CEN Case Reports (2018) 7:44–47
https://doi.org/10.1007/s13730-017-0289-7

CASE REPORT

Xanthogranulomatous pyelonephritis: a case with rare adhesion to pancreas

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Received: 15 October 2017 / Accepted: 18 November 2017 / Published online: 25 November 2017

Abstract
Xanthogranulomatous pyelonephritis (XGP) is a rare benign condition with unknown aetiology and chronic infection of kidney. Commonly, most cases are related with urinary tract obstruction, nephrolithiasis, infection, diabetes, and/or immune compromise. XGP is associated with destruction of the renal parenchyma and granulomatous inflammation with foamy lipid-laden macrophages resulting from obstructive uropathy. It closely mimics a malignancy, exhibiting local tissue invasion and destruction. Adjacent organs especially duodenum as well as very rarely pancreas or spleen may be involved. Additionally, XGP is known as notorious for fistulisations, such as pyelocutaneous and ureterocutaneous fistulae, which have been reported as well described. XGP may be indistinguishable from renal cell carcinoma by radiographic and clinic consultation so it must be diagnosed based on the histopathologic examinations. Furthermore, macroscopic appearance of XGP is a mass of yellow tissue with focal haemorrhage besides necrosis and in this regard, it grossly resembles renal cell carcinoma. Here, we report the case of a 32-year-old female, preoperatively diagnosed as malignancy by clinical examination. Our further pathological evaluations revealed very rarely adhesion of XGP to pancreas tissue.

Keywords Xanthogranulomatous pyelonephritis · Pseudotumor · Adhesion · Pyelonephritis

Introduction

Xanthogranulomatous pyelonephritis (XGP) is a long-term destructive granulomatous inflammation of renal parenchyma and occurs in approximately 1% of all renal infections [1]. The most common symptoms were declared as flank or abdominal pain, lower urinary tract symptoms, palpable mass, fever, gross hematuria, and weight loss [2]. Also, the most common associated factors were reported as urinary tract obstruction and infection. XGP is often difficult to differentiate from renal cancer preoperatively [3].

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Case report

Here we present the case of a 32-year-old female, investigated during 1-year period for gradually progressive pain in left lumbar region. Also, complaints of anorexia, nausea, dysuria and pollakuria were present. The patient was treated for 1 year with an initial diagnosis of urinary tract infection. Urinar ultrasonography revealed a heterogeneous hypoechoic lesion, measuring 77 × 52 cm in the median part of the left kidney, causing a defect in the medulla cortex differentiation and having no definite borders. Contrast-enhanced computed tomography examination revealed a solid lesion which had lobular contours and extended from the middle portion to the upper pole of the left kidney and not clearly distinguished from the pancreatic tail (Fig. 1).

The patient was scheduled for surgery with an initial diagnosis of malignancy. During the operation, the pancreatic tail was invasive with mass and the patient underwent distal pancreatectomy at the same time. Macroscopically, the kidney had a 8.2 × 7.5 × 3 cm sized yellow-white lesion located on the cortex and medulla. The lesion overtook the kidney capsule and showed pancreatic adhesions. Microscopic examination of the surgical specimen revealed the

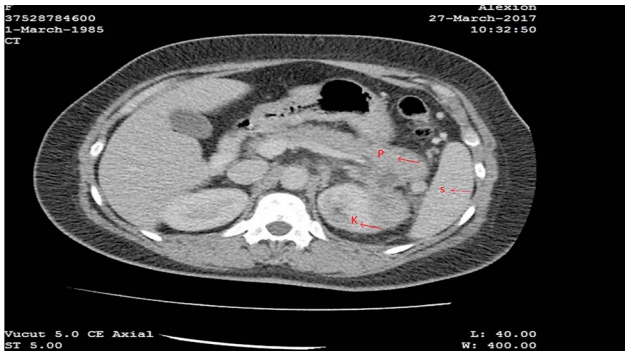

Fig. 1 Contrast-enhanced computed tomography examination shows a solid lesion extending from the middle portion to the upper pole of the left kidney and not clearly distinguished from the pancreatic tail (K: Kidney, P: Pancreas, S: Spleen)

