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A rare case of pancreatic cancer presenting as pseudoachalasia

"Pseudo" (or secondary) achalasia is a rare entity that it isn't easily distinguishing from idiopathic achalasia by manometry, radiological examination and endoscopy. Usually a neoplastic process of the esophago-gastric region is associated with this clinical condition. However, it has been reported that other neoplastic processes may lead to the development of pseudoachalasia, such as mediastinal masses, gastrointestinal tumours (pancreas, liver, biliary tract and other organs) and non gastrointestinal malignancies. We present a case of pseudoachalasia in which the primary cause of the disease was not an esophago-gastric cancer.

KEY WORDS: Achalasia, Manometry, Pseudoachalasia.

Introduction

"Pseudo" (or secondary) achalasia is a rare entity that it isn't easily distinguishing from idiopathic achalasia by manometry, radiological examination and endoscopy [1-3,10]. Usually a neoplastic process of the esophago-gastric region is associated with this clinical condition: benign masses as stromal tumours, amyloidosis or oesophageal leiomyomatosis [5,6,13,15,16,34,39] and malignant masses as esophago-gastric cancers [14,17-26,38]. However, it has been reported that other neoplastic processes may lead to the development of pseudoachalasia, such as mediastinal masses [29], gastrointestinal tumours (pancreas, liver, biliary tract and other organs) [4,35,36,47,48] and non gastrointestinal malignancies [12,27,28,30-33,37,40-42]. Mechanisms proposed to explain the clinical features of pseudoachalasia include a circumferential obstruction of the distal oesophagus or a malignant infiltration and destruction of inhibitory neurons within the myenteric plexus of the oesophageal wall [8,9]. Rarely, a distant neoplasm may cause this syndrome as a paraneoplastic process [11]. We present a case of pseudoachalasia in which the primary cause of the disease was not an esophago-gastric cancer.

Case report

A 71-years-old man presented with disphagia for solids and liquids, lasting for 6 months, regurgitation of indigested food with a weight loss of 15 kg over the same period of time. Laboratory tests were normal. The clinical past history reported an ischemic cardiopathy and chronic obstructive pulmonary disease. Upper endoscopy revealed a difficult transit through the cardial region with

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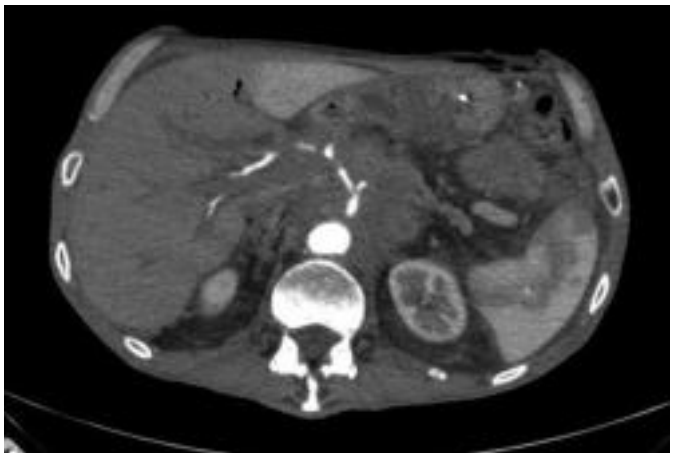

Fig. 1: Pancreatic cancer involving hepatic and spleen arteries.

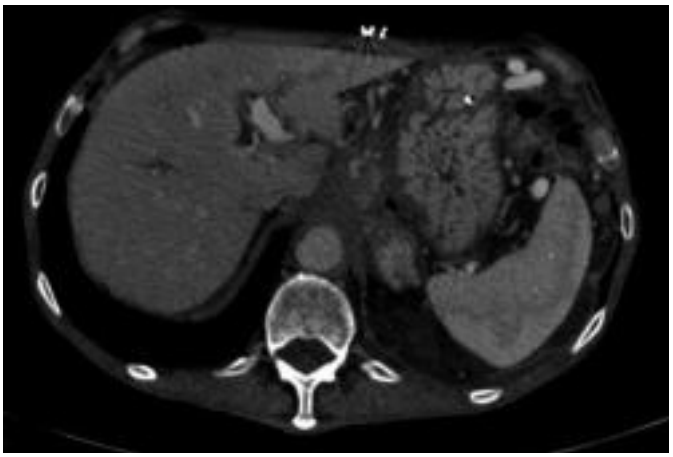

Fig. 2: Pancreatic cancer infiltrating esophago-gastric region.

