Gastroblastoma described in adult patient

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ABSTRACT

Gastroblastoma is a rare gastric epitheliomesenchymal biphasic tumor composed of spindle and epithelial cells. All cases have been reported in children or young adult, and as such, in view of the similarity with other childhood blastomas it has been postulated to refer to this condition as gastroblastoma. We report a woman of 53 years who has been diagnosed after realize an upper digestive endoscopy for peptic complaints, submitted a laparoscopic resection and followed without evidence of recurrence or metastasis 18 months after surgery. Therefore, this is the first case of gastroblastoma in the adult population, leading us to assume that is a tumor not exclusive of young patients.

Keywords: Adult gastroblastoma, Epitheliomesenchymal tumor, Gastric biphasic tumor

INTRODUCTION

Gastroblastoma which is a rare entity described as gastric epitheliomesenchymal biphasic tumor first reported by Miettinen in 2009 [1]. Only nine cases have been reported since then and all described cases arising in the stomachs of young adults [2]. An epitheliomesenchymal tumor in duodenum was already described by Poizat [3]. Regardless Poizat mentioned morphology close to what Miettinen described for gastroblastoma, the expression profile of immunohistochemical markers was distinct [3]. The etiopathogenesis, malignant potential and appropriate treatment for the gastroblastoma remains unknown [2], but recently Graham et al. have identified an oncogenic fusion gene that may serve as a diagnostic biomarker [4]. Here we report the first case of gastroblastoma in adult people and performed a review of the literature.

CASE REPORT

In December 2016, a 53-year-old woman was sent to our hospital by the medical assistant with complaints of heartburn and dyspepsia. The patient brought an upper digestive endoscopy (UDE) followed by a computed tomography (CT) of the abdomen with divergent information; UDE revealed the presence of a submucosal lesion that appeared to protrude from the great curvature near the gastric antrum (Figure 1) and CT images failed to show the described lesion.

We asked for an endoultrasonography that showed a 22.7 mm x 21.8 mm hypoechoic lesion with cystic components, irregular limits, presence of calcifications, apparent in continuity with muscle and growth into the peritoneal cavity without perilesional adenopathies (Figure 2A–D). A fine needle aspiration was done...
(Figure 2E) and evaluated by anatomopathologist that diagnosed as gastrointestinal stromal tumor (GIST).

She underwent laparoscopic atypical gastrectomy and the tumor was completely resected. The patient is still alive without evidence of recurrence and metastasis 18 months after surgery. On gross inspection, the partial gastrectomy specimen was described as 3.7 cm x 3.3 cm of distal stomach with a multinodular well-circumscribed tumor, exhibit 2.3 cm in its largest dimension. On the cut surface, the tumor was multinodular with color areas yellow and whitish. Microscopically, the tumor was centered in the muscular layer and had solid proliferation of variable cellularity, with biphasic areas well-demarcated (Figure 3A–B); one area with oval to spindle cells correspondent to the mesenchymal component (Figure 3C) and other with moderate pleomorphism relative to the epithelial component (Figure 3D). The mitotic index was fewer than 2 per 50 high-power fields and has a central sclerosed area, without foci of necrosis. There was no evidence of lymphovascular or perineural tumor invasion.

The proliferation index (Ki67) was low, estimated at under 5% (Figure 4). At immunohistochemical analysis the mesenchymal component had diffuse positivity for vimentin and CD10, multifocal positivity for CD56 and negativity for CD34, CD117, Cam 5.2, MNF, AE1/AE3, desmin, caldesmon, HMB45, WT1 and synaptophysin, and equivocal for DOG1 (Figure 5A); the epithelial component had diffuse/multifocal positivity for Cam 5.2, MNF and AE1/AE3, positivity focal for CD10 and CD56, negativity for CD117 and equivocal for DOG1 (Figure 5B). Reviewing the final anatomopathological information, the diagnosis was corrected for gastroblastoma and confirmed in an independent pathology service.

