Xanthogranulomatous Pyelonephritis : Diagnosis using Computed Tomography

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Introduction

Xanthogranulomatous pyelonephritis (XGP) is an uncommon serious, debilitating chronic inflammatory disorder of the kidney characterized by an infectious phlegmon arising in the renal parenchyma. It is seen in patients who are usually immunocompromised and have associated urinary tract infection and/or urolithiasis. The clinical presentation is nonspecific so that differential diagnosis with other kidney diseases becomes difficult. Computed Tomography (CT) has shown to be a reliable and accurate imaging modality in diagnosing XGP and characterizing its extrarenal involvement. The treatment is surgical and consists of nephrectomy, while a conservative nephron saving approach is limited to focal form of cases. We report a case of XGP in a patient of chronic renal failure that was diagnosed using CT.

Case Report

A 38 year old male patient suffering from chronic renal failure on haemodialysis presented with high grade fever associated with chills and acute severe pain in the left flank of two days duration. Past history revealed that this patient had sustained a fall four years back leading to spinal injury and paraplegia with urinary retention, for which he was on long term urinary catheterization. He developed repeated urinary infection leading to chronic pyelonephritis and subsequently developed chronic renal failure for which he was on haemodialysis.

Clinical examination revealed a well built male who was febrile, had extreme tenderness over the left renal angle with generalized guarding. Ultrasonography of the abdomen revealed a contracted right kidney with increased cortical echotexture, the left kidney could not be visualised, instead a large predominantly hyperechoic mass lesion measuring 16x8 cm in the left renal fossa was seen. No renal calculus was seen on either side.

A non contrast CT scan was done which revealed a globally enlarged, predominantly hyperdense left kidney measuring about 15x7.5 cm, with well defined hypodense areas within it. The right kidney was contracted and measured 6.4x3.6 cm.

No calculus was seen in either of the kidneys.

Contrast enhanced CT scans revealed areas of irregular and nonhomogenous enhancement within the left renal parenchyma, especially around the low density areas (Fig 1). There was extensive extra-renal extension of the disease process with involvement of the peri-nephric space, anterior and posterior para-renal spaces, ipsilateral conal fascia, psoas and the posterior abdominal wall muscles (Fig 2). The right kidney showed no evidence of any enhancement of the parenchyma nor any opacification of the pelvicalyceal system. Based on the clinical features and CT findings a diagnosis of XGP of the left kidney was made. The patient was put on parenteral antibiotics and supportive therapy and subsequently taken up for nephrectomy.

Per-operatively the gross specimen revealed a large mass of yellow tissue with multiple areas of necrosis and haemorrhage, superficially resembling a renal cell carcinoma. Histopathology showed lipid-laden “foamy” macrophages accompanied by both chronic and acute-phase inflammatory cells. A final diagnosis of XGP was made. The patient had an uneventful post-operative recovery and is on haemodialysis awaiting renal transplantation.

Discussion

XGP is a serious, chronic inflammatory disorder of the kidney associated with indolent bacterial infection. The disease process is characterized by an infectious phlegmon which begins in the renal pelvis and extends into the medulla and cortex which are gradually destroyed and replaced by lipid laden macrophages (Xanthomam Cells); the infective process then can extend into the extra-renal tissues [1]. XGP is seen in approximately 1% of all renal infections, is four times more common in women than men and is usually noted in the fifth and sixth decades of life, although most cases of XGP are unilateral, bilateral disease has been reported and is generally fatal [2]. The focal form of this disease accounts for 12.5% of all cases of XGP and has similar clinical features, laboratory and imaging findings [3].

The exact etiology of XGP is unknown, but it is generally accepted that the disease process requires

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Fig. 1 : Contrast enhanced CT scan showing globally enlarged left kidney with multiple hypodense areas and irregular enhancement

long-term renal obstruction and infection. Calculi, frequently of staghorn proportions, may be seen in 75 to 86% of these patients, but are not required to make the diagnosis [4]; our patient did not have any renal calculus. There is a common association with *Proteus* and *E Coli* infection and in 30-40% of cases these organisms are cultured from the urine [5]. XGP is often seen in diabetics or in immunocompromised individuals, our patient had chronic renal failure due to chronic pyelonephritis as a result of repeated infection. Patients present with systemic symptoms of malaise, fever, chills and weight loss along with urinary complaints of flank pain, increased frequency of micturition, dysuria and nocturia.

Contrast enhanced CT is a reliable method to diagnose XGP and to establish the presence and extent of extrarenal involvement. The CT findings of XGP are calculi (75-86%), increase in renal volume (55-60%), hydronephrosis (80%), hypodense areas with density measurements from 15 to 25 HU representing focal areas of parenchymal destruction filled with pus and / or debris, rim enhancement of these low density areas and extrarenal involvement, which is seen in 15-20% of cases [4]. In the focal form of the disease, CT reveals a poorly enhancing mass adjacent to a calyx or in one pole of a kidney with a duplicated collecting system [3]. This disease process shares many characteristics with a true neoplasm in terms of its imaging appearance and ability to involve adjacent structures or organs. XGP is notorious for fistulization pyelo-or uretero-cutaneous fistulae are well described, and occasionally may involve the bowel resulting in pyeloenteric fistula [6]. Medical

Fig. 2 : Contrast enhanced CT scan showing extensive extra-renal extension of the disease involving the peri-nephric, para-renal spaces and the posterior abdominal wall muscles

therapy alone is inadequate to treat XGP; antibiotics are a temporary measure for patients requiring medical work-up prior to nephrectomy. Total nephrectomy is the gold standard of treatment for XGP, unless both sides are affected in which case partial nephrectomy is done. These are often challenging cases, particularly if there is local organ involvement, the aim is to remove all involved granulomatous tissue, and if this is not accomplished, the remaining infected tissue may lead to cutaneous fistulae [7].

Traditionally, imaging of renal infections was largely a multimodality approach. Excretory urography, ultrasonography, nuclear scintigraphy and CT scans have all played major roles; however in recent years, CT has increased in prominence in the imaging and evaluation of renal infection. The reason for this trend includes improvement in the quality of CT scanners which provide excellent resolution of the tissues and their easy accessibility. Spiral scanning technology has greatly increased the quality and usefulness of the information CT provides. Most uncomplicated cases of renal infection in adults do not require imaging for diagnosis and treatment. When imaging is indicated plain and contrast enhanced CT is always the study of choice [8].

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A man needing a heart transplant is told by his doctor that the only heart available is that of a sheep. The man finally agrees and the doctor transplants the sheep heart into the man. A few days after the operation, the man comes in for a checkup. The doctor asks him “How are you feeling?” The man replies “Not BAAAAD!”