

What kind of a gastric tumor is this?

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ABSTRACT: This case report describes a gastric small submucosal tumor endosonographically resembling a gastrointestinal stromal tumor (GIST). The presence of calcifications inside prompted for the surgical excision, and the pathologic examination revealed in fact the presence of a glomangioma (glomus tumor). The presented case is an illustration of the fact that not all masses arising from the muscularis propria layer are GISTs, and the endosonographer must always think of alternative diagnoses.

KEYWORDS: gastric glomus tumor, gastric glomangioma, submucosal tumor

Introduction

Submucosal tumors of the gastric wall represent a relatively common indication for endoscopic ultrasonography (EUS). However, not all of the tumors located in the 4th submucosal layer are gastrointestinal stromal tumors (GISTs), and an alternative diagnosis must always be in the mind of the endosonographer.

Case Report

We would like to report the case of a 51 year-old male patient who was referred for upper gastrointestinal endoscopy complaining of epigastric discomfort, with the finding of a small submucosal tumor located on the gastric angulus. The endosonographic examination further characterized the small (13/9 mm) mass as isoechoic, originating from the 4th hypoechoic layer of the gastric wall, with small calcifications inside (Fig.1). The suspicion of a GIST with one of the characteristics for malignant behavior (echogenic foci) was raised.

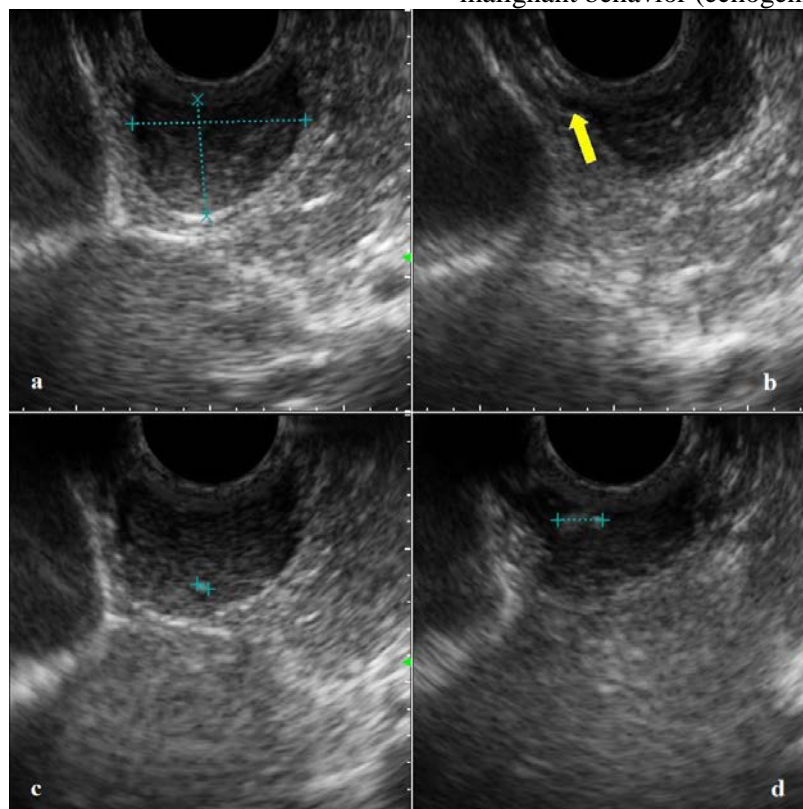


Fig.1. Endosonographic aspect of the small isoechoic mass of the gastric wall (a), originating from the 4th hypoechoic layer (arrow, b), with small calcifications inside (measured, c-d).

After surgical excision, which was chosen by the patient after being informed about the different possible management strategies, the pathologic examination revealed in the gastric serosa and muscularis propria a benign proliferation composed of large vascular channels separated by stroma including nests of round or cuboidal monomorphic cells with

plump nuclei and scant eosinophilic cytoplasm, immunohistochemically positive for smooth muscle actin and vimentin, focally for synaptophysin, and negative for S100 and cytokeratins (Fig.2). The histopathologic diagnosis was that of a glomus tumor (glomangioma).

