Case Report

Duodenal diverticulosis in autosomal dominant polycystic kidney disease

Sumedha Kumar¹, Magdalena Adeva¹, Bernard F. King², Patrick S. Kamath³ and Vicente E. Torres¹

¹Department of Internal Medicine, Division of Nephrology and Hypertension, ²Department of Diagnostic Radiology and
³Department of Internal Medicine, Division of Gastroenterology, Mayo Clinic College of Medicine, Rochester,
MN 55905, USA

Keywords: autosomal dominant polycystic kidney disease; duodenal diverticulosis

Introduction

Autosomal dominant polycystic kidney disease (ADPKD) accounts for approximately 5% of end-stage renal disease (ESRD) in USA and Europe. It is a systemic disease with multiple extrarenal manifestations, including colonic diverticulosis. Diverticulitis and diverticular perforation contribute to the morbidity and mortality of ADPKD. Whether extracolonic diverticulosis is associated with ADPKD is uncertain. We report a series of eight ADPKD patients with duodenal diverticulosis. In the index case, the clinical presentation was symptomatic diverticular disease.

Case reports

A 72-year-old white woman on haemodialysis for 6 years for end-stage ADPKD, presented with a weight loss of 40 pounds. She had experienced episodes of emesis, mostly 3–4 h post-prandially, for approximately 1 year and diarrhoea for the past 6 months. Physical examination revealed large polycystic kidneys occupying both flanks down to pelvis, bilateral pedal oedema and a ventral hernia. Laboratory evaluation revealed a serum albumin of 2.8 gm/dl and an International Normalized Ratio (INR) of 1.9. Serum folate and B-12 levels were normal. Serum ferritin was 228 mcg/l. Stool fat was 8 g/24 h. Tests for sprue were negative. MRI showed large cystic kidneys and a large duodenal diverticulum with an air fluid level (Figure 1). Esophagagogastroduodenoscopy revealed a non-occlusive Schatzki’s ring; gastric mucosal atrophy and a post-bulbar duodenal diverticulum. Culture of duodenal aspirate showed more than 100,000 aerobic bacteria per ml, consistent with bacterial overgrowth. Colonoscopy showed sigmoid colonic diverticulosis. She was treated with ciprofloxacin 500 mg daily for 2 weeks with resolution of the symptoms.

To find additional cases of duodenal diverticular cases associated with ADPKD, we searched our ADPKD database. We identified seven additional cases in which the diagnosis of duodenal diverticulosis appeared to be incidental. Their clinical presentation, laboratory parameters, and associated findings are summarized in Table 1.

Discussion

Colonic diverticular disease is often considered to be an extrarenal manifestation of ADPKD. Schef et al. [1] found diverticulosis on barium enema studies in 10 of 12 haemodialysis patients with ADPKD (83%), compared with 13 of 31 haemodialysis patients with chronic renal failure from causes other than ADPKD (32%). McCune et al. [2] found diverticular disease on colonoscopy or barium enema in 7 of 14 patients with ADPKD (50%), and 13 of 86 patients with other causes of ESRD (15%) aged >50 years evaluated for renal transplantation. Lederman et al. [3] found that 12 of 59 patients with ESRD secondary to ADPKD (20%) had a history of acute diverticulitis, whereas only 4 of 125 patients with ESRD due to other causes (3%) had diverticulitis. Whether the increased risk for diverticulosis and its complications extends to patients with ADPKD prior to ESRD is uncertain, since the prevalence of diverticular disease in 55 non-ESRD patients with ADPKD was not different from that in the general population in a study from the University of Colorado [4].

Whether ADPKD patients are at increased risk for extracolonic diverticular disease is not known. Only one case of this association has been reported;
a 75-year-old woman on haemodialysis for end-stage ADPKD [5]. This patient had multiple jejunal and ileal diverticula and presented with a diverticular perforation. Here, we report eight patients with duodenal diverticulosis. Of these, four had ESRD (three of them on haemodialysis and one after renal transplantation) thus suggesting an increased occurrence of duodenal diverticular disease in ADPKD patients with ESRD, as it is the case with colonic diverticular disease. The average age of diagnosis of diverticular disease in these patients was 64 years, consistent with duodenal diverticulosis as mainly a disease of the elderly. Four out of five patients with colonoscopies or barium enemas had associated colonic diverticular disease.

