MAEDICA – a Journal of Clinical Medicine
2023; 18(1): 157-160
https://doi.org/10.26574/maedica.2023.18.4.157

MÆDICA - a Journal of Clinical Medicine

CASE REPORT

Polycystic Kidney Disease and Polycystic Liver Disease Associated to Advanced Gastric Cancer: an External Complication of Potter III Disease

Danilo COCO[a], Silvana LEANZA[b]

[a]Department of General Surgery, Ospedali Riuniti Marche Nord, Pesaro, Italy

[b]Department of General Surgery, Carlo Urbani Hospital, Jesi, Ancona, Italy

ABSTRACT

Autosomal dominant polycystic kidney disease (ADPKD) is the most frequent of the hereditary diseases affecting the kidney but it can also be associated with cysts in the pancreas, liver, arachnoid, seminal vesicles. In particular, ADPKD plus adult polycystic liver disease (APLD) is defined as Potter type III. In the literature, the association of malignant neoplasm with APLD and ADPKD is extremely rare. We have found only nine cases. Only one report described multiple gastric carcinomas associated with Potter type III cystic disease. We described a case of a man suffering from ADPKD and APLD plus a stage IV (TNM) gastric adenocarcinoma.

Keywords: autosomal dominant polycystic kidney disease (ADPKD), adult polycystic liver disease (APLD), Potter III disease, gastric carcinoma.

INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is the most frequent of the hereditary diseases affecting the kidney, but it can also be associated with cysts in the pancreas, liver, arachnoid, seminal vesicles. It is caused by defects and malfunctions in the PKD-1 or PKD-2 genes. This results in a non-functioning of polycystin proteins, which is an integral membrane protein that functions as a regulator of calcium permeable cation channels and intracellular calcium homeostasis. It seems that polycystin-1 is encoded by the PKD-1 gene, which can be considered in its own right as a tumor suppressor gene that not only induces apoptosis and cell

Address for correspondence:
Danilo Coco, MD
Ospedali Riuniti Marche Nord, Pesaro, Italy
Email: webcostruction@msn.com

Article received on the 23rd of September 2022 and accepted for publication on the 10th of March 2023

cycle arrest in the G0/G1 phase in cancer cells but also prevents cancer cells from migration. So, PKD genes inhibit cancer progression. In addition, mutations in PRKSCH and SEC63 genes have been found to cause adult polycystic liver disease (APLD). Defective PKD-1 and PKD-2 may be a carcinogenic condition. In 1964, the studies of Osathanondh and Potter (1) classified ADPKD plus APLD as Potter type III. In the literature, we have found only nine cases. Just one report described multiple gastric carcinomas associated with Potter type III cystic disease.

In the present work, we described a case of a man affected of ADPKD plus APLD plus a stage IV (TNM) gastric adenocarcinoma. ❑

CASE REPORT

A 70-year-old man was admitted to our hospital for asthenia, easy fatigue and dyspnea. Past medical history included essential hypertension, diabetes mellitus and polycystic kidney disease as well as polycystic liver disease, stage IV kidney disease, BPH, cerebral vasculopathy. He stated that none of his family had suffered from polycystic kidney disease. Active physical examination admission revealed distended abdomen associated with open alvus in the absence of signs of peritonism. Laboratory examination revealed hemoglobinemia of 7.5 g/dL. An abdominal computed tomography (CT) scan showed multiple cysts in the liver and cystic lesions on both sides in the kidney and diffuse thickening of the body and gastric antrum. For this reason, he was subjected to esophagus-gastro-duodenoscopy, which highlighted a vegeto-ulcerated neoformation of the small gastric curvature involving the antrum and the angulus in its entirety, with part of the fundus reaching less than 1 cm below the cardia and proximal antrum. Histology revealed an infiltrating adenocarcinoma. Staging was completed with abdominal magnetic resonance imaging (MRI), fluorodeoxyglucose PET (PET-FDG) and tumor marker assay. The MRI scan of the abdomen showed a concentric parietal thickening of the body and gastric antrum with focal infiltration of the serosa and encroachment of the peritoneal adipose tissue anterior to the body and absent cleavage plane of the small curvature at the hepatic margin S2, perigastric lymph nodes, and confirmed the occurrence of polycystic kidney

