Int J Clin Exp Pathol 2015;8(9):11819-11822
www.ijcep.com /ISSN:1936-2625/IJCEP0012475

Case Report

A rare case of xanthogranulomatous pyelonepheritis with hepatic angiomyolipoma

Gongting Zhou, Weijian Hu, Haili Bao, Qiyu Zhang

Department of Hepatobiliary Surgery, The First Affiliated Hospital of Wenzhou Medical University, Fu-Xue Xiang, Wenzhou 325000, Zhejiang, P. R. China

Received July 6, 2015; Accepted August 21, 2015; Epub September 1, 2015; Published September 15, 2015

Abstract: Xantogranulomatous pyelonephritis (XGP) is a rare and chronic variant of pyelonephritis typically associated with obstruction and infection. Xantogranulomatous pyelonephritis can infiltrate liver and connect with hepatic dysfunction. Here we report on a 52-year-old female with a liver mass and a left renal mass detected by uitrasonography and magnetic resonance imaging. Right lobectomy and partial nephrectomy were successively performed. Subsequent pathologic examination indicated hepatic angiomyolipoma and xanthogranulomatous pyelonepheritis, respectively. To the best of our knowledge, this is the first case of such rare combination.

Keywords: Xanthogranulomatous pyelonepheritis, angiomyolipoma, hepatectomy, nephrectomy

Introduction

Xanthogranulomatous pyelonephritis (XGP) is a complicated and chronic inflammatory pathology of the kidney [1]. XGP is a well-recognized pyelonephritis characterized by destruction of the renal parenchyma and granulomatous inflammation with foamy lipid-laden macrophages (xanthoma cells) [2]. The disease usually occurs in middle-age females. Hepatic angiomyolipoma (AML) is a rare hepatic mesenchymal neoplasm which commonly occurs in the kidney and liver [3]. Symptoms of XGP are usually mild and delayed diagnosis is not rare [4]. Therefore, early and accurate diagnosis is imperative for better prognosis. Liver dysfunction can be observed in XGP or AML [5, 6]. The association of renal and hepatic diseases has attracted a great deal of interest [7].

Cases of combination of XGP and liver benign mass have been reported. To our knowledge, a co-existence of xanthogranulomatous pyelonephtitis with hepatic angiomyolipoma is rare and has not been reported yet. Here we present with a case report of a 52-year-old female with xanthogranulomatous pyelonepheritis and a large hepatic angiomyolipoma.

Case presentation

A 52-year-old Chinese woman with hypertension for five years was hospitalized with epigastric pain for one day in August 2014. She denied any history of abdominal discomfort or urinary symptoms.

On admission, physical examination was unremarkable. Laboratory findings showed white-cell count 13.3 × 10^9/L, erythrocyte count 3.85 × 10^{12}/L, hemoglobin 117 g/L, platelet count 166 × 10^9/L, total bilirubin 30 μmol/L, albumin 48.8 g/L, alanine aminotransferase 160 U/L, alkaline phosphatase 275 U/L, prothrombin time 13.1 s, prothrombin activity 103% and activated partial thromboplastin time 45.2 s. There were no abnormalities on viral markers and tumor markers. Magnetic resonance imaging showed a well-defined 10 cm × 10 cm mass occupying in the right lobe and a wedge-shaped mass measuring 2.0 cm × 1.6 cm in left kidney (**Figure 1A**). Magnetic resonance imaging revealed the kidney mass with hypointensity on T1-weighted, hypointensity on T2-weighted (**Figure 1A**, **1B**). Contrast-enhanced T1-weight imaging revealed that both kidney and liver masses are nonenhancing lesions (**Figure 1C**). The combination of hepatic hamartoma and

