An Unusual Presentation of Polyarteritis Nodosa
A Case Report
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ABSTRACT

Polyarteritis nodosa with gallbladder involvement is a rare condition. Autosomal dominant polycystic kidney disease is also a rare condition and rarely complicated. We describe an extremely rare case of Polyarteritis nodosa, involving gallbladder and ureter without obstruction, in a patient with autosomal dominant polycystic kidney disease. To the best of the authors’ knowledge, such a case has not been reported previously.

Case Report

A 41-year-old man was referred to our clinic with the complaints of right upper quadrant pain, fever, nausea, vomiting, loss of weight and weakness. Physical examination revealed right upper quadrant and bilateral lumbar tenderness with fever (38.7 °C). The white blood cell count was (12700/mm3), erythrocyte sedimentation rate (115 mm/h) (5–20mm/hour for men), concentration of C-reactive protein 196 mg/dL (0–5 mg/dL). Ultrasound imaging showed multiple cysts in both kidneys with diverticula in the first and second sections. Ultrasound imaging of the abdomen revealed a dilated common bile duct with a diameter of 1 cm, diffuse wall thickening of the gallbladder and liver, and dilatation of the gallbladder. Laboratory tests revealed an elevation of liver enzymes, bilirubin, and alkaline phosphatase. The diagnosis of Polyarteritis nodosa with gallbladder involvement was made based on the clinical presentation and imaging findings. The patient was treated with prednisolone and aspirin, and the symptoms improved significantly. To our knowledge, this is the first reported case of Polyarteritis nodosa involving gallbladder and ureter without obstruction in a patient with autosomal dominant polycystic kidney disease.
mg/dL), serum alkaline phosphatase 801 IU/l (66 to 220 IU/L), C3 1.028 g/L (0.50–0.90 g/L), total protein; 5.9 g/dL (6.4-8.3 g/dL) and albumin 2 g/dL (3.5–5.5 g/dL) levels. Urinalysis and other biochemical tests were within normal limits. Viral hepatitis and hydatid cyst screening tests were negative.

The contrast-enhanced computed tomography (CT) scan revealed multiple well defined hypodense cystic lesions of different size and location predominantly in the left lobe of the liver. The gallbladder wall was thickened diffusely with partial contraction (Fig. 1). The largest cyst was 35 mm in diameter. One of the cysts in the left lobe was thought to be infected because of its minimal high attenuation (16 HU). There were large cysts in the right kidney and multiple small ones in the left with minimal bilateral nephromegaly (Fig. 2).

The ultrasonographic findings were similar to those of the CT scan. Bilateral dilated proximal ureters were also noted (Fig. 3). The intravenous urography (IVU) study showed dilated collecting system of both kidneys and ureters. Retrograde urography showed no obstruction.

The patient was treated with gentamicin and ampicillin for nine days and metronidazole during the first five days. During the first nine hospital days, the white blood cell count, erythrocyte sedimentation rate and alkaline phosphatase levels fell, but the fever and pain persisted. At surgery, multiple cysts in the liver were observed, predominantly in the left lobe and the gallbladder was contracted and thickened. The kidneys were larger than normal and had multiple cysts. The fluid was clear and the cysts were not associated with the biliary tract as demonstrated by intraoperative cholangiography. Cholecystectomy was done and biopsies were taken from the cysts. Also, multiple lymph nodes were excised from the hepato-duodenal ligament, head of pancreas and superior pancreatic region. Histology revealed arteritis and polycystic liver hyperplasia. In addition, p-antineutrophil cytoplasmic antibodies (p-ANCA) was positive. Retrospectively, the cause of the bilateral hydronephrosis without obstruction was thought to be due to PAN.
ADPKD, in contrast to patients with noncystic hepatic abscesses in whom polymicrobial infections are most common (7).

Computed tomography has greater than 90% sensitivity for the detection of hepatic abscesses, which appear as low attenuation, rounded masses on both non-contrast and contrast enhanced scans (8). The attenuation ranges between 0 and 45 HU and overlaps with that of other lesions such as cysts, bilomas, and neoplasm (8).