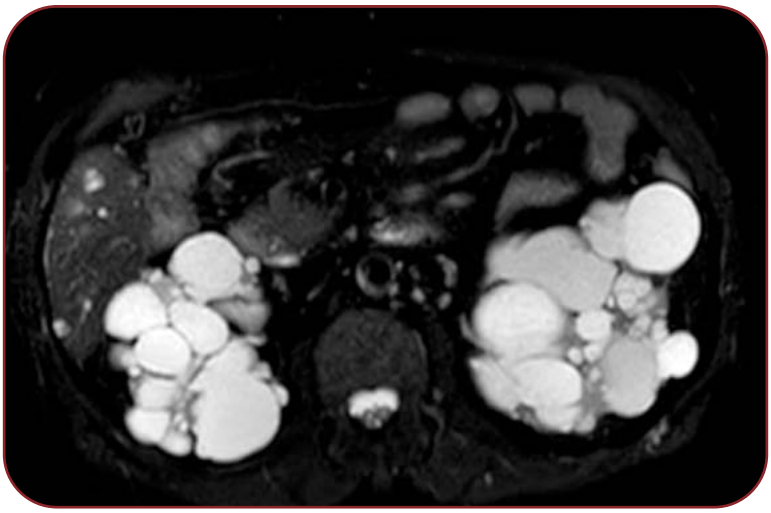

FIGURE 1. Autosomal dominant polycystic kidney disease (ADPKD)

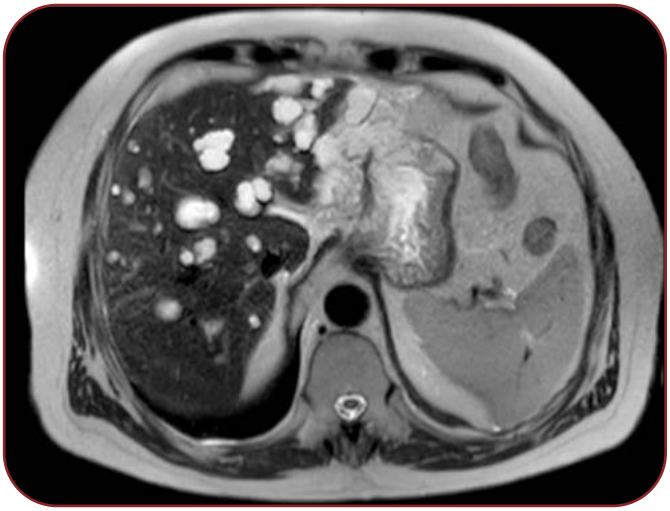

FIGURE 2. Adult polycystic liver disease (APLD)

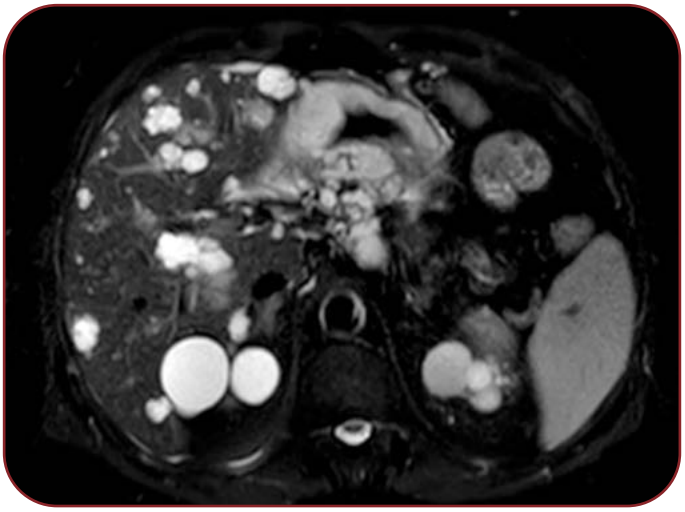

FIGURE 3. Stage IV (TNM) gastric adenocarcinoma of the antrum and angulus of the stomach

disease and polycystic liver disease associated with advanced gastric cancer (Figures 1, 2, 3). PET FDG demonstrated the presence of focal hepatic hyperaccumulation at the level of S7 (SUV 5.2), suggestive of probable metastasis. CEA was 125 ng/mL, Ca19.9:142 U/mL. After discussion between multidisciplinary teams, six cycles of neoadjuvant chemotherapy (CHT) according to the OLFi scheme, reduced to 50% of the dose for stage IV CRF, and subsequent surgical re-evaluation were indicated. Re-evaluation CT scan revealed a thickening of the small gastric curvature with extension to the peritoneal adipose tissue and perigastric and mesenteric lymphadenopathy. Post-CHT PET showed an accumulation at the level of antrum and duodenum (SUV 5.9) not significant accumulation of tracer at seg VII and lymph nodes. The MRI scan of the abdomen showed that the heteroplastic tissue of the antral body reached the serosa with associated lymphadenopathy of the small curve and hepatic hilum was unchanged. After chemotherapy and subsequent restadiation, the patient underwent surgical exploration. An exploratory laparoscopy was performed, which demonstrated the presence of peritoneal carcinosis with the peritoneal carcinomatosis index (PCI) of 10. Conclusive diagnosis was gastric adenocarcinoma cT3N+M1, HER2 negative, stable MSS, DPYD absent polymorphisms. At the time of discharge, the patient was apiretic, in good general condition, and received a personalized diet. ❑