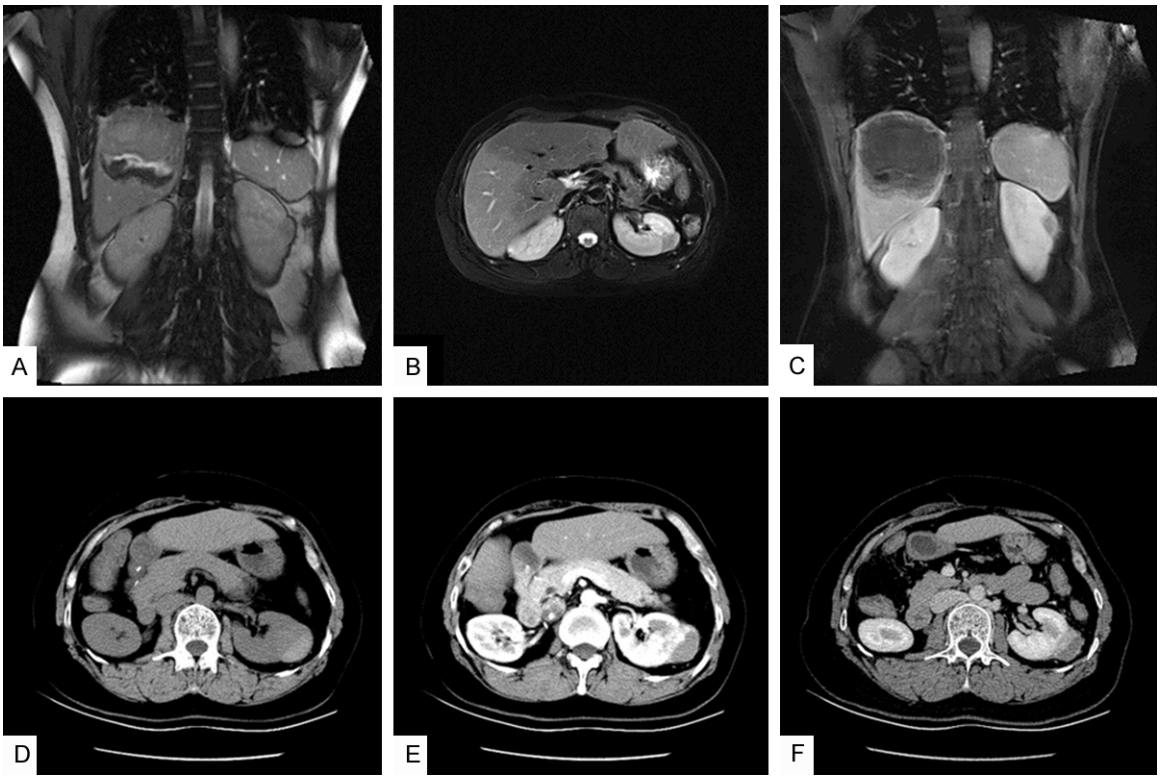

Figure 1. A: Magnetic resonance imaging showed a left kidney mass (2.0 cm × 1.6 cm) with hypointensity on T1-weighted and a heterogeneous mass (10 cm × 10 cm) on T1-weighted in liver. B: MRI showed the left kidney mass with hypointensity on T2-weighted. C: Contrast enhanced T1-weight imaging showed the absence of enhancement of the kidney mass and the liver mass. D: The kidney mass was high-density on computed tomography. E, F: Enhanced CT showed a kidney mass (3.8 cm × 3.3 cm) with the absence of early-phase and late-phase enhancement.

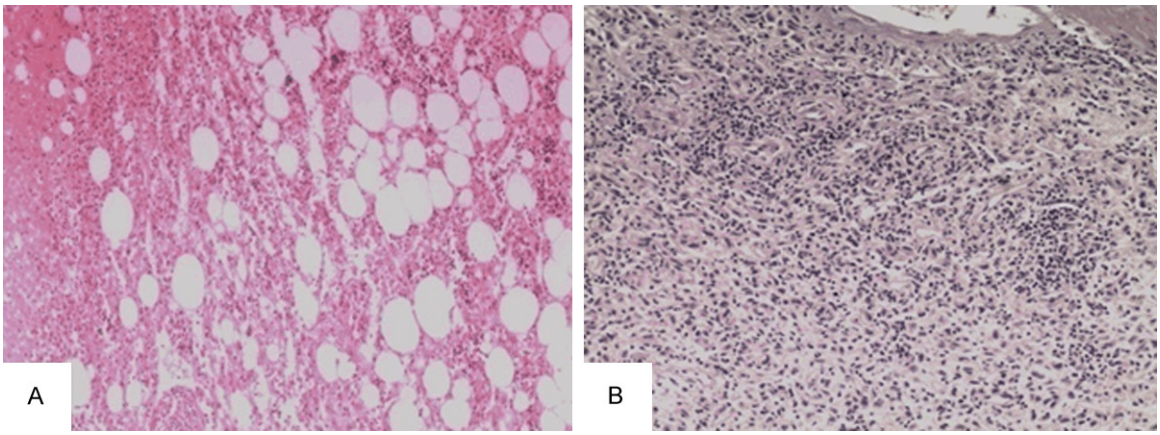

Figure 2. A: The hepatic mass consisted of a mixture of smooth muscle cells, mature adipose tissue and abnormal thick-walled blood vessels. B: Photomicrograph from the resected kidney mass showed spindle cells mixed with abundant foamy cells and scattered lymphocytes (hematoxylin and eosin stain [H&E], A with original magnification × 100, B with original magnification × 100).

kidney hamartoma was suspected. Due to the acute epigastric pain and suspection of internal hemorrhage of the large liver mass, right lobectomy was performed preferentially. Postoperatively, pathologic evaluation of the specimen indicated that the liver mass was a hepatic angiomyolipoma (**Figure 2A**). Immediate treatment for the kidney mass was considered unnecessary.

