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DOI: [10.1159/000350727](https://doi.org/10.1159/000350727)**Ruptured Cerebral Aneurysm and Acute Bilateral Carotid Artery Dissection in a Patient with Polycystic Kidney Disease and Polycystic Liver Disease**Christian Roth^{a,b}, Jens Kleffmann^b, Carsten Bergmann^c,
Wolfgang Deinsberger^b, Andreas Ferbert^aDepartments of ^aNeurology and ^bNeurosurgery, Klinikum Kassel, Kassel, and ^cCenter for Human Genetics, Bioscientia, Ingelheim, Germany*Background*

Both autosomal dominant polycystic kidney disease (ADPKD) and polycystic liver disease (PCLD) follow an autosomal dominant pattern of inheritance, and cerebral vascular manifestations have been described as occasional part of the disease spectrum [1].

Case Report

A 39-year-old woman complained about sudden headache with vomiting while watching a soccer match. She developed mild weakness on her left side. Upon admission, she still had a headache and was awake, but displayed no neurological deficits, e.g. Horner syndrome or meningism. Further clinical examination showed that her

weight was normal but that the size of her abdomen was massively enlarged. A CT scan showed a small hyperintensity in the area of the right Sylvian fissure. Subarachnoidal hemorrhage was suggested and digital subtraction angiography showed three aneurysms of the middle cerebral artery on the right side. Moreover, a dissection of the right internal carotid artery was suspected due to a wedge-shaped high-grade stenosis at the base of the skull. After interdisciplinary discussion, a decision was made to clip the aneurysm. Intraoperative blood clots were found perianeurysmal. Postoperatively, the patient again showed a mild hemiparesis on the left side. Over the following days she developed an organic psychosis. Five days after admission, MRI presented a middle cerebral artery infarction on the right side. Furthermore, fat-suppressed axial T1-weighted images showed a semilunar-shaped hematoma in the arterial wall of both internal carotid arteries with a high-grade stenosis on the right side and only a low-grade stenosis on the left side (fig. 1). A CT scan of the abdomen revealed a massively enlarged polycystic liver reaching down to the pelvis minor, but with only a few cysts of the kidneys (fig. 1). Family history yielded several relatives with polycystic kidney and/or liver disease on both the paternal and maternal sides. Our patient was the only one to have suffered from cerebral vascular disease. Five weeks after admission, the patient could be transferred to a rehabilitation clinic with only a mild cognitive impairment. Liver transplant was performed 20 months after the initial event. Molecular analysis of the patient revealed both a homozygote mutation in the *PKD1* gene (ADPKD) as well as in the *ADPLD* gene (PCLD); we have described this previously [2].

Fig. 1. **a** MRI with fat-suppressed axial T1-weighted images shows semilunar-shaped hematoma in the arterial wall of the both internal carotid arteries (circle). **b** A CT scan of the abdomen shows a massively enlarged polycystic liver reaching down to the pelvis minor.

Discussion

This is the first report of ruptured aneurysm of the middle cerebral artery in combination with acute bilateral artery dissection. Liver cysts and cysts of the subarachnoid space, in particular, are described as extrarenal manifestations of ADPKD. The prevalence of intracerebral aneurysms in patients with ADPKD is 8–10% and therefore more common than in the general population [1, 3, 4]. However, dissections of the cerebral arteries in patients with ADPKD have rarely been described in the literature [5–7]. We suggest that in the case in our study the extended degree of cerebrovascular involvement was due to the mutation load with a homozygote mutation in this dominant disease. Furthermore, cerebral aneurysms are suggested to be part of the spectrum of PCLD [8]. The combination of two life-threatening cerebral vascular diseases is extremely rare. In patients with multiple liver and/or kidney cysts and a history of headache, subarachnoidal hemorrhage as well as carotid dissection must be considered.

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