

Endoscopic Management of a Rare Entity: Gastric Calcifying Fibrous Tumor

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Keywords

Calcifying fibrous tumor · Submucosal tumor · Stomach · Endoscopic mucosal resection

Tratamento endoscópico de uma entidade rara: tumor fibroso calcificante do estômago

Palavras Chave

Tumor fibrose calcificante · Lesão subepitelial · Estômago · Resseção endoscópica da mucosa

A 55-year-old woman with no relevant past medical history was referred to our department for reevaluation of a gastric lesion detected on a esophagogastrosocopy. The lesion was described as a bulge at the gastric body, measuring 10 mm, with overlying normal appearing mucosa (Fig. 1). Endoscopic ultrasound (EUS) revealed a 9 × 12 mm hypoechoic, homogeneous lesion mainly localized within the submucosa, with well-defined and regular limits. Taking into account the reduced lesion size, ultrasonographic characteristics and patient preference, an en-bloc cap-assisted endoscopic submucosal resection was

performed, with no complications (Fig. 2, Fig. 3). Histologically, a complete (R0) resection was observed with a submucosal sharply delineated hypocellular spindle cell proliferation, haphazardly arranged, set in an abundant collagenous hyalinized stroma. Peripheral lymphoid cuffs, patchy lymphoplasmacytic aggregates frequently associated with small vessels, sometimes with germinal centers and dispersed foci of dystrophic calcification were noticed (Fig. 4). Immunohistochemical staining showed positivity with vimentin and CD10. A patchy, granular positivity with CD68 was also obtained (Fig. 4). Tumor cells were negative for CD34, CD117, DOG-1, BCL2, desmin, SMA, HHF35, S100, AE1/AE3, Cam5.2, ALK-1, and beta-catenin. A diagnosis of gastric calcifying fibrous tumor (CFT) was made. Six months after endoscopic resection, the patient presented no signs of tumor recurrence.

CFTs are rare benign mesenchymal tumors with a predilection for the soft tissue and the subserosal abdominal cavity that occur mainly in children and young adults. CFTs of the gastrointestinal tract are quite rare and very few cases have been reported in the gastric wall. Gastric CFTs seem different from their soft tissue counterpart in their smaller tumor size, higher age at presentation, and no tendency for local recurrence. No sex predominance



Fig. 1. Endoscopic view of the lesion, appearing as a bulge at the gastric body, about 10 mm, with overlying normal appearing mucosa.

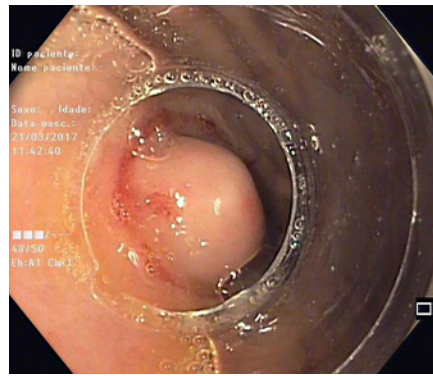


Fig. 2. En-bloc cap-assisted endoscopic submucosal resection.

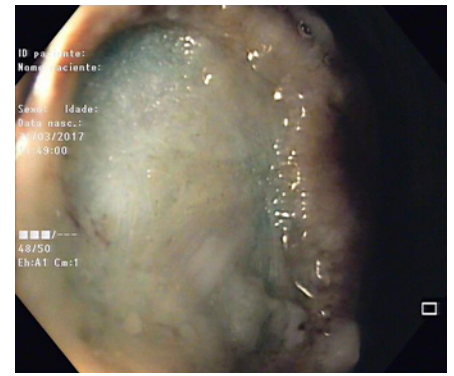


Fig. 3. Gastric scar after endoscopic resection of the lesion.