Figure 1: The upper digestive endoscopy revealed the presence of a submucosal lesion that appeared to protrude from the great curvature near the gastric antrum.

Figure 2: The endoultrasonography showed a 22.7 mm x 21.8 mm hypoechogetic lesion with cystic components, irregular limits, presence of calcifications, apparent in continuity with muscle and growth into the peritoneal cavity without perilesional adenopathies (A–D). A fine needle aspiration was done and evaluated by anatomopathologist (E).

Figure 3: Microscopically, the tumor was centered in the muscular layer and had solid proliferation of variable cellularity, with biphasic areas well-demarcated (A-B); the mesenchymal component with oval to spindle cells (C); the epithelial component with moderate pleomorphism (D).
DISCUSSION

GISTs are the most common mesenchymal tumors of the gastrointestinal tract and most frequently occur in the stomach, but typically not occur in young people. As gastric teratomas, inflammatory myofibroblastic tumors, synovial sarcomas and sarcomatoid carcinomas (carcinosarcomas) all may be differential diagnoses considering the clinical behavior. Our case presents clinical and morphological features similar to those epithelio-mesenchymal tumors of the stomach first described by Miettinen [1]. These cases described by Miettinen occurred in young adults and proposed to call them gastroblastomas in analogy to other childhood biphasic neoplasms for which the term blastoma is used [3]. However, when compared to other blastomas, such as pleuropulmonary blastoma, hepatoblastoma, and nephroblastoma, a type of tumors that tend to have immature elements and clear malignant potential, the gastroblastomas tend to demonstrate indolent behavior and excellent prognoses, probably related to a more developed architecture and low-grade cytologic features [5]. It has been proposed that gastroblastoma may relate better to the spindle epithelial tumor with thymus like differentiation and the desmoplastic nested spindle cell tumor of liver rather than blastomas [5].

The features reviewed by Toumi [2] in the other nine cases of gastroblastomas already reported tend to remain almost the same, as large tumor size, multinodular shape, two components with low-grade features and relatively low-mitotic activity, low overall atypia, absence of conspicuous nuclear pleomorphism, low malignant potential and disease limited to the stomach without metastatic potential or recurrence after curative resection. However, some differences had been reported, as one of the nine cases where Wey [5] mentioned existence of regional nodal and distant metastases at diagnoses, and other patient of this group described by Toumi [2] who developed loco-regional recurrence and local and distant lymph node metastases. Our case have brought some differences too, contrary to the postulate that gastroblastoma has been a disease only reported in young people, usually between the ages of 9–30 [6], mean age of 22 years [2], our patient is the first reported with 53 years. In addition, is the fourth patient female in a predominantly male disease population. Before our case, the tumors were relatively large by the time of diagnosis, ranging from 3.5 cm to 15 cm in the largest dimension (mean size 7.9 cm) [7], different from ours that appears with 2.3 cm. In adult population the use of endoscopic exams is more permissive what may justify the earlier diagnosis and the smaller size of the tumor find in our patient.

For diagnosing purposes using a biopsy specimen is probably impossible based on their different histological characteristics, so diagnostic confirmation is only possible with resection specimen and histopathological examination. Surgical resection with clear margins has been the preferred treatment of choice and laparoscopic approach is useful for small tumors less than 5cm located in the anterior surface of the stomach away from the gastroesophageal junction.

In literature, the follow-up periods are from 3 months to 14 years [7], with only one case reporting recurrence six months after curative resection [2]. So, gastroblastoma seem to have low malignant potential, but findings of metastatic disease suggest that at least some gastroblastomas are malignant [5].

CONCLUSION

Gastroblastomas is a distinct clinic-pathological entity and given the limited number of reported cases the prognosis is uncertain, being necessary more data to understand the biological and clinical behavior of the tumor.
REFERENCES


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Author Contributions
Diogo Nogueira Pinto – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

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