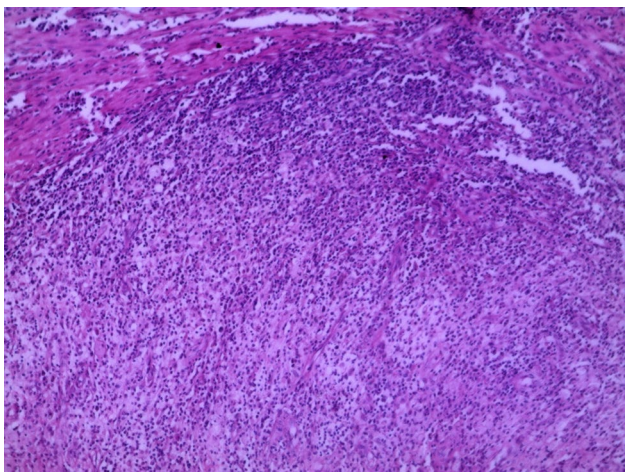

Fig. 2 Histological features of the resected kidney show a granulomatous inflammation with foamy lipid-laden macrophages as well as lymphoplasmacytic inflammation (hematoxylin and eosin stain; original magnification ×10)

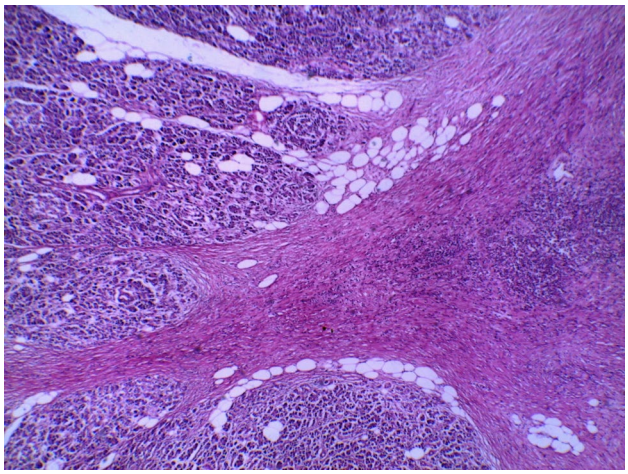

Fig. 3 Granulomatous inflammation extending to pancreas (hematoxylin and eosin stain; original magnification ×10)

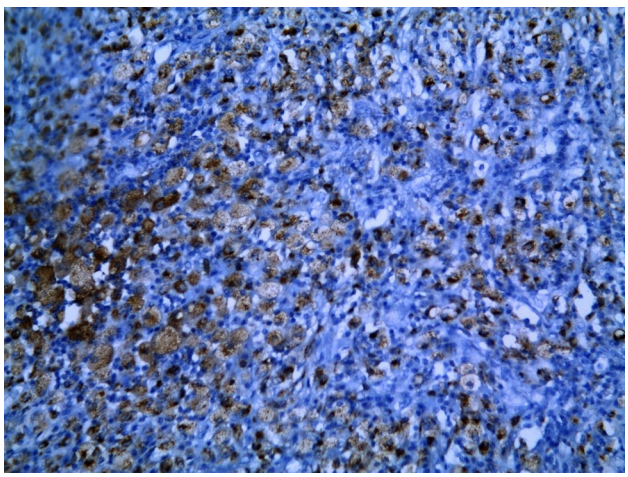

Fig. 4 CD68 positivity in immunohistochemical staining of foam cells (original magnification ×20)

lipid-laden xanthomatous cells (Fig. 2). These cells were extending to pancreas (Fig. 3) and diffusely positive for CD68 (Fig. 4) and negative for CD10 (Fig. 5) and PANCK (Fig. 6) in the immunohistochemical examination.

Discussion

We report a rare case of XGP with adhesion to pancreas and preoperatively diagnosed as malignancy by clinical examination. XGP lesion is considered as a granulomatous reaction causing from severe obstruction and secondary to calculus or rarely tumor [4]. Two forms of morphological involvement are reported; more common diffuse form which spread with pelvic communication and relatively rarer focal form within the renal cortex [5]. Since, focal form strictly mimics renal carcinoma; distinguishing focal XGP from renal cancer is preoperatively difficult [6].

The exact etiology of XGP is unknown; however, it is generally reported that the disease process requires long-term renal obstruction from nephrolithiasis. Additionally, XGP is chiefly associated with *Proteus* or *Escherichia coli* infections; also, *Pseudomonas* species have been implicated [7]. XGP is associated with repeated chronic inflammations, and therefore various fistulas occur including those to the intestine are most common [8].

