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Letter to the Editor

# Metastatic Intrahepatic Cholangiocarcinoma Presenting as an Achalasia-like Syndrome

## Case

The authors present a case of a 53-year-old woman with cutaneous lupus erythematosus without systemic involvement and no current medication, who was evaluated at our department due to a 4-month history of significant weight loss (approximately 25%) and an initially selective dysphagia for solids, that progressively became non-selective for liquids and solids. The patient denied recent endemic trips, ingestion of caustics, or exposure to radiation. Esophagogastroduodenoscopy revealed a slight dilatation of the esophageal lumen with food stasis, a regular and easily traversed gastroesophageal junction (GEJ), and absence of gastric and duodenal lesions. Esophageal biopsies excluded eosinophilic esophagitis. High-resolution manometry revealed 100% of failed contractions (10% with esophageal pressurization) with an increased integrated relaxation pressure (33.9 mmHg), characteristic of a type I achalasia (Chicago classification version 3.0) (Fig. 1). Pneumatic dilation at 30 mm and 35 mm (Rigiflex, Boston Scientific, Boston, Massachusetts) was performed, without symptomatic improvement. CT detected a solid mass with central hypodensity and peripheral uptake with 4.4 × 3.0 cm in segment II, without clear margins within GEJ structures suggesting infiltration (Fig. 2). In segments V and IV, 2 other hypodense lesions with 2.2 cm and 2.0 cm were identified, suggestive of secondary nature. Histology of the lesion in segment II was obtained through a percutaneous biopsy, showing the presence of an adenocarcinoma with diffuse expression of Cytokeratin (CK)-19 and CK-20, multifocal of Caudal type homeobox-2, and focal of CK-7 (without expression of Thyroid transcription factor-1, aspects favoring a primary neoplasia of the gastrointestinal tract, without microsatellite instability expression. Colonoscopy did not reveal an invasive colorectal neoplasia and subsequent CT for staging presented a dimensional increase in the main lesion and mesenteric enlarged lymph nodes. Positron emission tomography study (F-18 fluorodeoxyglucose) corroborated pericardial, peritoneal, ganglionic, and hepatic metastasis. In a multidisciplinary team discussion, intrahepatic cholangiocarcinoma with distant metastasis was assumed, conditioning invasion at the GEJ level with a consequent pseudoachalasia. The patient was proposed for palliative chemotherapy with cisplatin/gemcitabine and enteral feeding.

## Discussion

Pseudoachalasia is an achalasia-pattern like syndrome presenting with dilatation of the esophagus due to the narrowing of the distal esophagus from causes other than primary denervation.[1] It is a rare entity, and could be difficult to differentiate from achalasia even in the manometric findings (aperistalsis of esophageal body and incomplete relaxation of lower esophageal sphincter) and, in a recent systematic review, 23.6% of patients were primarily treated for a mistakenly diagnosed achalasia.[2] Generally, symptoms with less than 6 month-duration, age older than 50 years old and a marked weigh loss were considered to be clinical features that help distinguish pseudoachalasia from achalasia.[2,3] According to a recent European guideline,[4] it is suggested an additional testing using CT or endoscopic ultrasound should be performed in patients with 2 or more of the following risk factors: duration of symptoms of less than 1 year, age over 55 years old, weigh loss over 10 kg, and severe difficulty passing the lower esophageal sphincter with an endoscope. In our case, regardless of patient's age, the significant weight loss and short symptom duration was relevant to prompt a CT. The most common cause of pseudoachalasia is adenocarcinoma of GEJ

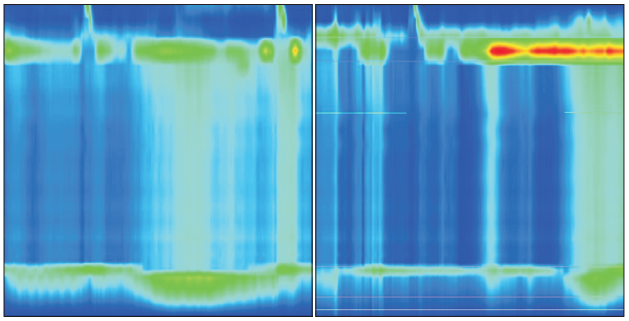

**Figure 1.** High-resolution manometry: 100% of failed contractions (10% with esophageal pressurization) with an increased integrated relaxation pressure (33.9 mmHg), characteristic of a type I achalasia (Chicago classification version 3.0).

(70%) followed by other lymphoproliferative disorders, metastatic carcinomas and small cell lung carcinomas.[2] To our knowledge, this is the second case in the literature reporting an intrahepatic cholangiocarcinoma with GEJ infiltration conditioning a pseudoachalasia syndrome.[2]

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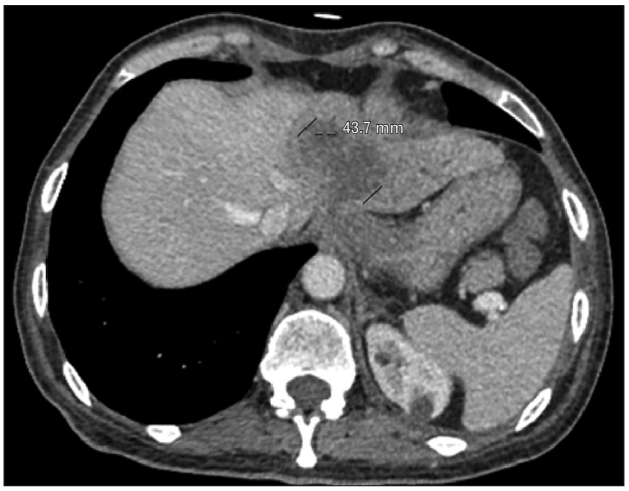


**Figure 2.** Findings on CT. Solid hepatic mass with 4.4 × 3.0 cm in segment II infiltrating gastroesophageal junction structures.

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