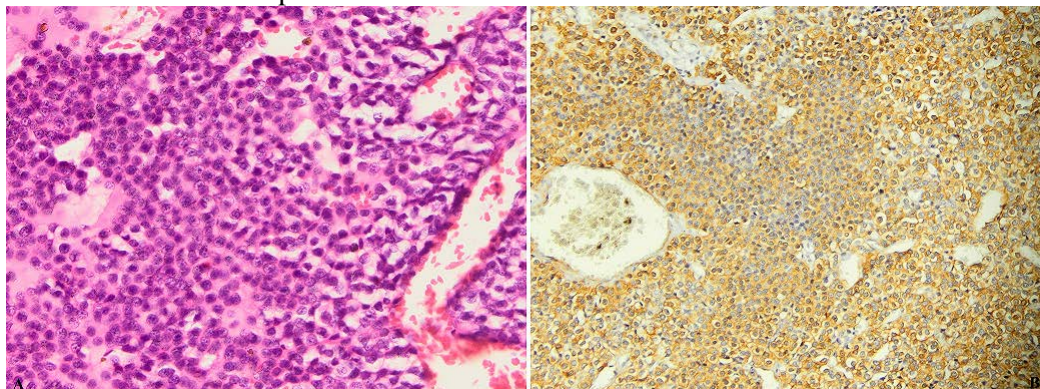


Fig.2. Histopathologic appearance of the excised submucosal tumor (H&A stain, A), immunohistochemically positive for smooth muscle actin (B).

Discussion

Gastric glomangiomas are rare, usually benign solitary neoplasms that arise from the smooth muscle cells of the glomus body, a type of neuromyoarterial receptor that plays a role in the regulation of arterial blood flow [1]. They appear as smooth submucosal masses with or without ulceration [2,3], and although EUS helps in identifying the layer of origin, there are no specific findings on gastroscopy or EUS allowing for a convincing preoperative diagnosis [4], most of the cases being in fact considered to be GISTs [5,6].

The most common location of glomangiomas is reported to be the gastric antrum [4] and their imaging features are variable, from sharply demarcated masses located in the 3rd or 4th layer of the gastric wall with hypoechoic pattern and heterogeneous echogenicity in most of the cases, sometimes with internal hyperechoic and/or echo-free structures which correspond to calcification and hemorrhage, respectively [2,4,7], to even the presence of a marginal halo in others [3], these imaging features being also encountered in GISTs of the gastric wall, a far more frequent disease entity [6]. Moreover, EUS features that are suggestive of malignancy in GISTs may be present in benign gastric glomus tumors [3,4]. These features include irregular borders, necrotic or cystic internal areas and the presence of tiny flecks of calcification [4], as in this case. The only more specific EUS feature

that could point to the diagnosis is the color Doppler examination, which might show a prominent vascular signal corresponding to the hypervascular nature of the tumor [8].

However, due to the intramural location, which precludes a diagnosis by standard endoscopic biopsy, and the lack of characteristic imaging features, glomus tumors are commonly diagnosed histologically and immunohistochemically only after surgical resection. It is important therefore for endoscopists not to be influenced by unusual findings of an otherwise “typical GIST” lesion, and thus to ignore the possibility of other diagnoses. Indeed, studies have shown that a definite diagnosis of GIST based solely on EUS features as described above is difficult to establish – specificity of 92%, but sensitivity of only 63% for GISTs was reported in a retrospective study [9]. Moreover, while EUS evaluation of the layer of origin of a submucosal tumor (SMT) is widely accepted as an important factor in providing a correct diagnosis of GIST, only around 80% of all SMTs are correctly characterized in this regard [10].

Considering these limitations, EUS guided fine needle aspiration (FNA) has been proven particularly useful in enhancing the diagnostic capabilities of the endoscopist by providing cytological material [11]. This also is not a perfect tool, the tissue yield of EUS-FNA for gastric SMTs being around 75%, with location in the lower third of the stomach and younger

age correlating with inadequate tissue yield [12]. However, EUS-guided FNA biopsy has been reported to yield adequate sampling for an accurate diagnosis of gastric glomus tumor [13]; it usually results in hemorrhagic material and although the cytomorphic features of glomus tumor are quite distinctive, immunohistochemical stains (e.g., alpha-SMA and vimentin positivity) can further help to ascertain the diagnosis [14].

Unfortunately, the cytological sample obtained by EUS-FNA does not provide sufficient cells to assess the mitotic index and, consequently, falls short of delivering data regarding the malignant potential of the tumor [15]. And as the criteria for malignancy in glomus tumors include besides location and size also a combination of high nuclear grade and mitotic activity (>5/50 HPFs) [16], there is definitely a need for core tissue sampling. In this regard, the introduction of EUS-guided trucut biopsy (TCB) can theoretically provide the examiner with sufficient material to allow a correct and complete diagnosis; however, this method is not commonly used in the diagnosis of SMT of the stomach wall and has not been studied in gastric glomangiomas.

In conclusion, EUS-guided fine needle aspiration (FNA) can be considered in such cases that might be mistaken for GISTs, maneuver reported to successfully diagnose glomus tumors (presence in the aspirate of benign vascular epithelioid cells) [5,6] and possibly sparing a patient from extensive surgical resection. However though, the assessment of the tumor's clinical behavior that cannot be fully reached by aspiration cytology and the risk of bleeding in these highly vascular tumors represent major limitations of EUS-FNA [4], and usually the diagnosis needs to be confirmed on the surgically resected specimen.

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