After 7 months of follow-up, plain CT scan showed that the size of the kidney mass had

obviously increased, reaching size of 3.8 cm × 3.3 cm (**Figure 1D**). She was admitted to our hospital in 2015 for further examination of the left kidney mass which firstly discovered 7 months back. We found the absence of enhancement after intravenous injection of contrast medium (**Figure 1E**, **1F**). Left partial nephrectomy was performed. The diagnosis of xanthogranulomatous pyelonepheritis was confirmed by immunohistochemical examination (**Figure 2B**).

The patient recovered well with uneventful post-operative period and was discharged on postoperative day 9. The patient was last seen at 4 months follow up and has progressed well.

Discussion

Xanthogranulomatous pyelonephritis, first described by Schlagenhaufer in 1916, is a rare and chronic pyelonephritis which is usually associated with nephrolith, urinary tract obstruction and infection. Many cases result in a nonfunctioning kidney associated with severe obstruction, calculus, stricture or rarely tumor [8].

Gross morphological changes of XGP include kidney enlargement, hydronephrosis, peripelvic fibrosis and lobulated yellow masses [2]. The etiology of XGP is still undefined, however impairment of the urinary flow and chronic bacterial infection have been regarded as consistently associated factors that connect with the development of XGP [9]. *E. coli* and *P. mirabilis* have been identified as the most commonly implicated organisms [10]. Patients with XGP may experience flank pain, fever, weight loss, gross haematuria, malaise, anorexia, draining sinus and flank mass [11, 12].

Angiomyolipoma is a rare mesenchyme-derived neoplasm that most commonly occurs in the kidney, but can also be found in the liver [3, 13]. Most hepatic AML is benign and rare malignant hepatic AML has been reported [14, 15]. The etiology remains unknown, however, there are reports showed some hepatic AML associated with liver dysfunction. With regard to liver dysfunction, the combination of xanthogranulomatous pyelonephritis and hepatic AML make clinical features vaguer, as liver dysfuntion in our case. For liver dysfunction may occurs in XGP or AML [5, 6].

XGP usually simulates a renal tumor. Although, in addition to clinical features, sensitive radiologic investigations (CT, MRI) have made preoperative diagnosis of XGP possible [16]. However, the correct diagnosis of XGP is confirmed mostly by pathology examination after surgery such as our patient.

Several cases of xantogranulomatous pyelonephritis were reported to be in contact with the liver. After performing a literature search, we found 2 reports of liver inflammatory extension infiltrated from XGP. Several cases of xantogranulomatous pyelonephritis were reported to be in contact with the liver. Taskinen et al reported a kidney mass combined with caliceal diverticulum infiltrating the liver [17]. In another report, Karaman et al reported a patient with XGP with inflammatory infiltration in an unconnected liver segment [18]. Moreover, there was a case report of co-existence of xanthogranulomatous pyelonepheritis with renal angiomyolipoma, who was successfully treated by radical nephrectomy and who is free of disease for the benign nature [19]. The case of XGP combined with a large hepatic angiomyolipoma had not been published. Radical surgery with nephrectomy or partial nephrectomy remains standard and necessary treatment for XGP. Partial nephrectomy is recommended whenever possible, especially in focal type [18].

It is recommended to resect all diseased tissue in cases of XGP. In our case, partial nephrectomy was performed for resection of the increasing tissue. Series have shown that poor functioning or functionless kidneys were seen in 83% of patients presenting with XGP, implying delayed presentation [20]. Early diagnosis and treatment of such case can keep larger segment of kidney.

Although preoperative renal prognosis may be uncertain, early surgery and medical treatment can be effective. The diagnosis and treatment for such coexistence of unique lesions was considered worth mentioning. In summary, although co-existence of xanthogranulomatous pyelonepheritis with hepatic angiomyolipoma is extremely rare, a possible diagnosis should not be ignored. In our case, both xanthogranulomatous pyelonepheritis and hepatic angiomyolipoma were misdiagnosed as hamartoma. As surgery has been the preferable method of treatment for xanthogranulomatous pyeloneph-

eritis [6], preoperative evaluation and discussion must be made with caution in similar case. Such combination must be ruled out before a treatment decision with neglect of xanthogranulomatous pyelonepheritis, particularly for lager liver mass which may make symptom more ambiguous.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Qiyu Zhang, Department of Hepatobiliary Surgery, The First Affiliated Hospital of Wenzhou Medical University, Fu-Xue Xiang, Wenzhou 325000, Zhejiang, P. R. China. E-mail: zqydoc@163.com

References

[1] Chang JW, Chen SJ, Chin TW, Tsai H, Pan CC, Chu YK, Tiu CM and Yang LY. Xanthogranulomatous pyelonephritis treated by partial nephrectomy. Pediatr Nephrol 2004; 19: 1164-1167.

