Case Report

Atypical presentation of focal xanthogranulomatous pyelonephritis

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ABSTRACT

Xanthogranulomatous pyelonephritis (XGP) is a chronic condition caused by granulomatous reaction to chronic renal infection. The diffuse form is more common where the kidney is enlarged while still retaining the reniform shape. The focal form is very rare and is pathologically similar to diffuse form but limited to one pole or less. To the best of our knowledge, all reported cases of XGP in the literature report a history of chronic obstructive uropathy such as calculus, stricture, or mass. We are presenting here a case report of focal XGP in a 58-year-old woman with no known history of obstructive uropathy and presented with nonspecific symptoms.

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Introduction

Xanthogranulomatous pyelonephritis (XGP) is a chronic condition caused by granulomatous reaction to chronic renal infection. Microscopically, areas of abscess formation and chronic inflammation can be seen with diffuse infiltration by lipid laden macrophages that give it a foamy yellow appearance microscopically. Most patients that develop XGP have a history of chronic obstructive uropathy caused by calculus, stricture, or mass [1]. Patients may present with acute symptoms such as fever, chills, and flank pain, however, many present with nonspecific chronic symptoms such as weight loss, fatigue, and anorexia. The condition affects females more than males most commonly between the ages of 45-60 [2,3].

There are 2 recognized forms of XGP, focal and diffuse. The diffuse form is more common where the kidney becomes enlarged, still retaining its reniform shape. Diffuse form is then further staged based on the extent of renal and perirenal involvement. The focal form is very rare, pathologically it is like the diffuse form but limited to one pole or less [4]. The treatment for diffuse form or advanced XGP is nephrectomy with pre- and postsurgical antibiotics. The focal form, however, is treated with antibiotics and drainage initially. If it fails to resolve, nephrectomy may be considered [5,6].
Here we present an atypical presentation of focal XGP in a 58-year-old woman with no known history of obstructive uropathy presenting with nonspecific symptoms.

Case report

A 58-year-old female presented to the emergency department with chief complaints of intermittent epigastric and right upper quadrant pain with associated nausea and vomiting. Past medical history is notable for hypertension. She denied the history of renal calculus but admits to having “kidney infection” during her teenage years. She was alert and oriented with mild distress on presentation. Her vital signs were blood pressure 140/80 mmHg, heart rate of 82 beats per minute, respiratory rate of 12 breaths per minute and oxygen saturation of 98% on room air. The abdominal exam revealed tenderness in the right upper quadrant and epigastric area. Her urinalysis and urine culture were found to be negative. Laboratory evaluation revealed leukocytosis with WBC of 16,000/mm3. Given her symptoms, ultrasound (US) of the right upper quadrant was ordered that demonstrated cholelithiasis and a right renal mass (Fig. 1). Subsequently, contrast-enhanced CT of the abdomen and pelvis demonstrated a 9.8 cm heterogeneous, mildly enhancing renal mass arising from the interpolar region of the right kidney (Fig. 2). MRI was also obtained. On T2-weighted images, the mass demonstrated heterogeneous T2 signal with a thick T2 hypointense rim (Fig. 3). On T1-weighted images, increased T1 signal was seen suggestive of blood products (Fig. 4A). On contrast-enhanced
subtracted images, there was mild diffuse wall thickening with irregular mural-like nodular enhancement (Fig. 4C). Given the overall appearance, differential diagnosis included neoplasm such as papillary renal cell carcinoma (RCC) and renal abscess.

This mass was biopsied with an initial pathology report of necrotic cellular material raising the possibility of necrotic neoplasm. The sample, however, did not show significant pan-kertib staining characteristic for RCC. XGP was considered in the differential, however cytological findings were considered discordant with this diagnosis. Culture of the lesion yielded no growth. Patient was started on ceftriaxone and upon further consultation with urology the decision was made to perform short term follow up imaging.

Patient was initially lost to follow-up; however, repeat CT imaging in 2 years showed interval decrease in the size of the mass to 3.0 × 4.0 cm. Faint peripheral calcifications were present without significant enhancement (Fig. 5). Given the decrease in the size and resolution of clinical symptoms, continued follow up imaging was decided upon. Follow up MRI imaging after another year demonstrated minimal decrease in the size of mass. The mass showed T2 hypointensity (Fig. 6A) and loss of signal on in-phase imaging (Fig. 6B) suggesting hemosiderin content. The mass was heterogeneously hyperintense on T1-weighted images (Fig. 7A) with...
linear and nodular peripheral enhancement on subtraction images (Fig. 7C). Repeat biopsy was subsequently performed that revealed sheets of foamy, lipid-laden histiocytes, chronic inflammation, and areas of fibrosis. An immunohistochemical stain for CD68 highlights abundant histiocytes (Fig. 8). Findings were consistent with a diagnosis of focal XGP.

Discussion

Focal XGP is a rare form of XGP. Most cases of XGP are preceded by obstructive uropathy, most commonly renal calculi. This is an unusual presentation of a focal form of XGP given lack of known renal obstruction. We present here the progression of disease over 5 years with multi-modality imaging evaluation prior to and following treatment and described how its imaging and microscopic features can overlap with renal neoplasm. The patient presented with nonspecific symptoms and was incidentally found to have right renal mass. The first biopsy was inconclusive with features raising the possibility of necrotic neoplasm. Over time the mass decreased in size with areas of internal hemorrhage. A second biopsy 5 years later confirmed the diagnosis of focal XGP. This case demonstrates that not all cases of focal XGP have a history of known obstruction. Imaging and microscopic features of XGP can sometimes have similar findings as renal neoplasm, therefore close follow up is recommended.

Patient consent

No patient identifier information is used in the paper. Written informed consent for the publication of this case report was obtained from the patient.

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