

ACHALASIA AND BILATERAL SYNCHRONOUS RENAL ADENOCARCINOMAS: A CASE REPORT

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INTRODUCTION

When treating dysphagia and suspected achalasia, it is important to determine whether it is primary achalasia with loss of ganglion cells in the myenteric plexus of the distal esophagus or pseudoachalasia, which is most often due to tumor infiltrations of the lower esophageal sphincter (Zaninotto et al. 2018, Oude Nijhuis et al. 2020). In this paper, a unique case is reported regarding bilateral synchronous renal adenocarcinomas, initially presented with progressive dysphagia and significant unintended weight loss, in fact, with achalasia.

CASE REPORT

A 39-year-old male patient with dysphagia was initially referred from a provincial hospital to the National Reference Center for Functional Gastrointestinal Disorders in Zagreb, Croatia. During the last 8 months, he reported difficult and painful swallowing; at first he managed to swallow solid food with liquid but with time the symptoms progressively worsened. On admission to the Reference Center, he also had difficulty swallowing fluid and stated that food was getting stuck behind the sternum. He complained of chest and epigastric pain along with frequent vomiting. Since suffering these complications, he has involuntarily lost 20 kilograms of body weight. He has no history of serious diseases and no family history of digestive system tumors. The patient had a decreased body mass index (Body Mass Index – BMI 19.6 kg/m²), he was at nutritional risk (Nutrition Risk Screening - 2002 was 3) and during clinical examination there was accentuated pain present regarding epigastral palpation. His appetite was weakened but stool and urination were without major problems. He denied allergies; he is a long-term smoker, 20 cigarettes/day, but has reduced to 5 cigarettes/day since the beginning of symptoms. In therapy he used proton pump inhibitors (PPIs) - pantoprazole 40 mg daily.

An initial laboratory diagnosis was made (complete blood count, C-reactive protein, erythrocyte sedimen-

tation rate, urine, liver enzymes, serum urea and creatinine, blood glucose, serum protein electrophoresis, serum iron, ferritin, thyroid-stimulating hormones, tumor markers, antibodies to celiac diseases, etc.) and all the findings were within reference ranges. The esophagram with barium showed dilatation of the esophagus (grade II) with barium retention in the lumen and the narrowing of the distal part in the form of a "bird's beak" (Figure 1A).

An upper endoscopy (esophagogastroduodenoscopy - EGD) showed a dilated esophagus with food retention in the lumen, without peristalsis with narrowing in the distal part that passes with some resistance while the EGJ area appeared unchanged (Figures 1B and 1C). Esophageal biopsies were taken for PHD; no malignancy was found. High-resolution esophageal manometry (HREM) findings were characteristic for achalasia type II: panesophageal pressurization; a LES resting pressure of 38 mmHg and an integrated relaxation pressure (IRP) of 18 mmHg (Figure 1D).

The Eckardt symptom score was 11 points (stage III). An ultrasound examination of the abdomen revealed tumors on both kidneys. In the frame of further analysis, a multislice computed tomography (MSCT) of the thorax showed no focal or other changes in the lung parenchyma; no lymphadenopathy was found but a wider esophageal lumen was observed. Abdominal and pelvic MSCTs with digital subtraction angiography (DSA) showed an expansive process of 60 mm × 50 mm in the distal third of the left kidney bounded within the renal capsule, inhomogeneously imbibed by contrast, with ponds of contrast, suggesting a tumor (Figure 1E). Also, these examinations showed that the right kidney was ectopically located, in the pelvic position, in the small pelvis, which in its middle part showed a nodal expansive process with a diameter of 40 mm × 30 mm that was post-contrast imbibed (Figure 1F). No liquid or other pathological contents were found in the bottom of the pelvis, nor significant abdominal lymphadenopathy. The cytological examination of urine did not reveal the presence of malignant cells.