[2] Muta T, Takamatsu Y, Morita S, Kikuchi I and Tamura K. Xanthogranulomatous pyelonephritis. Intern Med 1997; 36: 586-90.

[3] Kreel L. Renal and hepatic angiomyolipoma. Postgrad Med J 1985; 61: 325-327.

[4] Dwivedi US, Goyal NK, Saxena V, Acharya RL, Trivedi S, Singh PB, Vyas N, Datta B, Kumar A and Das S. Xanthogranulomatous pyelonephritis: our experience with review of published reports. ANZ J Surg 2006; 76: 1007-1009.

[5] Kuo CC, Wu CF, Huang CC, Lee YJ, Lin WC, Tsai CW, Wu VC, Chen YM, Wu MS, Chu TS and Wu KD. Xanthogranulomatous pyelonephritis: critical analysis of 30 patients. Int Urol Nephrol 2011; 43: 15-22.

[6] Chang Z, Zhang JM, Ying JQ and Ge YP. Characteristics and treatment strategy of hepatic angiomyolipoma: a series of 94 patients collected from four institutions. J Gastrointestin Liver Dis 2011; 20: 65-69.

[7] Vermillion SE, Morlock CG, Bartholomew LG and Kelalis PP. Nephrogenic hepatic dysfunction: secondary to tumefactive xanthogranulomatous pyelonephritis. Ann Surg 1970; 171: 130-136.

[8] Claes H, Vereecken R, Oyen R and Van Damme B. Xanthogranulomatous pyelonephritis with emphasis on computerized tomography scan. Retrospective study of 20 cases and literature review. Urology 1987; 29: 389-393.

[9] Antonakopoulos GN, Chapple CR, Newman J, Crocker J, Tudway DC, O'Brien JM and Considine J. Xanthogranulomatous pyelonephritis. A reappraisal and immunohistochemical study. Arch Pathol Laborat Med 1988; 112: 275-281.

[10] Chuang CK, Lai MK, Chang PL, Huang MH, Chu SH, Wu CJ and Wu HR. Xanthogranulomatous pyelonephritis: experience in 36 cases. J Urol 1992; 147: 333-336.

[11] Grainger RG, Longstaff AJ and Parsons MA. Xanthogranulomatous pyelonephritis: a reappraisal. Lancet 1982; 1: 1398-401.

[12] Fernandez Miranda C, Vigil Merino L, Palacio Perez-Medel A, Camacho Herreros M, Usera Sarraga G and Navas Palacios JJ. [Xanthogranulomatous pyelonephritis. Study of 18 cases with special reference to hepatic dysfunction]. Rev Clin Esp 1981; 163: 259-264.

[13] Zeng JP, Dong JH, Zhang WZ, Wang J and Pang XP. Hepatic angiomyolipoma: a clinical experience in diagnosis and treatment. Digest Dis Sci 2010; 55: 3235-3240.

[14] Zhou YM, Li B, Xu F, Wang B, Li DQ, Zhang XF, Liu P and Yang JM. Clinical features of hepatic angiomyolipoma. Hepatobiliary Pancreat Dis Int 2008; 7: 284-287.

[15] Wang WT, Li ZQ, Zhang GH, Guo Y and Teng MJ. Liver transplantation for recurrent posthepatectomy malignant hepatic angiomyolipoma: a case report. World J Gastroenterol 2015; 21: 3755-3758.

[16] Zugor V, Schott GE and Labanaris AP. Xanthogranulomatous pyelonephritis in childhood: a critical analysis of 10 cases and of the literature. Urology 2007; 70: 157-160.

[17] Taskinen S, Giordano S and Rintala R. Xanthogranulomatous pyelonephritis infiltrating the liver. J Pediatr Surg 2008; 43: e7-9.

[18] Karaman A, Samdanci E, Dogan M, Aksoy RT, Sigirci A and Demircan M. Xanthogranulomatous pyelonephritis with unconnected liver lesion. Urology 2011; 78: 189-191.

[19] Nabi G, Greene D, Marsh R and Viljoen A. A rare co-existence of focal xanthogranulomatous pyelonepheritis, angiomyolipoma and renal cysts simulating renal cell carcinoma. Intl Urol Nephrol 2002; 34: 465-466.

[20] Samuel M, Duffy P, Capps S, Mouriquand P, Williams D and Ransley P. Xanthogranulomatous pyelonephritis in childhood. J Pediatr Surg 2001; 36: 598-601.