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Gastritis cystica profunda mimicking a GIST – A diagnostic challenge

Gastritis cystica profunda simulando GIST – un caso de desafío diagnóstico

Gastritis cystica profunda (GCP) is a rare hyperplastic lesion with unclear pathogenesis histologically characterized by the presence of gastric glands in the submucosa and even muscularis propria of the stomach with normal overlying mucosa.[1,2] There are two histological patterns of gastritis cystica poliposa: gastritis cystica superficialis, in which cystic glands are limited to the mucosal layer; and GCP in which the cystic lesion locates within the submucosa and muscularis propria.[3] Clinical manifestations of GCP are variable and can include gastrointestinal bleeding, epigastric pain and weight loss.[1] An unspecified mucosal insult or injury is widely accepted as etiological mechanism but the pathophysiology is unknown.[1–4] We present a case of acute gastrointestinal bleeding caused by gastritis cystica profunda mimicking a gastrointestinal stromal tumor in a patient without previous gastric surgery.

A 67 year-old man was admitted for melena during 4 days associated with syncope. He had a previous history of atrial fibrillation and was medicated with warfarin. At admission, he was hemodynamically stable and had no abdominal pain. Laboratory tests revealed normocytic anemia with hemoglobin 8.8 g/dl (previous value: 13.5 g/dl) and an INR of 2.3. Upper gastrointestinal endoscopy (UGIE) revealed a 40 mm polypoid lesion in the gastric body with normal mucosa surface and a central 15 mm ulcerated bleeding as which was very suggestive of a gastrointestinal stromal tumor (GIST) (Fig. 1). There were no esophageal or duodenal visible bleeding lesions. Biopsies performed during UGIE revealed aspects of chronic active gastritis and *Helicobacter pylori* was identified. Upper endoscopic ultrasonography (EUS) showed 40 mm submucosal a hypoechogenic and heterogeneous mass with cystic areas and no perilesional adenopathies. The EUS findings could also correspond to a GIST. EUS guided fine-needle aspiration (FNA) using 19 gauge needle was preformed but the sample was insufficient for evaluation. A full body computed tomography was performed and revealed a 4 × 6 cm gastric intraluminal lesion without signs of invasion or metastatic disease. The diagnosis of a gastric GIST was assumed and the patient was proposed to surgical resection of the lesion. Macroscopically, a polypoid lesion with a nodular surface and a central ulcer was observed, with multiple cysts and solid areas on cross-section (Fig. 2). Histologically (Fig. 2), the gastric mucosa showed focal lesions of chronic atrophic gastritis with activity, hemorrhage and ulceration, the submucosa and muscular propria displayed an abundant cystically dilated pyloric-type and foveolar-type glandular proliferation, without mitoses or atypia; surrounding the glands there was a thin layer of lamina propria and fibromuscular hyperplasia. The diagnosis of Gastritis cystica profunda was made. Surgical margins were free of lesion.

In the majority of reported cases, GCP occurs in patients with a history of gastric surgery, in particular Billroth II procedure.[2,3] It is unclear if it is secondary to chronic inflammation as consequence of duodenal reflux, foreign body reaction or ischemic injury as a result of the surgery.[1,2,5,6] Nevertheless, the interruption of the muscularis mucosae appears to allow migration of epithelial cells into the submucosal layer and subsequent cystic dilation.[2,7] In the unoperated stomach, the cause may be congenital in origin in patients with no prior gastric ulceration or trauma history.[3] It is more common in men and most frequently develops in the gastric body,[3] as seen in this case. Our patient had a history of chronic atrophic gastritis with *H. pylori* infection which in this case can be considered as a possible etiological factor. The presentation symptoms are not specific and, endoscopically it is impossible

