Case Report

Thoracic aortic dissection complicating autosomal dominant polycystic kidney disease

H. E. Paynter, A. Parnham, T. G. Feest and C. R. K. Dudley

Gloucestershire Royal Hospital, Gloucester, and Southmead Hospital, Bristol, UK

Key words: autosomal dominant polycystic kidney disease; thoracic aortic dissection

Introduction

A number of vascular abnormalities have been associated with autosomal dominant polycystic kidney disease (ADPKD), including heart valve lesions (mitral valve prolapse, mitral incompetence, tricuspid regurgitation, tricuspid valve prolapse, and bicuspid aortic valve) [1], cerebral aneurysms [2], aortic root dilatation [3], coarctation of the aorta [3], and abdominal aortic aneurysm [4]. Thoracic aortic dissection has been reported rarely [5–8]. We present a case of thoracic aortic dissection occurring in a young woman with ADPKD.

Case report

A 36-year-old Caucasian female was referred to us in June of 1994 with known polycystic kidney disease and a serum creatinine of 200 μmol/l. Her father had died in his thirties of renal failure, and her brother had required dialysis at the age of 33. Four years previously she had been diagnosed as having coeliac disease, based on a biopsy and response to a gluten-free diet. She smoked 20 cigarettes a day.

At initial referral she was hypertensive (200/120), and this was treated with angiotensin-converting enzyme inhibitors (initially captopril, later lisinopril). Her blood pressure during follow-up varied between 130/80 and 150/100. Nifedipine was later substituted for lisinopril because of hyperkalaemia. In 1995 she fell in the bath and sustained fractured ribs, and a traumatic right pneumothorax, which was treated by insertion of a chest drain. Over the next few months her renal function continued to deteriorate, and it was predicted that she would reach end-stage renal failure by mid-1996.

In May 1996 she was admitted as an emergency with severe indigestion-like chest pain, radiating to her neck and jaw. In association with this, she was complaining of breathlessness and palpitations.

On examination she was of normal habitus with no evidence of Marfan’s syndrome. Her pulse was 60 b.p.m., regular, and collapsing in nature. Her blood pressure was 150/50 in the right arm, 130/50 in the left, and there was no elevation of her jugular venous pressure. She had a long, loud early diastolic murmur, most marked at the left sternal edge. Her left radial, femoral, and dorsalis pedis pulses were weaker than the right. Her chest had coarse bibasal crepitations to the mid-zones, and apart from large palpable polycystic kidneys, her abdominal examination was normal. She had no focal neurological signs.

An ECG showed biphasic T waves in the anteroseptal leads. Emergency CT of the thorax demonstrated dissection within the descending thoracic arch (Figure 1). She was transferred immediately for aortic replacement.

At operation an intimal tear was identified within the left coronary sinus, and the false lumen extended into the arch of the aorta, rupturing near the innominate artery. A composite aortic graft and valve were implanted.

Her recovery was complicated by the development of haemopericardium and cardiac tamponade requiring surgical evacuation. She required haemofiltration immediately after the bypass surgery, and failed to recover her residual renal function. Subsequently she has made a good recovery and is now established on continuous ambulatory peritoneal dialysis.

Histology of the excised aorta was normal; in particular it showed no cystic medial necrosis.

Discussion

This 36-year-old woman with mild to moderate hypertension, and ADPKD, suffered a type I thoracic aortic dissection. The degree of hypertension is unlikely to be causative, as there were no histological changes of
hypertension in the excised aorta. Marfan’s syndrome can be excluded on both histological and phenotypic grounds. The dissection was not immediately preceded by trauma, and there is no evidence that aortic dissection may be a late complication of trauma. In this case, therefore, it seems likely that there is a direct association between ADPKD and aortic dissection.

One autopsy series describes thoracic aortic dissection to be seven times more common in patients with ADPKD than in the general population [8]. However, only a few cases have been reported in the literature. They form a heterogeneous group in terms of age, sex, degree of renal failure, and habitus (Table 1). Our case is the first clearly reported to be in the presence of a normal habitus. The histology of the excised aorta has not always been described, but cystic medial necrosis is the only reported finding. This is the histological abnormality characteristic of aortic dissection in Marfan’s [9] and Ehlers–Danlos [10] syndromes. Interestingly, the aorta resected in our patient was normal.

The association of ADPKD with berry aneurysms, vertebral artery dissection, and abdominal aortic aneurysms, in addition to thoracic aortic dissection, suggests an abnormality of the structure of the arterial wall. Connective tissue abnormalities have been observed in ADPKD. Extracellular matrix components such as collagen type IV, proteoglycan, fibronectin, undulin, and tenascin have all been demonstrated to be abnormal in pattern of deposition and proportion [11–13].

Aortic dissection is a rare but potentially fatal complication of the disease, and clinicians should maintain a high index of suspicion when dealing with patients with ADPKD who present with chest pain or collapse. Further characterization of the polycystin gene may provide a better understanding of the nature of the vessel wall abnormality.

Table 1. Characteristics of reported cases of ADPKD and thoracic aortic dissection

<table>
<thead>
<tr>
<th>Age at dissection</th>
<th>Sex</th>
<th>ESRF</th>
<th>Histology</th>
<th>Habitus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biagini et al. [7]</td>
<td>63</td>
<td>F</td>
<td>No</td>
<td>–</td>
</tr>
<tr>
<td></td>
<td>45</td>
<td>F</td>
<td>No</td>
<td>–</td>
</tr>
<tr>
<td>Hartman [5]</td>
<td>72</td>
<td>M</td>
<td>Yes</td>
<td>–</td>
</tr>
<tr>
<td>Somlo et al. [6]</td>
<td>60</td>
<td>F</td>
<td>No</td>
<td>–</td>
</tr>
<tr>
<td></td>
<td>38</td>
<td>M</td>
<td>Yes</td>
<td>–</td>
</tr>
<tr>
<td></td>
<td>31</td>
<td>M</td>
<td>No</td>
<td>Cystic medial necrosis</td>
</tr>
<tr>
<td>This case</td>
<td>36</td>
<td>F</td>
<td>Yes</td>
<td>Normal</td>
</tr>
</tbody>
</table>

ESRF, end-stage renal failure; M, male; F, female.
Thoracic aortic dissection complicating ADPKD

References


Received for publication: 17.2.97
Accepted: 26.3.97