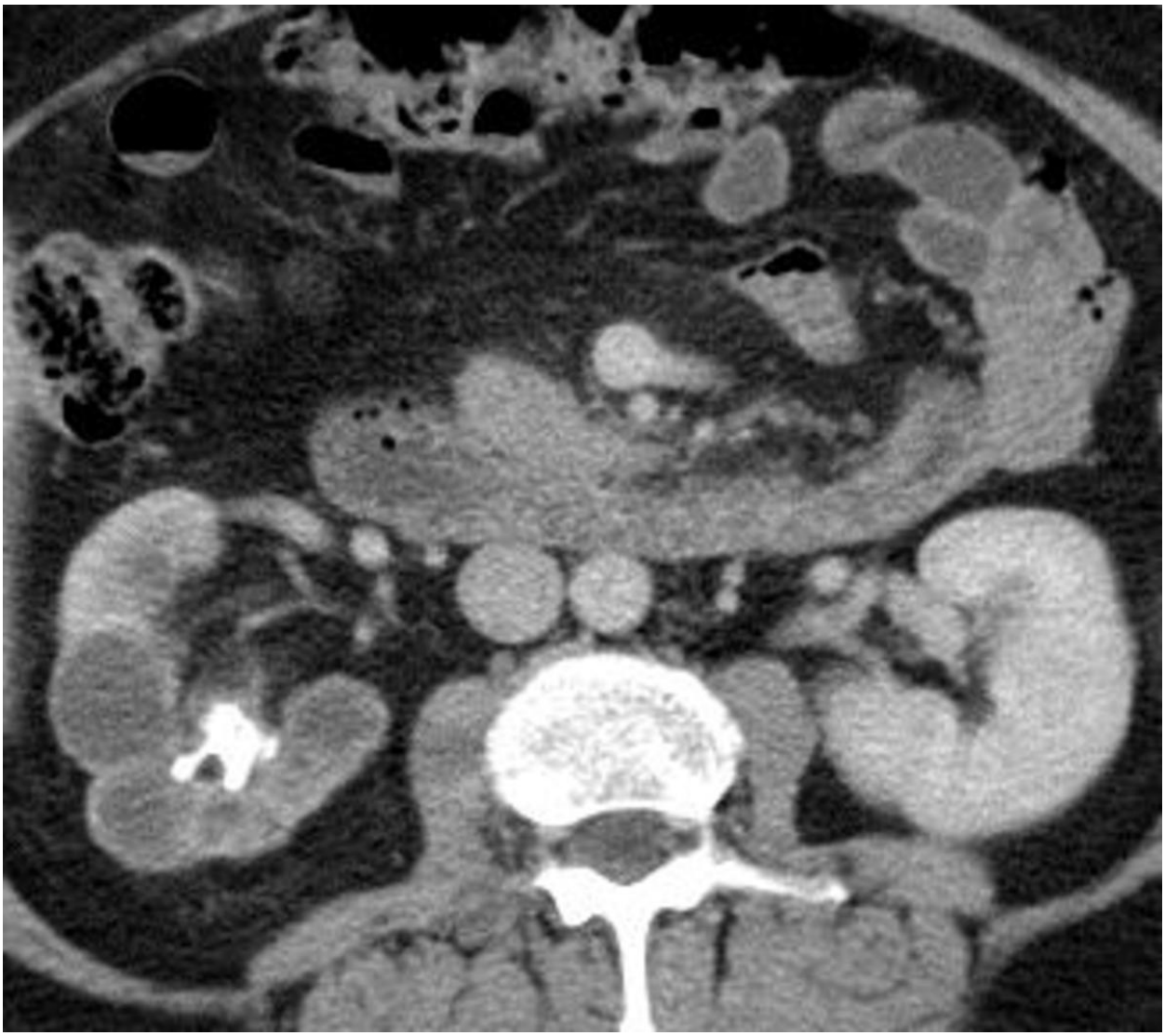

Fig. 8: Staging of xanthogranulomatous pyelonephritis according to the degree of extrarenal involvement. In stage 1 inflammation is limited to the kidney.

