NEPHROLOGY PICTURES



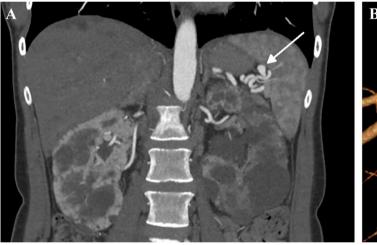
Aneurysms of splenic artery in a patient with autosomal dominant polycystic kidney disease

Julia Borowiecka¹ · Zofia Jankowska¹ · Monika Gradzik² · Mariusz Niemczyk¹ o

Received: 10 January 2024 / Accepted: 24 March 2024 © The Author(s) 2024

A 37-year-old woman with autosomal dominant polycystic kidney disease (ADPKD) and normal kidney function, with well-controlled arterial hypertension, and rupture of an intracranial aneurysm at 27 years of age presented to our institute. Ultrasound led to a suspicion of two splenic artery

aneurysms, which was subsequently confirmed by computed tomography (Fig. 1). The patient was referred to a vascular surgeon who recommended observation alone. To date, the aneurysms remain clinically silent.



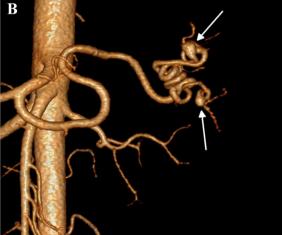


Fig. 1 A Contrast-enhanced computed tomography of the abdominal cavity, coronal plane. Aneurysm of the splenic artery (arrow).

Polycystic kidneys. **B** Contrast-enhanced computed tomography (3D reconstruction). Two aneurysms of the spenic artery (arrows)

Published online: 08 May 2024



Mariusz Niemczyk mariusz.niemczyk@wum.edu.pl

Department of Transplantology, Immunology, Nephrology, and Internal Medicine, Medical University of Warsaw, Warsaw, Poland

Department of Clinical Radiology, Medical University of Warsaw, Warsaw, Poland

ADPKD is the most common monogenic disease of the kidney. It affects approximately 1 in every 1000 people. As well as being a cause of chronic kidney disease, it is accompanied by numerous extra-renal manifestations, including aneurysms. Due to possible dramatic consequences of rupture, intracranial aneurysms, observed in approximately 10% of ADPKD patients [1], attract the most attention. However, aneurysms can also be found in other arteries [2].

Knowledge of the pathogenesis of aneurysms in ADPKD remains piecemeal. ADPKD is caused by a mutation in the PKD1 or PKD2 gene, leading to disturbed structure and function of their protein products, polycystin-1 (PC-1), or polycystin-2 (PC-2), respectively. Mutations in PKD1 or PKD2 lead to dysregulation of various intracellular signaling pathways. Both PKD1 and PKD2 are expressed in the smooth muscle and myofibroblasts of blood vessels, and polycystins play an important role in maintaining the normal structure and function of blood vessels. Additionally, involvement of vascular enothelial growth factor (VEGF), collagen genes and genes of the TGF-β pathway in vascular remodeling was shown; however, exact mechanisms were not elucidated. These changes, together with arterial hypertension, which is common in ADPKD, may result in aneurysm development [S1, S2—see Suppl. file].

Rupture of an intracranial aneurysm in a patient's medical history is considered a risk factor for the development of new intracranial aneurysms in ADPKD [1]. Our case reminds us that new aneurysms should also be expected in other locations.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s40620-024-01946-3.

Author contributions JB and ZJ, literature search, writing the manuscript; MG, imaging; MN, management of the patient, idea of the paper, writing the manuscript.

Funding No funding was received to support this work.

Data availability Additional data are available from the corresponding author on justifiable request.

Declarations

Conflict of interest None.

Ethical approval The manuscript was prepared in accordance with the Declaration of Helsinki. Approval from the institutional board was not required for this case report.

Informed consent The patient agreed to publication of her data and images in an anonymous form.

Open Access This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit http://creativecommons.org/licenses/by/4.0/.

References

- Chapman AB, Devuyst O, Eckardt KU et al (2015) Autosomaldominant polycystic kidney disease (ADPKD): executive summary from a kidney disease: improving global outcomes (KDIGO) controversies conference. Kidney Int 88:17–27
- Ruderman I, Menahem S (2014) Generalized aneurysmal disease in association with autosomal dominant polycystic disease. Clin Kidney J 7:416–417

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