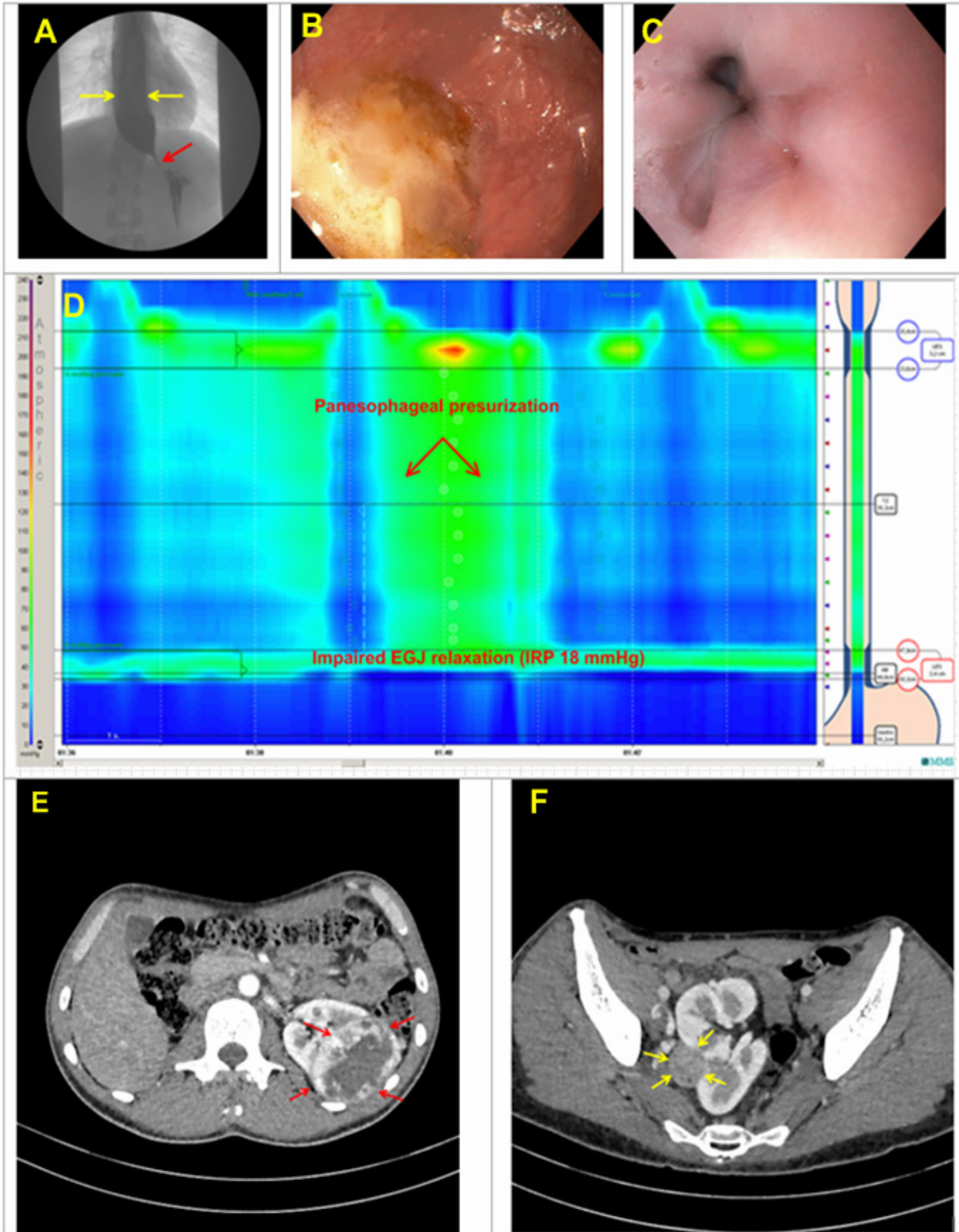


Figure 1. Diagnostic algorithm: (A) Barium esophagram - dilatation of esophagus (yellow arrows), "bird's beak" (red arrow); (B) Esophagogastroduodenoscopy (EGD) - dilated esophagus with food retention in lumen; (C) EGD - narrowing of distal part of esophagus and EGJ; (D) High-resolution esophageal manometry; (E) Abdominal MSCT - tumor in distal third of left kidney (red arrows); (F) Abdominal MSCT - ectopically located right kidney and tumor in its middle part (yellow arrows)