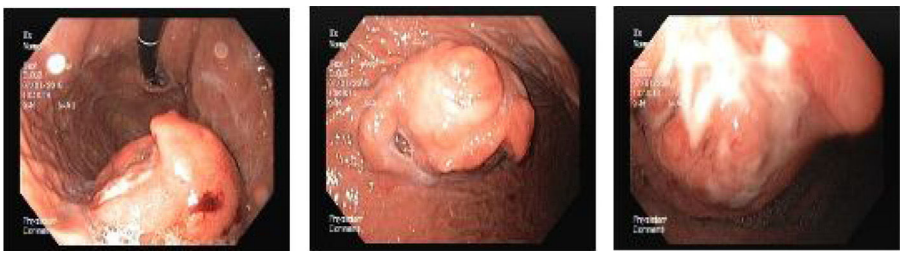

Figure 1 Upper gastrointestinal endoscopy showing a 40 mm polypoid lesion in the gastric body with normal mucosa surface and a central 15 mm ulcerated bleeding suggestive of a gastrointestinal stromal tumor.

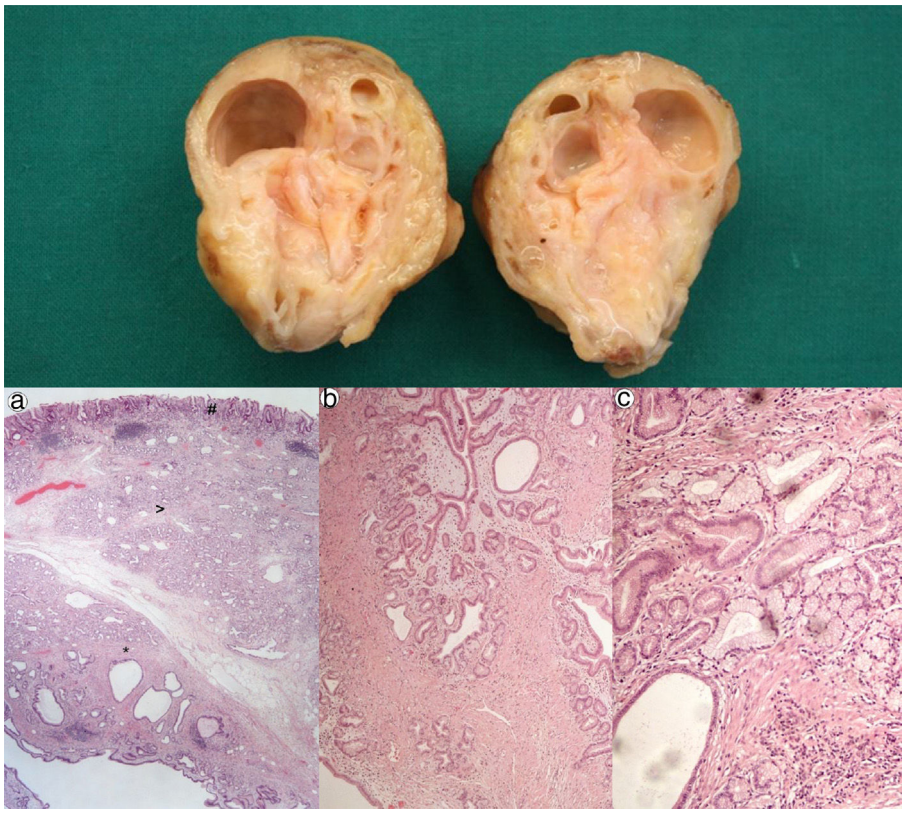

Figure 2 Cross-section shows multiple cysts with diameters between 0.2 and 1.5 cm, filled with transparent and mucinous-looking liquid. (a) (H&E ×20): gastric wall including mucosa (#), submucosa (>) and muscularis propria (*) with abundant cystic glandular proliferation; (b) (H&E ×40): surrounding the glands there is a thin layer of lamina propria and fibromuscular hyperplasia; (c) (H&E ×100): the glands are lined by pyloric-type and foveolar-type epithelium, without mitoses or atypia.