no mucosal lesions at the gastroesophageal junction. A barium swallow examination revealed a mildly dilated oesophagus with irregular oesophageal motility. On oesophageal manometry there was an incomplete post-swallowing relaxation of the lower oesophageal sphincter (LES) and a complete absence of primary peristalsis of oesophageal body. The patient was submitted to pneumatic dilatation with a mild improvement of symptoms. A strict follow-up was planned. He came back to our department 1 month later for complete arrest of bolus transit, regurgitation, weight loss of additional 5 kg and a posterior para-vertebral pain. A CT scan of the abdomen was performed and revealed a large mass arising in the pancreatic body with extension to the cardial region. Staging laparoscopy confirmed a large irresectable tumour of the pancreatic body extending to the gastroesophageal junction with intramural infiltration. Peritoneal cytology was positive for well differentiated pancreatic adenocarcinoma and biopsy of tumoral tissue confirmed the diagnosis. A jejunostomy for enteric nutrition was performed. A course of chemotherapy was started but the patient died a few weeks later.

Discussion

The first two cases of secondary, malignancy-induced, achalasia, the so-called "pseudo"- achalasia, were described by Horwath early in the past century [43]. The prevalence of pseudoachalasia is considered to be in the 2-4% range among patients with manometric findings suggestive of achalasia [2,3,24]. Primary gastroesophageal cancer is considered the most common malignancy causing pseudoachalasia, but also liver, lung, pancreas, breast cancer, haematological cancers and metastases from unknown primaries have been reported in association with achalasia [14,17-26,27-42]. Benign causes of pseudoachalasia are extremely rare [50]. Three cases of pseudoachalasia in patients with pancreatic cancer have been described until now [4,47,48]. We have reported another case of pancreatic adenocarcinoma with pseudoachalasia.

The difficulty of differentiating achalasia from pseudoachalasia is well known [1-4,7-10,44,45,49]. Older age, short clinical history and significant weight loss in a short period of time are usually considered suggestive of pseudoachalasia. However, as reported in scientific literature, the positive predictive value of these parameters is low, because of similar clinical and radiological presentation [45]. The presence of a dilated esophagus on barium swallow examination and a normal-appearing cardial region on endoscopy are common findings in both primary and secondary achalasia and they are usually of little help in differentiating the two conditions. The most sensitive test to diagnose primary achalasia is oesophageal manometry, typically with incomplete LES relaxation and complete aperistalsis of the oesophageal body. The presence of peristalsis in some swallows should itself raise the suspicion of pseudoachalasia, but in some cases as ours, the complete aperistalsis of the esophageal body should exclude it. Endoscopic ultrasound (EUS) and CT scan are radiological examinations that we perform in the evaluation of the tissue surrounding the oesophageal wall in esophagogastric tumours [46,49]. However, in the assessment of achalasia they are not routinely performed [51,52]. Sometimes it is very difficult to distinguish primary achalasia from pseudoachalasia with routinely examinations and, in doubtful cases, it is mandatory to perform a CT scan in order to exclude the second clinical condition [7]. In conclusion, although rare, pseudoachalasia is a clinical condition to consider in the differential diagnosis of achalasia in patients presenting with disphagia keeping in mind that tumours other than oesophageal or gastric may be responsible for this condition.

Riassunto

La pseudoacalasia, o achalasia secondaria, è un'entità rara non facilmente distinguibile dall'acalasia idiopatica sia con la manometria, che con gli esami radiologici e le indagini endoscopiche.
Generalmente un processo neoplastico che si sviluppi in corrispondenza della regione esofago-gastrica si associa ad una condizione clinica paragonabile all'acalasia dello sfintere esofageo inferiore. Sono inoltre stati descritti altri processi neoplastici che possono determinare una paseudoacalasia, come masse mediatiniche, tumori gastrointestinali (pancreas, fegato, vie biliari extraepatiche e di altri organi) ed altre condizioni gastrointestinali non neoplastiche.
Si presenta qui il caso di una pseudoacalasia in cui la causa primitiva non era rappresentata da una neoplasia esofago-gastrica.

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