The patient was presented to a multidisciplinary team that included a gastroenterologist, urologist, oncologist, radiologist, and pathologist. Due to dominant symptoms (progressive dysphagia), an endoscopic pneumatic balloon dilatation of the LES was performed first. The procedure was performed under general anesthesia under the supervision of an anesthesiologist. The LES was dilated according to a standardized protocol with a balloon catheter under 10-12 Psi pressure. The procedure went smoothly; a laceration on the cardia was found; post-procedurally without any complications. Shortly afterwards, the patient was hospitalized at the Urology Clinic and a radical left nephrectomy was performed. The histological findings according to the WHO classification corresponded to an adenocarcinoma of the kidney, a clear cell histological type, with nuclear grade 3 (PHD: Adenocarcinoma renis. pT1bNXMX. G3).

After pneumatic dilatation (PD) was performed, the patient was able with proper chewing and pureed diet, to eat food properly and gain weight. After recovery from the left nephrectomy, surgery on the right kidney tumor was scheduled. However, recurrent dysphagic problems (difficulty swallowing food but fluid passed properly) occurred before the scheduled surgery. A control EGD was performed where no more food retention was observed in the lumen of the esophagus. The distal part of the esophagus was passed with some less resistance. For these reasons, 3 months after the previous PD, another PD of the LES was performed according to standardized protocol. Dilatation as well as post-procedural monitoring was conducted without complications. Shortly afterwards, the patient was hospitalized at the Urology Clinic, but at his request, no radical nephrectomy of the right kidney was performed, only an enucleation of the tumor. The histological finding corresponded with chromophobic renal cell carcinoma (PHD: Adenocarcinoma renis).

The patient has check-ups with a urologist every 6 months with blood tests and pelvic kidney Doppler; all findings have been normal thus far. Clinically, significant improvement has been achieved (regular food ingestion) and significant body weight gains (BMI 21.3 kg/m²). In therapy, he took pantoprazole 40 mg daily and oral enteral supplements. During control EGDs, there was no retention of contents in the esophageal lumen and the LES was passed without significant resistance. Moreover, repeated biopsies were taken from the distal part of the esophagus; histologically without malignancy. Additionally, MSCTs of the thorax and abdomen were repeated; the findings were without the presence of tumors or metastases. During follow-up, a positron emission tomography - computed tomography (PET-CT) was performed with regular ultrasound and endoscopic examinations since the last operation on the right kidney was more than 14 months ago. According to the findings, there were no signs of recurrence

of the renal tumor process or metastasis. At the last manometric findings, the resting LES pressure was not elevated but the aperistaltic activity of the esophageal body was still present. The Eckardt symptom score was 2 points (stage I) and the achalasia therapy was considered as successfully performed. Certainly, the further monitoring of the patient is necessary, both in relation to the symptoms of achalasia and the monitoring of the pelvic right kidney and renal function.

CONCLUSION

Generally speaking, pseudoachalasia in patients with RCC is rare (Bhalme et al. 2009, Cabezas-Camarero et al. 2015, Lamm et al. 2018, Padda & Si 2019, Schizas et al. 2020). According to the available literature, this is the first case report of a patient with bilateral synchronous renal cell carcinoma clinically manifested by dysphagia and progressive weight loss, and in whom the manometric findings indicated achalasia. In our patient, the therapy of achalasia has proven to be an effective one, enabling the further treatment of the malignant disease; improving symptoms and nutritional status.

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Contribution of individual authors:

Rosana Troskot Perić: conceptualization, literature survey, diagnosis and follow-up of the patient, writing manuscript, supervision.

Danijel Bevanda: writing manuscript, literature survey, diagnosis and follow-up of the patient, conceptualization, methodology.

Sandra Zgodić & Barbara Paušak: data acquisition, literature survey, proof reading.

Marina Madunić & Dubravka Jandrić: project administration, supervision.

All authors approved the final version.

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