to differentiate from other entities like polyps or GIST.[2] As biopsy samples are restricted to the spared mucosal the results are often not diagnostic.[3,8] CT can show a heterogeneously iso- to hypoattenuating intraluminal lesion with multiple small cysts but the appearance may be similar to GIST.[3,9] On EUS, GCP can appear as a polymorphic, homogeneous cystic mass with a minimal solid component within the gastric mucosae which is also not specific of GCP.[1,3] However, the accuracy of EUS-FNA with immunostaining in preoperative GIST, diagnosis has been reported at 91%–100%.[10] Some patients might have to undergo gastric resection when it is impossible to make a definite diagnosis with radiologic study or endoscopic biopsy, as seen in this case. Case reports have revealed the possibility of a malignant transformation from GCP, even in an unoperated stomach, but the incidence of malignancy in GCP patients remains unknown.[3] Epstein–Barr virus might have a role as a premalignant factor in cancer tissue with GCP.[10] Given the lack of a pathognomonic endoscopic or radiographic appearance of GCP, diagnostic and surveillance guidelines are not available and further studies are required.

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References

1. Machicado J, Shroff J, Quesada A, Jelinek K, Spinn MP, Scott LD, et al. Gastritis cystica profunda: endoscopic ultrasound findings and review of the literature. Endosc Ultrasound. 2014;3:131–4.
2. Laratta JL, Buhtoiarova TN, Sparber LS, Chamberlain RS. Gastritis cystica profunda: a rare gastric tumor masquerading as a malignancy. Surgical Sci. 2012;3:158–64.
3. Park CH, Park JM, Jung CK, Kim DB, Kang SH, Lee SW, et al. Early gastric cancer associated with gastritis cystica polyposa in the unoperated stomach treated by endoscopic submucosal dissection. Gastrointest Endosc. 2009;69:47–50.
4. Park JS, Myung SJ, Jung HY, Yang SK, Hong WS, Kim JH, et al. Endoscopic treatment of gastritis cystica polyposa found in an unoperated stomach. Gastrointest Endosc. 2001;54:101–3.
5. Moon SK, Kim KO, Park SH, Yoo KS, Park CH, Kim JH, et al. Gastritis cystica profunda accompanied by multiple early gastric cancers. Korean J Gastroenterol. 2010;55:325–30.
6. Tsuji T, Iwahashi M, Nakamori M, Ueda K, Ishida K, Naka T, et al. Multiple early gastric cancer with gastritis cystica profunda showing various histological types. Hepatogastroenterology. 2008;55:1150–2.

7. Kalra VB, Gilbert JW, Mitchell KA, Salem RR, Israel GM. AIRP best cases in radiologic–pathologic correlation: gastritis cystica polyposa. Radiographics. 2013;33:109–14.
8. Tomizuka T, Mazaki T, Mado K, Henmi A, Ishii Y, Masuda H, et al. A case of gastritis cystica profunda. Surgery. 2008;143:449–50.
9. Akahoshi K, Oya M. Gastrointestinal stromal tumor of the stomach: how to manage? World J Gastrointestinal Endosc. 2010;2:271–7.
10. Choi MG, Jeong JY, Kim KM, Bae JM, Noh JH, Sohn TS, et al. Clinical significance of gastritis cystica profunda and its association with Epstein–Barr virus in gastric cancer. Cancer. 2012;118:5227–33.