XGP occurs in a wide range of age, from newborn to elderly, with female-to-male incidence ratio of 2:1. The typical presenting symptoms include flank or abdominal pain, palpable mass, gross hematuria, and weight loss. Most cases of XGP are unilateral and with deranged renal function. The

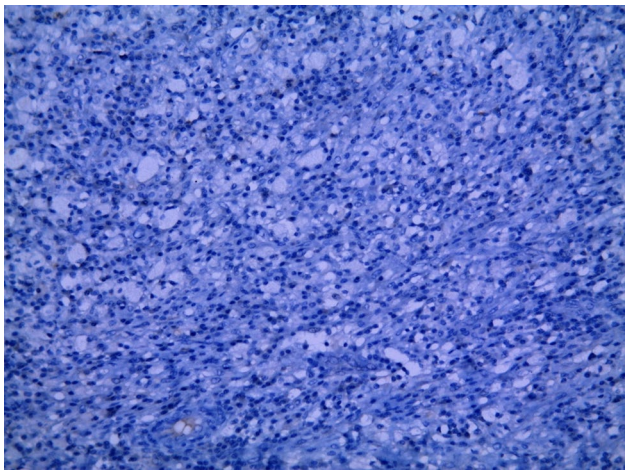

Fig. 5 Negative immunohistochemical staining for foam cell CD10 (original magnification ×20)

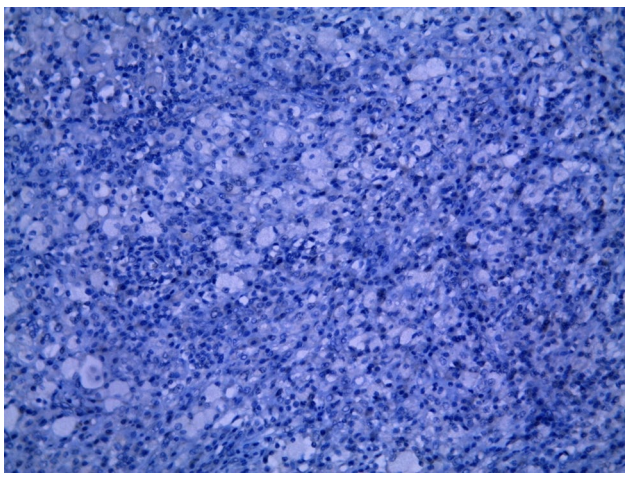

Fig. 6 Negative immunohistochemical staining for foam cell PANCK (original magnification ×20)

common laboratory findings are reported as leukocytosis and anemia. Urine cultures frequently emerge *Escherichia coli* and *Proteus mirabilis* [9].

XGP is further divided into three stages based on whether it is limited to the kidneys or spreads into the surrounding adjacent structures: stage I, nephric disease limited to the kidney; stage II, infiltration into the gerota fascia; and stage III, disease spreading into perinephric tissue. It has been described as a great imitator, even it is often misdiagnosed as a renal mass [10]. Fistula formations were reported as directed to skin, psoas muscle, and intestine tissues among stage III diseases [11]. Conversely, the present case was divergent because there was no determined obstructing calculus. Also, the pain in left lumbar region was the one of the prominent symptoms. Based on the severity of the present case and its adhesion to pancreas it was diagnosed as stage III. Furthermore, we could not find any case report of XGP showing pancreatic adhesions.

The bear's paw sign could also be visualized and reported in some cases. Radiologically, unilateral large kidney, renal pelvis stone, and a nonfunctioning or poorly functioning kidney could also found on computed tomographic imaging. Nevertheless, definitive diagnosis must be performed by histopathological examination of specimens [12].

Computed tomography is the mainstay of diagnostic imaging for XGP; moreover, it reveals the extrarenal extent of the disease and in this way, gives a lead in surgical planning. Treatment of XGP is performed by partial or total nephrectomy, as well as appropriate administration of antibiotics [1]. In the present case, computed tomography exhibited a heterogeneous hypoechoic lesion of kidney extending to pancreas and no evidence of obstructing calculi was detected. Therefore, left total nephrectomy was performed due to preliminary tumor diagnosis. The differential diagnosis of XGP includes clear cell renal cell carcinoma, sarcomatoid renal cell carcinoma, malakoplakia, and leiomyosarcoma [9].

The kidney affected by XGP is nonfunctional, and also threatens life if it is not diagnosed and treated properly [13]. Focal forms of XGP may be treated with partial nephrectomy [14]. Laparoscopic nephrectomy is feasible for some cases of XGP and few cases have been cured by this operative modality and its advantage over radical nephrectomy is being explored [15]. The advantage of kidney preservation must be weighed against the possible additional morbidity compared to standard procedure with nephrectomy [7]. In septic patients and treatment-resistant cases, the emergency nephrectomy is the treatment of choice [16].

XGP exhibits pseudotumoural appearance and treatment should be conservative. Lack of knowledge of this disease may explain the high rate of nephrectomies [6]. Although radical surgery is the main treatment of choice for patients with diffuse XGP, nephron sparing surgery is an alternative for patients who have the focal form, if technically possible [17]. The combination of a nonfunctioning enlarged kidney, a central calculus within a contracted renal pelvis, expansion of the calices, and inflammatory changes in the perinephric fat is strongly suggestive of XGP. Atypical findings are less common and include massive pelvic dilatation, absence of stones, and renal atrophy with or without accumulation of perinephric fat [1].