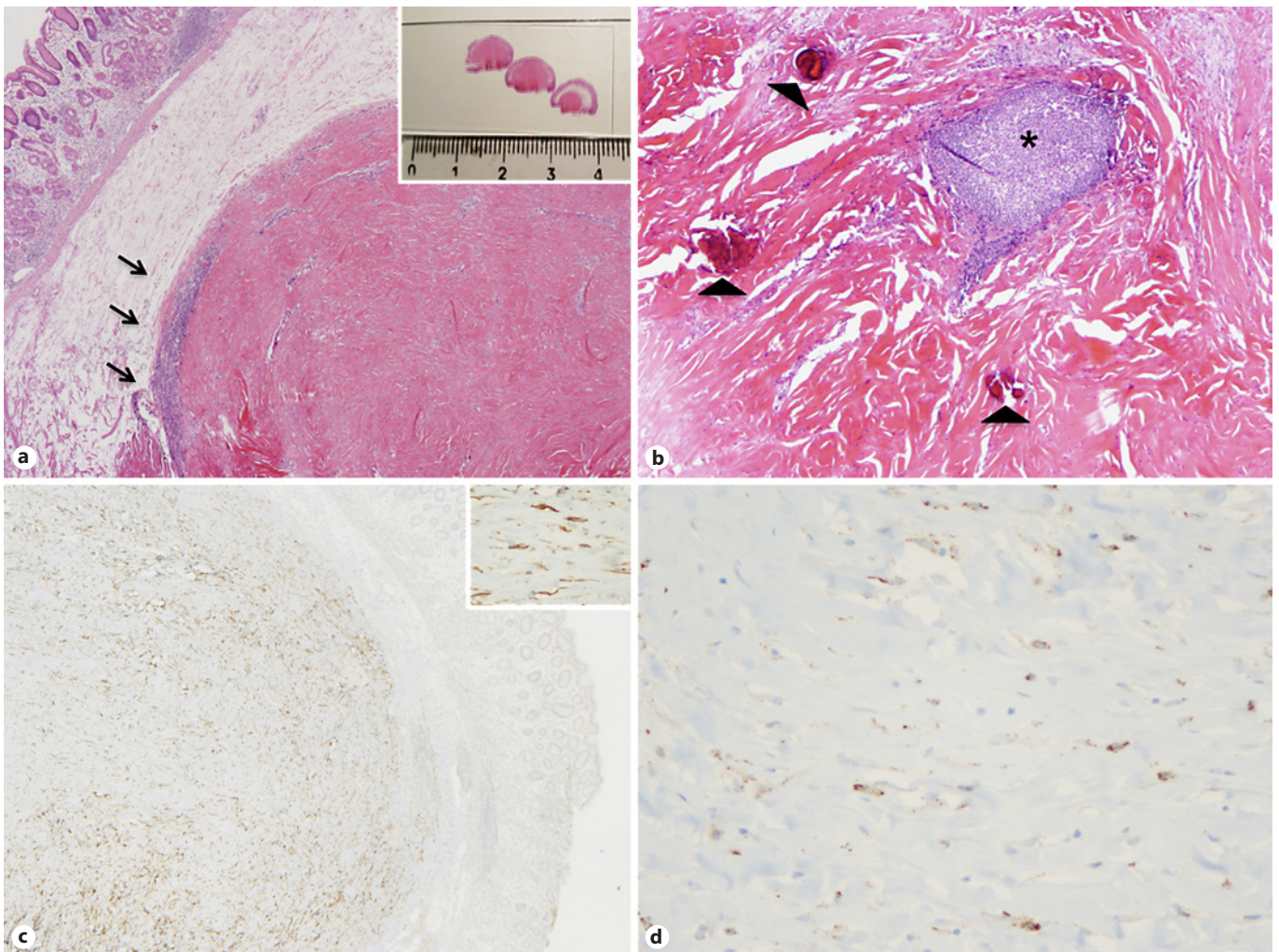


Fig. 4. a Well-circumscribed submucosal nodule, with focal lymphoid cuff (arrows). **b** Lymphoid aggregate with germinal center (asterisk) and foci of dystrophic calcification (arrowheads) in a hyalinized/sclerotic area. **c** Consistent universal positivity with vimentin (not shown) and with CD10 (inset with detailed cytoplasmic positivity).

d Weak and patchy granular cytoplasmic positivity with CD68 in the proliferative spindle cells as well as some inflammatory cells.

has been documented, age of presentation is usually in the fifth or sixth decade, and the majority are located in the gastric body [1–4].

Awareness of CFT and differential diagnosis with other mesenchymal tumors is important but not always straightforward. The main differential diagnosis is probably gastrointestinal stromal tumor (GIST), which is usually more cellular than CFT and lacks the hyaline stroma, the lymphoplasmacytic infiltrate, and lymphoid cuff. More importantly, GIST has typical expression of CD34, CD117, and/or DOG-1, which were negative in this case. Moreover, CD68 is negative in GIST. CD10 positivity was observed in our case and has not yet been reported in the literature since it has never been tested in any published series of CFT. Other tumors can enter the differential diagnosis such as schwannoma, hyalinized leiomyoma, reactive nodular fibrous pseudotumor, solitary fibrous tumor, inflammatory myofibroblastic tumor, and IgG4-related disease. Characteristic findings of these entities were lacking [1–2, 5].

With no specific symptoms or endoscopic or EUS findings, the diagnosis of CFTs relies largely on pathological evaluation. Given its excellent prognosis, differential diagnosis with other mesenchymal tumors is essential. In fact, malignant transformation has never been reported and rare recurrences occur primarily when resection is incomplete. Provided a complete resection can be ensured, endoscopy is a feasible and safe approach not only for diagnosis but also for management of this entity [1–4].

Disclosure Statement

The authors do not have any interest which might be interpreted as influential in this report. This report did not receive any support from corporations, industrial or private.

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