In this case, one of the cysts in the liver was thought to be infected because of its minimally high attenuation (16 HU), the clinical and laboratory findings (fever, weakness, anorexia, high sedimentation rate and white blood cell count, decreased albumin and total protein) were in accordance with findings of general infection. In addition, thickening of the gallbladder wall alone at CT is not specific for acute cholecystitis and can be seen in a contracted gallbladder, hepatic cirrhosis, hepatitis, ascites, pancreatitis, congestive heart failure, renal failure, hypoalbuminaemia, adenomyomatosis, portal hypertension, multiple myeloma and gallbladder carcinoma (8).

If multiple cysts are concentrated in one aspect of the liver, resectional therapy may be indicated (9). Surgery is typically reserved for refractory or recurrent abscesses or abscesses with other intra-abdominal pathologies (9). In this case, cysts were predominantly located in the left lobe of the liver and preoperatively, it was decided to resect the left lobe and enucleate the cysts in the right lobe.

When patients without previously established PAN diagnosis are referred because of abdominal pain and fever, diagnosis is difficult and surgery is directed towards the lesion(s) producing the acute symptoms, most often leading to resection of the pathologic tissue and/or control of bleeding. Histologic examination establishes the diagnosis, and is usually followed by medical evaluation and treatment (2). The index patient was operated on with the initial diagnosis of infected ADPKD and thickened gallbladder wall, but it was observed during the operation that the cystic fluid was not infected. Histopathological diagnosis was consistent with arteritis. In addition, p-ANCA was found to be positive. Following immunosuppressive treatment, regression of signs and symptoms was observed, together with the disappearance of p-ANCA.

Determination of ANCA and its specificities is a useful adjunct to the classification of patients with biopsy-proven necrotizing arteritis (10). Serial measurement of p-ANCA titers in patients with ANCA-associated vasculitis during remission can help predict relapses and pre-emptive increase in immunosuppression therapy following four-fold titer rise reduces the risk of relapses. Moreover, adjustment of immunosuppression based on lesser titer changes appears to result in a favourable outcome (10). Persistence of positive p-ANCA may be a marker of an underlying disease process, but does not adequately reflect disease activity, thus, in no case.

The patient was treated with prednisone at a dose of 1 mg/kg/day for 1 month. The daily dose was decreased by 2.5 mg every week until a dose of 10 mg/day was reached at approximately the sixth month. Then, the dose was decreased by 1 mg every week. Cyclophosphamide was infused monthly at a dose of 750 mg/month for 8 months. After this treatment, the patient recovered and p-ANCA was decreased to normal levels after four months (Fig. 4).

DISCUSSION

Autosomal dominant polycystic kidney disease is a rare condition with a prevalence of approximately 0.1% (4, 6). Complications of hepatic cysts are uncommon and infection is one of them. Secondary infection with severe consequences, may occur after an episode of acute cholangitis or septicaemia even though, usually, no direct communication exists with the biliary tract (4, 6).

Infected hepatic cysts develop in up to 3% of patients with ADPKD who have end-stage renal failure, but in less than 1% of those without. Monomicrobial infections with Enterobacteriaceae seem to predominate in patients with ADPKD, in contrast to patients with noncystic hepatic abscesses in whom polymicrobial infections are most common (7).

Fig. 4: After the treatment for PAN for two years, both of the pelvicalyceal systems and ureters seem to be normal on intravenous urogram. The simple cyst compressing the upper pole of the right kidney was still present.
should it be the only indication for therapeutic intensification (10).

There was bilateral hydronephrosis without obstruction which was ascribed to PAN retrospectively. Ureteric involvement at presentation of PAN is unusual. In most cases, single or multiple narrowed areas were described in one or both of the ureters which in pathological examination revealed vasculitis involving predominantly the adventitial layer of the ureter, but also the muscular layer as well. To our knowledge, there is only one case report in the literature about bilateral hydronephrosis without obstruction (3). One of the most striking features of this case was bilateral dilatation of the collecting systems without obstruction. Retrograde urography showed no obstruction. Retroperitoneal fibrosis was not detected on CT. After the operation there was low-grade fever in the patient which subsided after corticosteroid and cytotoxic agent therapy.

This case study illustrates the potential difficulty in making the diagnosis of polyarteritis nodosa. This condition is rare and polyarteritis nodosa commonly presents with non-specific constitutional symptoms (eg fever, malaise and weight loss). The diagnosis of this disease is especially difficult if the initial diagnosis is masked by other symptoms.

REFERENCES