In conclusion, XGP remains an uncommon entity to encounter on the surgical pathology bench, and it is vital to be aware of the probability of the disease. Due to its similarity to the various benign and malignant conditions, accurate diagnosis needs an association of clinical presentation, imaging studies, as well as pathologic diagnosis which relies on the characteristic morphology of XGP.

Compliance with ethical standards

Conflict of interest All the authors have declared no competing interest.

Informed consent Informed consent was obtained from the patient in this case report.

References

1. Craig WD, Wagner BJ, Travis MD. Pyelonephritis: radiologic-pathologic review. Radiographics. 2008;28(1):255–77.
2. Korkes F, Favoretto RL, Bro´glio M, Silva CA, Castro MG, Perez MD. Xanthogranulomatous pyelonephritis: clinical experience with 41 cases. Urology. 2008;71(2):178–80.
3. Chaudhary R, Singh K, Jain N, Biswas R. Chronic flank pain, fever and an unusual diagnosis. BMJ Case Rep. 2011. https://doi.org/10.1136/bcr.08.2011.4646.
4. Dwivedi US, Goyal NK, Saxena V, Acharya RL, Trivedi S, Singh PB, Vyas N, Datta B, Kumar A, Das S. Xanthogranulomatous pyelonephritis: our experience with review of published reports. Anz J Surg. 2006;76:1007–9.
5. Ghoz HM, Williams M, Perepletchikov A, James N, Babeir AA. An unusual presentation of xanthogranulomatous pyelonephritis: psoas abscess with reno-colic fistula. Oxford Med Case Rep. 2016;7:150–3.
6. Chlif M, Chakroun M, Rhouma SB, Chehida MAB, Sellami A, Gargouri MM, Nouira Y. Xanthogranulomatous pyelonephritis presenting as a pseudotumour. Can Urol Assoc J. 2016;10(1–2):E36-40.
7. Kuo C, Wu CF, Huang CC, Lee YJ, Lin WC, Tsai CW, Wu VC, Chen YM, Wu MS, Chu TS, Wu KD. Xanthogranulomatous pyelonephritis: critical analysis of 30 patients. Int Urol Nephrol. 2011;43(1):15–22.
8. Holton-Burke RC, M Varughese. A case of xanthogranulomatous pyelonephritis associated with renoduodenal fistula. Hindawi Case Rep Med. 2017. https://doi.org/10.1155/2017/8069205 (Article ID 8069205).
9. Li L, Parwani AV. Xanthogranulomatous pyelonephritis. Arch Pathol Lab Med. 2011;135(5):671–4.
10. Arrighi N, Antonelli A, Zani D, Zanotelli T, Corti S, Cunico SC, Simeone C. Review renal mass with caval thrombus as atypical presentation of xantogranulomatous pyelonephritis. A case report and literature review. Urologia. 2013;80:44–47.
11. Snoj Z, Savic N, Regvat J. Late complication of a renal calculus: fistulisation to the psoasmuscle, skin and bronchi. Int Braz J Urol. 2015;41(4):808–12.
12. Loffroy R, Guiu B, Watfa J, Michel F, Cercueil JP, Krause D. Xanthogranulomatous pyelonephritis in adults: clinical and radiological findings in diffuse and focal forms. Clin Radiol. 2007;62(9):884–90.
13. Chow J, Kabani R, Lithgow K, MA Sarna. Xanthogranulomatous pyelonephritis presenting as acute pleuritic chest pain: a case report. J Med Case Rep. 2017;11:101.
14. Wang Peter S. Xanthogranulomatous pyelonephritis. Ultrasound Q. 2016;32:310–1.
15. Pastore V, Niglio F, Basile A, Cocomazzi R, Faticato MG, Aceto G, Bartoli F. Laparascopic-assisted nephroureterectomy for shaped urolithiasis and xanthogranulomatous pyelonephritis: case report and review of literature. Afr J Paediatr Surg. 2013;10(3):285–8.
16. Friedl A, Tuerk C, Schima W, Broessner C. Xanthogranulomatous Pyelonephritis with staghorn calculus, acute gangrenous appendicitis and enterocolitis: a multidisciplinary challenge of kidney-preserving conservative therapy. Curr Urol. 2014;8:162–5.
17. Çaliskan S, Özsoy E, Kaba S, Koca O, Öztürk MI. Xanthogranulomatous pyelonephritis. Arch Iran Med. 2016;19(10):712–4.