Case report: Xanthogranulomatous pyelonephritis presenting as psoas abscess in a 7-year-old girl

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ABSTRACT

Xanthogranulomatous pyelonephritis (XGP) is a rare variant of chronic pyelonephritis. We report a case of a 7 year girl with fever and recurrent left flank pain with a nonfunctioning left kidney. Left psoas abscess was aspirated under ultrasound guidance and appropriate antibiotics were administered. Left nephrectomy was done for the nonfunctioning kidney. Histopathology was suggestive of XGP. The patient is asymptomatic on follow up.

Key Words: Xanthogranulomatous pyelonephritis; psoas abscess; nephrectomy.

Introduction

Xanthogranulomatous pyelonephritis (XGP) is a chronic inflammatory disease of kidney characterised by suppuration, parenchymal destruction and lipid-laden foamy macrophages [1]. It was first reported by Schlagenhauffer in 1916 [2]. XGP occurs at any age, but is more common among females, particularly middle-aged women [3]. It is very rare in children and, the disease most frequently occurs in those under 8 years. Clinical signs and symptoms are generally nonspecific [4]. XGP can spread into the pelvic cavity leading to serious complications including emphysematous pyelonephritis, perinephric abscess, psoas muscle abscess and, nephro-cutaneous and colonic fistula [5]. Additionally, preoperative diagnosis of this condition is difficult because it may found with various clinical manifestations, mimicking renal tumor, renal tuberculosis, or other rare chronic renal inflammatory process like malakoplakia [1-5]. Here, we report a case of a 7 years old girl with xanthogranulomatous pyelonephritis complicated by psoas abscess.

Case report

A 7 year female child was admitted with fever and recurrent left flank pain of 4-5 days duration. Ultrasonography of abdomen
revealed 20cc hypoechoic collection in the left psoas muscle suggestive of abscess with grossly dilated calyces of left kidney filled with echogenic material. Psoas abscess was aspirated and patient was started on intravenous antibiotics. Her laboratory reports showed leukocytosis, microcytic anemia, and raised erythrocyte sedimentation rate. Urine analysis revealed 8-10 pus cells while urine culture was negative. In further imaging studies, contrast enhanced computed tomography showed enlarged left kidney with multiple cysts with hypodense areas and calcifications with no excretion from left kidney and normal functioning right kidney [Fig. 1].

Differentials of multicystic dysplastic kidney (MCDK) or infective etiology or renal Koch's were considered. Micturating cystography was normal. Renal scan was suggestive of non-visualized, non-functioning left kidney. Patient was posted for left nephrectomy in view of infective etiology and non-functioning kidney. Intra-operatively, left kidney was densely adherent and severely hydronephrotic with thinned out renal parenchyma and multiple cysts containing thick pus [Fig. 2].

Histopathology was suggestive of aggregation of foamy macrophages with inflammation and hemorrhage suggestive of XGP. Intra-operative pus culture showed *Escherichia coli*. Post-operatively patient improved well and on regular follow up.

**Discussion**

Xanthogranulomatous pyelonephritis occurs in approximately 1% of all renal infections. It is a disease predominantly noted in the fifth and sixth decades of adults but can occur in all age groups. It is rarely seen in children and neonates. In children, the disease most frequently occur below 8 years of age with male predominance and is focal, affecting left kidney predominantly [6]. Its etiology remains unclear, although common causes are urinary obstruction due to renal calculus, urinary tract infection, abnormal lipid
metabolism, lymphatic obstruction, altered immune response, and vascular occlusion [7]. On the basis of morphological involvement, two forms of XGP have been described: diffuse (83%-90%) and focal (10%-17%). The focal variety is often misdiagnosed as a renal mass. Malek and Elder proposed a staging system for XGP: Stage I - the lesion is confined to the kidney; Stage II - there is infiltration of the Gerota space; Stage III - extends to the perinephric space and other retroperitoneal structures [8].

The symptoms are often vague and non-specific. Some patients present as abdominal mass. Most of the children present with symptoms of chronic infection, like recurrent fever, weight loss, pallor and failure to thrive. Urinary symptoms such as dysuria, frequency, and hematuria are uncommon [6]. Additionally, urinary tract infections, pyuria, leukocytosis and raised erythrocyte sedimentation rate (ESR) are frequently associated with XGP [9]. Positive urine cultures and pathological urine sediment are predictive for XGP but are found in only 70% of all patients [10]. Positive urine culture has been reported in 50%-75% of cases of XGP. Escherichia coli and Proteus species, the most commonly implicated organisms, are found in 59%-95% of positive cultures [11].

Preoperative diagnosis of XGP is difficult due to its clinical and radiological similarities to renal malignancy. Ballasteros et al. [12] reported an 80% accurate preoperative diagnosis of XGP in adults by serial urinary cytology for foam cells. Ultrasonography is the recommended first step for diagnosis and may differentiate between the two forms of XGP. In the focal form of XGP, ultrasonography shows a localized hypoechoic mass, often misdiagnosed as a renal tumor [13]. In the diffuse form of XGP, ultrasonography shows an enlarged kidney with thinned parenchyma, calculus in renal pelvis, multiple fluid-filled masses, and dilated pelvicaliceal system [3]. Computer tomography is a choice radiological technique in evaluating patients with XGP and, diagnosis is made by the presence of an enlarged, non-functioning kidney with staghorn calculus, caliceal dilatation, low attenuation areas replacing the renal parenchyma secondary to inflammatory infiltrate and perinephric standing [13, 14]. Kidneys XGP with a non-functioning or poorly functioning is the most common finding on intravenous pyelogram (IVP) and MAG3/DTPA renal scan [13], as seen in our patient.

Final diagnosis is usually possible on histopathological examination. XGP is characterized as destruction of parenchyma, and accumulation of pathognomonic lipid laden macrophages which surround abscess cavities or present as discrete yellow nodules. These cells can be difficult to distinguish from clear cell carcinoma [3].

The long-standing mainstay of therapy for diffuse XGP disease is total nephrectomy but partial nephrectomy is advocated whenever possible (focal or bilateral lesions) [3,15-17]. However, there have been reports of successful treatments of focal XGP with medical intervention only [10]. Preoperative and postoperative broad-spectrum antibiotics should also be as key factors for successful management of XGP [16]. As was seen in our patient, psoas abscess is rare with XGP, but when it occurs, the diagnosis and management of the cases are more complicated. In our patient, the diffuse form XGP related to Escherichia coli was treated with abscess drainage, intravenous antibiotics and nephrectomy, and the disease completely resolved.
**Conclusion**

XGP is an uncommon and distinct type of chronic infective pyelonephritis. Clinical awareness and a high index of suspicion are required to achieve the correct preoperative diagnosis and appropriate management.

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**References**