DISCUSSION

Autosomal dominant polycystic kidney disease (ADPKD) is the most frequent of the hereditary diseases affecting the kidney, but it can also be associated with cysts in the pancreas, liver, arachnoid, seminal vesicles (2). In 1964, the studies of Osathanondh and Potter (1) classified the polycystic kidneys into three types. In particular, ADPKD defined as Potter type III involves one or more of several anomalies. The confirmation of this polycystic and multi-district pathology was highlighted by the studies on 399 patients carried out by Hatfield and Pfister (3), who noted that 58/399 of their subjects were affected by multiple cysts in both kidneys, liver and/or pancreas cysts; among study participants, a family history of cystic diseases was found in 60% of cases. However, as in our case, there may be no family history. No family history occurs in 20-40% of cases. Although not confirmed by genetic research for the PKD-1 or PKD-2, family history criteria, clinical findings, and imaging studies can confirm ADPKD. The likelihood of having Potter's disease in the same patient type III, namely APLD and ADPKD, and an associated neoplasm is shown to be very rare. In the literature we have found only nine cases, including five pancreatic neoplasms, three cholangiocarcinoma and one patient with Potter III disease associated with multiple gastric carcinomas. In 1977, a study by Cryer *et al* (4) discussed a case of a 66-year-old female with family history (FH) of ADPKD, who was suffering from pancreatic adenocarcinoma. In 1984, Landais *et al* (5) described two cases (a 52-year-old female without ADPKD-FH and a 56-year-old male with APDKD-FH) with inoperable chorangiocarcinoma. In 1997, Niv *et al* (6) reported a case of advanced pancreatic cystadenocarcinoma in a 68-year-old man ADPKD-FH. In 2001, Sakurai *et al* (7) described a case of Potter III disease associated with inoperable pancreatic ductal adenocarcinoma in a 63-year-old man. In 2002, Sasaki *et al* (8) reported an intrahepatic cholangiocarcinoma in a 60-year man with ADPKD. In 2005, Naitou *et al* (9) found a 43-year-old man with an intraductal papillary mucinous tumor fit for surgery associated with ADPKD-FH. He had cysts in kidney, liver and pancreas. In 2009, Sato *et al* (10) performed a pancreasectomy for intraductal papillary mucinous tumor in a 63-year-old man with kidney, liver and pancreas cystis without ADPKD-FH. In 2011, Mimatsu *et al* (11) performed a gastrectomy for multiple stomach tumors in a 65-year-old woman with multiple cysts in the kidney, liver and mesentery without ADPKD-FH. We felt that this case of the association of APLD with multiple renal cysts could be classified as Potter's type III cystic disease without ADPKD-FH. ❑

CONCLUSION

We described the case of a man suffering from ADPKD and APLD plus a stage IV (TNM) gastric adenocarcinoma, which was found as a complication in patients with polycystic disease, although it was unclear whether the development of inoperable gastric cancer was

directly or indirectly caused by ADPKD plus APLD, known as Potter III disease. ❑

Conflicts of interest: none declared.
Financial support: none declared.

References

1. **Osathanondh V, Potter EL.** Pathogenesis of polycystic kidneys: historical survey. *Arch Pathol* 1964;77:459-512.
2. **Torres VE, Harris PC, Prison Y.** Autosomal dominant polycystic kidney disease. *Lancet* 2007;369:1287-1301.
3. **Hatfield PM, Pfister RC.** Adult polycystic disease of the kidneys (Potter type 3). *JAMA* 1972;222:1527-1531.
4. **Cryer PE, Kissane JM.** Obstructive jaundice in a patient with polycystic disease. *Am J Med* 1977;62:616-626.
5. **Landais P, Grunfeld JP, Droz D, et al.** Cholangiocellular carcinoma in polycystic kidney and liver disease. *Arch Intern Med* 1984;144:2274-2276.
6. **Niv Y, Turani C, Kahan E, Fraser GM.** Association between pancreatic cystadenocarcinoma, malignant liver cysts, and polycystic disease of the kidney. *Gastroenterology* 1997;112:2104-2107.
7. **Sakurai Y, Shoji M, Matsubara T, et al.** Pancreatic ductal adenocarcinoma associated with Potter type III cystic disease. *J Gastroenterol* 2001;36:422-428.
8. **Sasaki M, Katayanagi K, Watanabe K, et al.** Intrahepatic cholangiocarcinoma arising in autosomal dominant polycystic kidney disease. *Virchows Arch* 2002;441:98-100.
9. **Naitou H, Shoji H, Ishikawa I, et al.** Intraductal papillary mucinous tumor of the pancreas associated with autosomal dominant polycystic disease. *J Gastrointest Surg* 2005;9:843-845.
10. **Sato Y, Mukai M, Sasaki M, et al.** Intraductal papillary-mucinous neoplasm of the pancreas associated with polycystic liver and kidney disease. *Pathol Int* 2009;59:201-204.
11. **Mimatsu K, Oida T, Kawasaki A, et al.** Multiple Gastric Carcinomas Associated with Potter Type III Cystic Disease. *Case Rep Gastroenterol* 2011;5:590-596. doi: 10.1159/000329179.