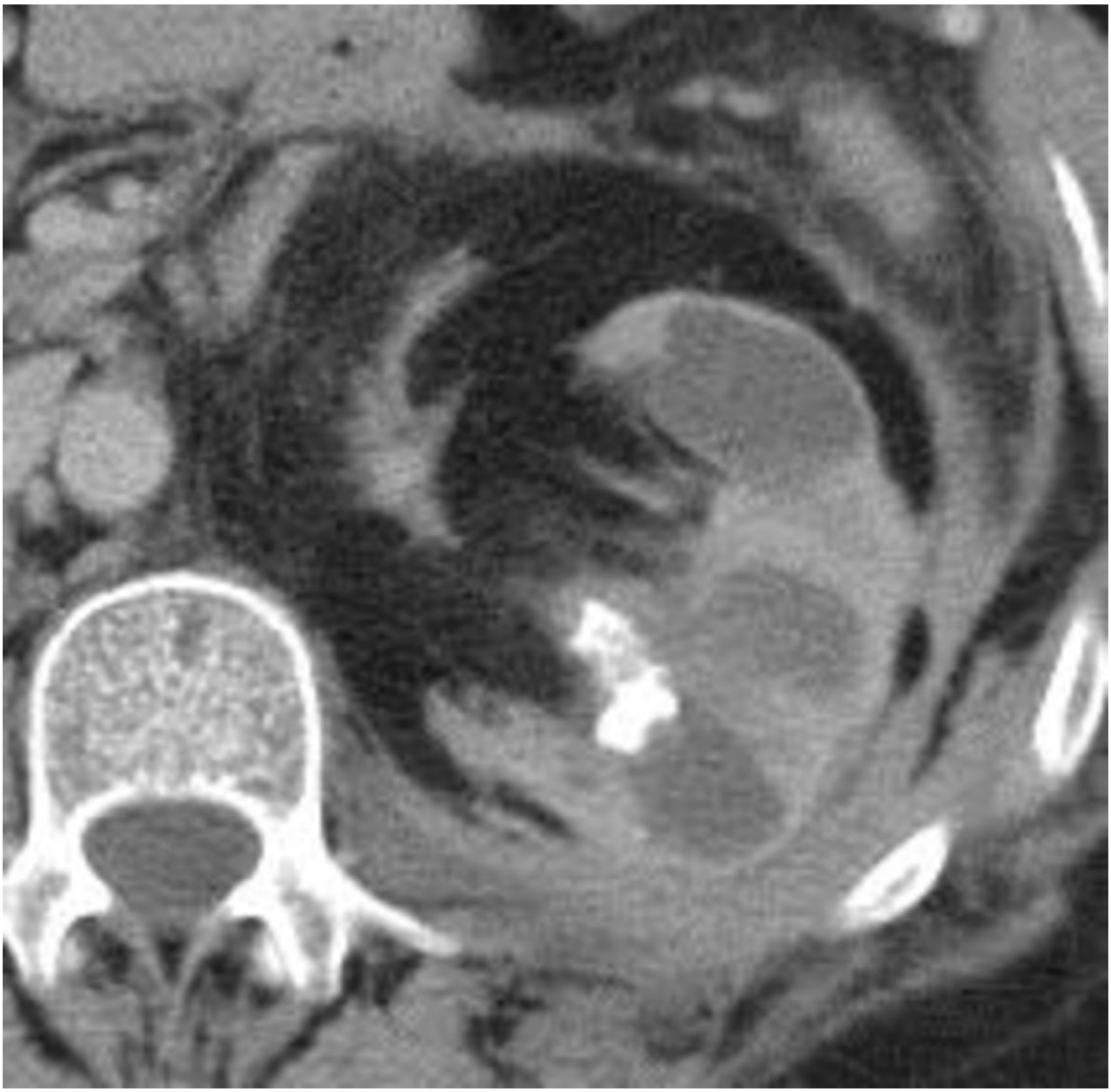

Fig. 9: Staging of xanthogranulomatous pyelonephritis according to the degree of extrarenal involvement. In stage 2 inflammatory changes extend into the perinephric space.

