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Case Report

Sacral radicular cysts in autosomal dominant polycystic kidney disease

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Abstract

This is the first report of a case of sacral radicular cysts in a patient with autosomal dominant polycystic kidney disease (ADPKD). A 46-year-old woman with ADPKD was found to have bilateral sacral radicular cysts discovered incidentally by magnetic resonance imaging (MRI). Cysts arising from arachnoid or spinal meningeal sac should be considered one of the manifestations of a more widespread connective tissue disorder associated with ADPKD.

Keywords: ADPKD; arachnoid cyst; magnetic resonance imaging; sacral radicular cyst

Introduction

Patients with autosomal dominant polycystic kidney disease (ADPKD) may have intracranial aneurysm, hypertensive intracerebral haemorrhage or cervicocephalic arterial dissection. Although arachnoid cysts have been associated with ADPKD [1–5] with a prevalence rate of 8% (10 times higher than in the general population), spinal meningeal cysts have been reported in only four patients [6,7]. The association of sacral radicular cysts with ADPKD has never been previously reported. We report a case of bilateral sacral radicular cysts in a patient with ADPKD discovered incidentally by magnetic resonance imaging (MRI). In addition to reporting this case, we review the literature.

Case

A 46-year-old woman was referred in October 2008 for a routine follow-up for ADPKD diagnosed 8 years before. Mild arterial hypertension was known for ∼8 years. Two months earlier, she presented an episode of macroscopic haematuria that was controlled by conservative treatment. The patient remained asymptomatic. Physical examination at that time was normal except for slight arterial hypertension. She was treated with telmisartan 80 mg daily. Neurological examination was normal. Her serum creatinine was 2 mg/dl (176.8 μmol/l) and creatinine clearance 34 ml/min/1.73 m^2 (0.56 ml/s/1.73 m^2). Abdominal ultrasonography disclosed enlarged cystic kidneys with a few liver cysts. MRI of the abdomen revealed both kidneys with numerous cysts of different sizes scattered throughout the parenchyma. Some cysts showed evidence of recent bleeding. The total kidney volume measured by MRI was 3053 ml (right kidney 1530 ml and left kidney 1523 ml). Moreover, radicular cysts at the sacral level were an incidental finding (Figure 1). The sacral radicular cysts had a fluid content with homogenous high signal intensity on T_2-weighted sequences similar to the signal of cerebrospinal fluid (CSF).

Discussion

Most of the cases of meningeal cysts occur in patient with neurofibromatosis, Marfan syndrome, Ehlers–Danlos syndrome and Lehman syndrome [8–10]. A primary defect in the organization of collagen, with the decrease in its tensile strength, leads to dural weakness to such an extent that it becomes ectatic. These abnormalities, also called arachnoid cysts, meningoceles or meningeal diverticula, are abnormal outpouching of the common dural sac, the spinal arachnoids or the nerve root sheath. These cysts may have thin walls and contain CSF, so they may sometimes be detected only by the mass effect they exert. MRI is the diagnostic procedure of choice because of its ability to demonstrate the exact location, extent and relationship of the arachnoid cyst with the adjacent brain or spinal cord. Myelography and CT myelography remain of diagnostic value, especially for cases that are not definitive on MRI. Most spinal meningeal cysts are asymptomatic and are discovered incidentally by MRI. Clinical features result from nerve root or spinal cord compression and vary with the location of the cyst. Lumbar cysts may present with low back pain and radiculopathy. Patients may present clinically with progressive spastic or flaccid para- or tetraparesis. Sacral cyst may be the aetiology of chronic perineal pain in many instances. Sensory deficits are less prominent. Bowel or bladder dysfunction may also occur in individuals with sacral cysts [11,12].

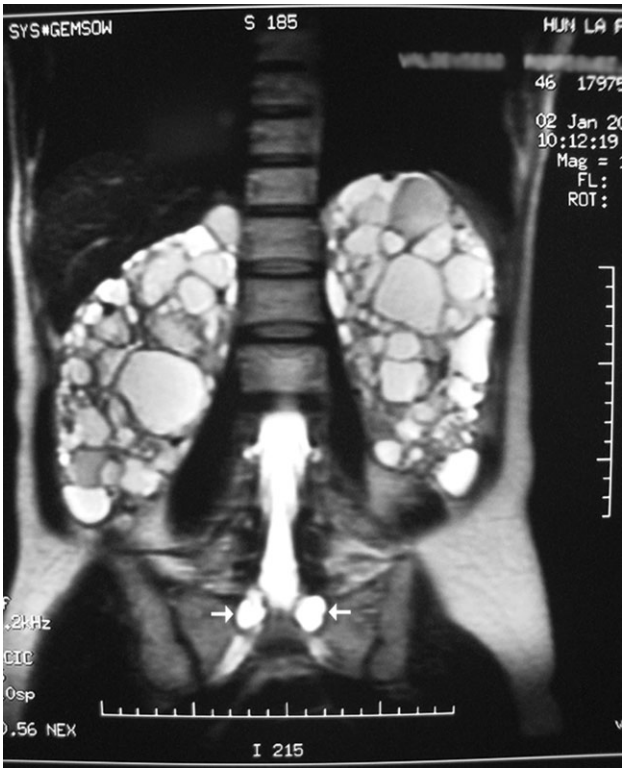

Fig. 1. Coronal T_2-weighted MRI of spine showing radicular cysts (arrows) at the sacral level. The high signal content was similar to the signal of CSF. Also note numerous cysts in both kidneys.

Although arachnoid cyst has been associated with ADPKD [1–5], spinal meningeal cyst has been rarely reported. Thus, spinal meningeal cyst has only been described in four adult patients with ADPKD [6,7]. The cysts were found at the thoracic level in three of them and the lumbar level in the fourth one. Although the aetiology of spinal meningeal cysts is unknown, it is likely that an underlying weakness of the meninges is involved. Some of these patients had a history of postural headaches caused by spontaneous intracranial hypotension [6,7]. However, the association of sacral radicular cysts with ADPKD has never been previously reported. In our patient, the sacral radicular cysts were an incidental finding that remains so far totally asymptomatic. Our patient exhibited no features of neurofibromatosis, Marfan syndrome, Ehlers–Danlos syndrome or Lehman syndrome. In this case, MRI provided sufficiently clear anatomical information and demonstration of the pathological entity.

In summary, we believe that the association between spinal meningeal cysts and ADPKD is not fortuitous. Cysts arising from arachnoid or spinal meningeal sac should be considered one of the manifestations of a more widespread connective tissue disorder associated with ADPKD. Our patient was asymptomatic. However, in other published cases meningeal cysts were complicated by postural headaches, cranial nerve palsies, visual blurring, upper or lower limb pain, weakness or numbness. Such signs and symptoms occurring without a clear explanation in a patient with ADPKD should lead the physician to perform an MRI of the spinal chord.

Conflict of interest statement. None declared.

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