Case Report
A rare case of xanthogranulomatous pyelonephritis with hepatic angiomyolipoma

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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 ultrasonography and magnetic resonance imaging. Right lobectomy and partial nephrectomy were successively performed. Subsequent pathologic examination indicated hepatic angiomyolipoma and xanthogranulomatous pyelonephritis, respectively. To the best of our knowledge, this is the first case of such rare combination.

Keywords: Xantogranulomatous pyelonephritis, angiomyolipoma, hepatectomy, nephrectomy

Introduction

Xantogranulomatous 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 pyelonephritis 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 pyelonephritis 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-weighted imaging revealed that both kidney and liver masses are nonenhancing lesions (Figure 1C). The combination of hepatic hamartoma and
kidney hamartoma was suspected. Due to the acute epigastric pain and suspicion 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 pyelonephritis 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 Schlegenhauer 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 dysfunction 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 xanthogranulomatous 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 xanthogranulomatous 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 pyelonephritis 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 pyelonephritis with hepatic angiomyolipoma is extremely rare, a possible diagnosis should not be ignored. In our case, both xanthogranulomatous pyelonephritis and hepatic angiomyolipoma were misdiagnosed as hamartoma. As surgery has been the preferable method of treatment for xanthogranulomatous pyeloneph-
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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 pyelonephritis, particularly for larger liver mass which may make symptom more ambiguous.

Disclosure of conflict of interest

None.

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References