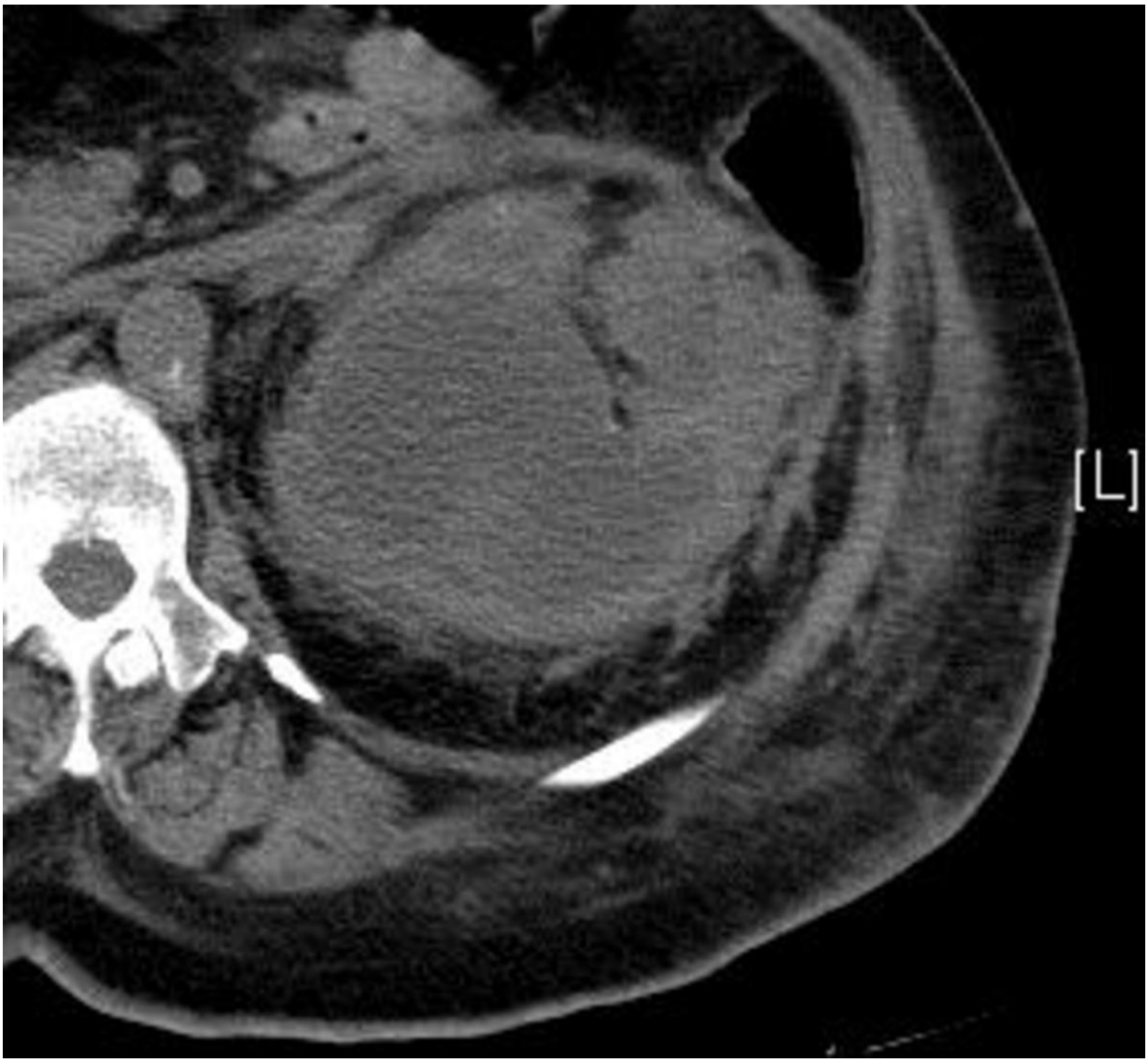

Fig. 10: Staging of xanthogranulomatous pyelonephritis according to the degree of extrarenal involvement. In stage 3 inflammation extends beyond the renal fascia involving adjacent tissues as the lateral abdominal wall shown in the image.

Conclusion

Xanthogranulomatous pyelonephritis is a chronic granulomatous process induced by recurrent bacterial urinary tract infection. Although US is useful in the diagnosis of this condition, CT is the main imaging tool, providing specific findings of nephromegaly, renal function impairment, urinary obstruction due to calculi and multiple hypodense edge shaped areas. Extra-renal extent is common. In a few atypical cases the preoperative diagnosis cannot be made by CT. The definitive diagnosis requires histological examination.

References

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