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Hidroadenocarcinoma ecrino perianal en el contexto de una enfermedad de Crohn fistulizante

Perianal eccrine hydroadenocarcinoma in the context of a fistulising Crohn's disease

El adenocarcinoma del canal anal es una neoplasia muy poco frecuente que representa el 5% de todas las anorrectales y un 1,5% de los tumores gastrointestinales[1,2]. De acuerdo con la Organización Mundial de la Salud, se pueden distinguir 3 tipos: el primero tiene su origen en la mucosa de transición del canal superior, el segundo deriva de las glándulas (ductos) anales y el último, de una fístula perianal crónica. Los pacientes con tumores del canal anal, independientemente de su tipo, presentan mayor porcentaje de enfermedad avanzada, metástasis a distancia y menor supervivencia global en comparación con los casos de carcinoma escamoso rectal. La escasa casuística publicada implica que no exista un esquema terapéutico plenamente comprobado[3,4], aunque la mayoría de los autores abogan por un tratamiento con quimiorradioterapia neoadyuvante seguido de cirugía radical[5]. Describimos a continuación un caso extremadamente raro de hidroadenocarcinoma ecrino perianal en un paciente a quien se diagnosticó previamente enfermedad de Crohn.

Varón de 42 años con enfermedad de Crohn de 20 años de evolución con múltiples fistulizaciones perianales crónicas, intervenido en 2006 con colostomía derivativa. El paciente fue derivado a cirugía para biopsia de adenopatía inguinal dolorosa y biopsia «trucut» perianal para descartar la degeneración de una fístula perianal crónica. En el estudio anatomopatológico, el ganglio linfático mostró una arquitectura totalmente reemplazada por una proliferación celular atípica de estirpe epitelial organizada en forma de nidos sólidos, adoptando en otras áreas un patrón ductal con morfología cribiforme focal. La celularidad neoplásica era de gran tamaño, con núcleos pleomórficos de cromatina granular, nucléolos evidentes y abundantes mitosis. El citoplasma variaba (pálido eosinófilo, granular o con cambio hacia célula clara). El inmunofenotipo de la célula era positivo para p53 (80%), CK8, CK19, antígeno epitelial de membrana (EMA), antígeno carcinoembrionario (CEA), CKAE1/AE3 y CAM 5.2 con inmunoexpresión negativa de 34BE12, calretinina, CD30, CD117, vimentina, a-inhibina, CK7, p504S, p63, PSA, CDX2, CK20, CD10 y S100[6]. El EMA, CK8, CK19 y el CEA suelen estar presentes en las glándulas ecrinas y apocrinas. La morfología y el inmunofenotipo descritos fueron coincidentes con los hallazgos de la biopsia «trucut», estableciéndose el diagnóstico de metástasis ganglionar de hidroadenocarcinoma derivado de glándulas ecrinas perianales.

Se realizó un estudio de extensión mediante TC toracoabdominal, RM pélvica y rastreo corporal con citrato de galio-67 (fig. 1). El estudio con galio-67 puso de manifiesto un depósito patológico de actividad en la región perianal compatible con un proceso inflamatorio-séptico.

En la TC se observaron múltiples adenopatías en las cadenas ilíaca interna y externa derechas e inguinales bilaterales. Se detectaron hallazgos de enfermedad perianal compleja; múltiples trayectos fistulosos y abscesificaciones. Adicionalmente, en la RM se identificó una gran masa heterogénea de aspecto infiltrativo que interesaba desde la porción más caudal del recto, musculatura del suelo de la pelvis, glúteo mayor derecho y hasta tejido graso subcutáneo y piel.

Con el diagnóstico de una afectación neoplásica infiltrativa del canal anal (T4) con diseminación adenopática, se discutió el caso en el Comité Multidisciplinar y, revisada la bibliografía, se decidió tratar mediante quimioterapia neoadyuvante[7]. Se administraron 12 ciclos con esquema FOLFOX (ácido folínico, fluorouracilo y oxiplatino) y 8 ciclos más sin oxiplatino (con 5-fluorouracilo).

Se reevaluó al paciente mediante RM abdominopélvica que confirmó progresión tumoral asociando fístulas complejas y abscesificación perilesional y se identificaron adenopatías en progresión a nivel inguinal e ilíacas externas. En el estudio de extensión (TC) no se constataron signos de enfermedad diseminada.

Se planificó la intervención quirúrgica y se procedió a una resección local, extirpándose una neoplasia perianal ulce-