Gastric Glomus Tumor Presenting as a Submucosal Lesion of the Fundus: A Case Report and a Review of the Literature

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Introduction

Glomus tumors originate from smooth muscle cells implicated in the thermoregulation within the glomus body, an arteriovenous anastomosis located in the dermis [1]. Therefore, the majority of glomus tumors are found in fingernails, hands and feet [2]. Gastric glomus tumors are rare [2]; the antrum being the most frequent location in the stomach [3]. Those tumors are usually benign, but few cases of malignant gastric glomus tumors are described [1]. Neither radiologic nor endoscopic findings are typical of gastric glomus neoplasms [3]. Thus, the immunohistochemical study of a tumor sample is needed to make the diagnosis [3]. We report a case of gastric glomus tumor followed by a discussion about the presentation and the diagnosis challenge of this lesion.

Case Report

A 63-year-old woman known for metastatic breast cancer treated with chemotherapy had, during follow-up, a contrast-enhanced CT of the abdomen and pelvis (Somatom 16, Siemens AG, Germany) showed a 21 mm enhancing, partially calcified parietal lesion of the gastric fundus (Figure 1). The lesion was present on previous scanners but was not described. She never complained of any digestive symptoms. The lesion was described with an upper endoscopy as a 20 × 10 mm firm submucosal mobile nodule (Figure 2) and endoscopic biopsies showed normal fundic mucosa. A homogenously hypoechogenic lesion that originated from the muscularis propria with regular borders and without marginal halo (Figure 3 and Video 1) was visible on EUS. There was no peri-gastric lymph node. Fine needle aspiration (FNA) was performed with a 19 gauge needle.

The histologic findings showed aggregates of well-differentiated and well-delimited small round cells with dense chromatin and a moderately abundant cytoplasm within a fibrous stroma (Figure 4). The mitotic index was low. Immunohistochemistry showed tumor positivity for vimentin, smooth-muscle actin and synaptophysin and negativity for the keratin 7, keratin AE1/AE3, keratin 20, keratin 5/6, chromogranin, KIT (CD117), CD34, CDX2, CD56, protein-S100, p63 and desmin antigens (Figures 5 and 6).
Figure 4: 19G FNA sample of the gastric glomus tumor (H&E x100).

Figure 5: IHC showing tumor cells positivity for smooth-muscle actin antigen.

Figure 6: IHC showing tumor cells negativity for keratin antigen.

All those findings were consistent with a benign gastric glomus tumor and the patient was sent back to her treating physician for oncologic follow-up.

Discussion

Previously, gastric glomus tumors were thought to be more prevalent in women [1]. However, in recent reviews, men seem to be more affected than women [3-7]. The mean age of presentation is in the fifth decade [1,2,7]. Even if some gastric glomus tumors are found incidentally, the clinical presentation is symptomatic in most cases, consisting of ulcer-like symptoms or upper GI bleeding with or without anemia [1-7]. The most frequent localisation is the antrum [3], but it has been described in the gastric fundus in rare cases as reported in our case.

On CT, glomus tumors have been reported as well circumscribed homogeneous parietal lesions that may be partially calcified. Strong early enhancement and persistent later phases enhancement have been described [4]. Peripheral nodular enhancement in the arterial phase and homogeneous significant enhancement in the delayed phase has also been reported. On MRI, they manifest as slightly hyperintense on T1-weighted images, slightly hyperintense on T2-weighted images, are hypervascular and show persistent enhancement on later phases [5].

The majority of gastric glomus tumors are benign [2-7]. The probability of malignant behavior is low [1,3,7], but some cases of malignant and metastatic tumors exist [1]. The size of the tumor is an important malignancy predicting factor; especially for lesions larger than 5 cm [1,3,7]. Those lesions do not always have a malignant behavior, but they need a close follow-up [7]. Nuclear atypia and spindle cell change are other malignancy factors [1]. However, metastatic glomus tumors with only mild nuclear atypia have been described [1-3] showing that this is not a valuable malignancy predictor. To date, there are no clear criteria for malignancy in gastric glomus tumors [7]. Our patient had bone and liver metastasis but they were linked to a breast cancer relapse. Moreover, the lesion size was near 2 cm, there was no nuclear atypia and the cells were round, without spindle change reinforcing the benign nature of the gastric glomus tumor exposed here.

The approach of a glomus tumor is the same as other submucosal tumors of the stomach, as CT and EUS are both key modalities to describe the lesion [7]. However, a diagnosis of glomus tumor cannot be made with the imaging results [3]. Upper endoscopy is useful to recognize a submucosal tumor, but it usually does not provide enough submucosal material to make the diagnosis with standard biopsies [2-3]. Thus, the next step is EUS with guided FNA. It will help to evaluate the layer where the lesion is located, if the tumor shows any sign of malignity and a tumor sampling can be performed [3]. Our experience is to sample submucosal tumors with a 19G needle to obtain as much tissue as possible for immunohistochemistry and mitosis count. The diagnosis is made with the pathology results [3]. Gastric glomus tumors are positive for smooth-muscle cells and vimentin antigens in most cases [1]. They are also positive for synaptophysin antigen in a few cases [1-7]. However, they are negative for the KIT (CD117), associated with GISTs, and the chromogranin antigens, associated with carcinoid tumors [6].

Surgery is the treatment of choice for gastric glomus tumors [2-7]. The wedge or segmental resection is preferred in most cases, as glomus tumors are usually benign [2]. Subtotal gastrectomy is practiced mostly when there is a predicting factor for malignancy [2].

The diagnosis of gastric glomus tumors is difficult to make. Their rarity and the fact that they are often not recognized as a possible diagnosis for submucosal tumors of the stomach explain this difficulty. Indeed, textbooks and literature reviews about submucosal gastric tumors usually include GISTs, leiomyomas, metastasis, schwannomas, heterotopic pancreatic tissue and carcinoid tumors as possible lesions. However, glomus tumors are rarely mentioned [8]. We think that glomus tumors should be included in the differential diagnosis of a gastric submucosal lesion, especially if it is located in the antrum, with a reminder that glomus tumors can be found anywhere in the stomach.
In conclusion, glomus tumors are lesions arising from the arteriovenous anastomosis in the dermis [1]. Gastric localisation is rare but they should be included in the differential diagnosis of gastric submucosal tumors given their benign nature. The CT, MRI and EUS appearance is almost indistinguishable from other muscularis propria lesions like GIST or leiomyoma reinforcing the need of EUS-FNA for immunohistochemical studies [3].

Author’s Contributions

- Conception and design: E. Désilets, A. Comtois
- Drafting of the article: A. Comtois
- Critical revision of the article for important intellectual content and final approval: E. Désilets, T. Manière, C. Meunier, A. Comtois, XV. Do

The article has been reviewed by an English speaker as a second language.

Disclosure of Interest

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