Diverticular disease of the duodenum was first described by Chomel in 1710 [6]. Prevalence of duodenal diverticulosis ranges from 1 to 5% in radiological studies and from 11 to 22% in autopsy series. Prevalence increases with age and it is more common in women than in men [7]. It is the second most common form of diverticulosis after colonic diverticulosis. In two-thirds of cases, it is found in the second portion of the duodenum, near the ampulla of Vater. Two forms are known: intraluminal and extraluminal, the latter being more common. It becomes clinically significant in a minority of persons. It may present clinically as nausea, vomiting, abdominal pain and malabsorption. Complications may include ulceration, haemorrhage, duodenal or common bile duct obstruction, pancreatitis and perforation into retroperitoneum. Perforation is the rarest complication but has a high mortality, 13–34% [6]. Choledocholithiasis, common bile duct dilatation, cholecystolithiasis and pancreatitis have been reported in association with juxtapapillary diverticulosis [8]. In fact, six of the eight patients in our series had biliary disease (Table 1) and three patients required an endoscopic retrograde cholangiopancreatography (ERCP). In patients with polycystic liver disease and

![Image](image-url)

Fig. 1. Axial, T2 weighted, image at the level of the kidneys. Note the diffuse involvement of polycystic disease in both kidneys. Situated between both kidneys, in the midline, is a large duodenal diverticulum with an air-fluid level.

Table 1. Clinical characteristics of eight patients with ADPKD and Duodenal Diverticulosis

<table>
<thead>
<tr>
<th>No.</th>
<th>Age/Gender</th>
<th>Clinical diagnosis</th>
<th>AST</th>
<th>ALT</th>
<th>ALP</th>
<th>Bilirubin</th>
<th>Creatinine at presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>72/Female</td>
<td>Duodenal diverticulosis with bacterial overgrowth</td>
<td>49</td>
<td>183</td>
<td>0.9</td>
<td>Haemodialysis</td>
<td>Yes</td>
</tr>
<tr>
<td>2</td>
<td>54/Male</td>
<td>Ascending cholangitis from reflux into choledochojejunostomy</td>
<td>35</td>
<td>407</td>
<td>1.2</td>
<td>Haemodialysis</td>
<td>Yes</td>
</tr>
<tr>
<td>3</td>
<td>66/Male</td>
<td>Cholelithiasis with stricture of right hepatic duct</td>
<td>25</td>
<td>75</td>
<td>0.9</td>
<td>Haemodialysis</td>
<td>Yes</td>
</tr>
<tr>
<td>4</td>
<td>64/Female</td>
<td>Gastroesophageal reflux</td>
<td>20</td>
<td>183</td>
<td>0.6</td>
<td>Post-renal transplant</td>
<td>Yes</td>
</tr>
<tr>
<td>5</td>
<td>54/Female</td>
<td>Benign stricture of right hepatic duct</td>
<td>126</td>
<td>262</td>
<td>0.5</td>
<td>Yes</td>
<td>2nd portion</td>
</tr>
<tr>
<td>6</td>
<td>66/Male</td>
<td>Cholecystolithiasis, sigmoid colon polyp</td>
<td>128</td>
<td>258</td>
<td>0.6</td>
<td>Yes</td>
<td>2nd portion</td>
</tr>
<tr>
<td>7</td>
<td>64/Female</td>
<td>Hepatic artery stenosis</td>
<td>125</td>
<td>198</td>
<td>1.7</td>
<td>Yes</td>
<td>4th portion</td>
</tr>
<tr>
<td>8</td>
<td>66/Female</td>
<td>Hepatic vein stenosis</td>
<td>25</td>
<td>108</td>
<td>1.7</td>
<td>Yes</td>
<td>4th portion</td>
</tr>
</tbody>
</table>
biliary symptoms, the possibility of a duodenal diverticulum should be kept in mind since a peri-ampullary duodenal diverticulum can make cannulation of the common bile duct at ERCP more challenging. Diagnosis of duodenal diverticulosis can be made on upper gastrointestinal (GI) series, MRI, CT, ERCP and esophagogastroduodenoscopy. Diverticulitis is best diagnosed by CT scan [9]. Management depends upon the status of the patient. In acute setting and when there is uncertainty about diagnosis, surgery is preferred, whereas in mild cases patients are treated with bowel rest and antibiotics covering both gram negative and anaerobic organisms. On occasion, total parenteral nutrition may be required if nutritional needs cannot be met by mouth.

The coexistence of ADPKD and extracolonic diverticulosis may be due to chance and not reflect a true association. Nevertheless, advances in the understanding of the molecular genetics and biology of ADPKD provide a possible explanation for such association. Polycystin 1 and polycystin 2 are expressed in vascular and intestinal smooth muscle [10,11]. Pkd2+/− haploinsufficiency in smooth muscle has been associated with abnormal contractility, possibly due to alterations on intracellular calcium homoeostasis [12,13]. Therefore, it seems possible that subtle alterations in the function and/or structure of intestinal smooth muscle directly linked to PKD1/PKD2 mutations may predispose ADPKD patients to diverticular disease. It has been recently hypothesized that the diverticulosis may be attributable to smooth muscle dysfunction, resulting from vagal attrition associated with ageing [14]. Diverticular disease may become more clinically relevant with the increasing life expectancy of ADPKD patients, resulting from improvements in the treatment of hypertension, dialysis and renal transplantation. A diagnosis of duodenal diverticulosis and its associated complications should thus be considered while treating patients of ADPKD with ESRD presenting with gastrointestinal symptoms.

Conflict of interest statement. None declared.

References


Received for publication: 27.5.06
Accepted in revised form: 13.6.06

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