Journal on Oncology

www.journalononcology.org

Case Report

Open Access, Volume 3

Resection of a Gastroblastoma in an Adult Patient: An Unsuspected and Unusual Tumor

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Abstract

Introduction: Since the first description of gastroblastoma in 2009, 19 cases have been previously reported, most of them in young adults and children. The prognosis is uncertain, however, malignant tumors have been described.

Case report: A female patient, 44-years-old was studied with upper endoscopy and abdominal tomography for an incidentally found, submucosal tumor of the gastric antrum. She was operated on, and an antrectomy was performed. The results of the biopsy reported the histological and immunohistochemical characteristics of a gastroblastoma.

Literature review: Since the first description of gastroblastoma only case reports have been published. Consequently we performed a detailed review of all available cases.

Discussion: By including the present case, only 20 cases of gastroblastoma have been published to date. Most of them have benign behavior. However, some malignant cases raise concerns regarding whether a second look more aggressive surgery should be performed after diagnosing one of these tumors by primary surgical resection. Gastroblastoma malignant potential should be considered, and lymphatic dissection should be performed at the surgery. Longterm follow-up is important and recommended to avoid missing early or late recurrence.

Keywords: Gastroblastoma; Epithelioid-mesenchymal biphasic gastric tumor; Rare gastric tumors.

Introduction

The term gastroblastoma was proposed and adopted in 2009 following the description by Miettinen of 3 cases of such a tumor resected in 3 younger adult patients [1]. In his report, Miettinen described these tumors as rare epitheliomesenchymal neoplasms of the stomach that does not fit into recognized categories of biphasic tumors such as high-grade carcinosarcomas, sarcomatoid carcinomas and synovial sarcomas [1]. Since then, 19 cases of gastroblastoma have been reported, most of them in young adults and children [2,3]. The prognosis is uncertain, however, malignant tumors have been described [4,5]. Herein we report the case of a middle age woman with an incidentally found gastroblastoma.

Patient report: A female patient, 45-years-old, with gastritis and gastroesophageal reflux symptoms, was submitted to perform an upper endoscopy showing a 2 cm diameter submucosal lesion in the antrum towards the posterior wall, eroded and covered with fibrin (Figure 1). The biopsy reports mild active chronic gastritis with focal intestinal metaplasia and Helicobacter Pylori bacilli.

Manuscript Information: *Received: Nov 20, 2023; Accepted: Dec 14, 2023; Published: Dec 21, 2023*

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Citation: *Beltrán MA, Dictter C. Resection of a Gastroblastoma in an Adult Patient: An Unsuspected and Unusual Tumor. J Oncology. 2023; 3(2): 1121.*

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Evaluated at the surgery clinic, a computed tomography scan (CT) of the abdomen was requested, reporting a hypervascular endoluminal solid nodule on the anterior wall of the pylorus with a diameter of 2 cm compatible with a gastrointestinal stromal tumor (GIST), and without any sign of dissemination.

Surgery was scheduled for laparoscopic antrectomy and Roux Y reconstruction. The surgery was performed with endoscopic control due to the endophytic growth of the tumor. The patient was discharged on the 4th day.

The surgical biopsy reports diffuse sheets of uniform fusiform and dominant spindle cells with round nuclei and the cytoplasm in part eosinophilic, and clusters and cords of epithelioid cells. There are no areas of necrosis. The mitotic index was 1 per high power field. Cellular immunoreactivity was positive for cytokeratins CK AE1/AE3, CD56, CK7, Vimentin, and CD10. Cells were not reactive to CK20, CDX2, S100, CD117, DOG1, Chromogranin, and Synaptophysin. With the diagnosis of neoplasm of undetermined origin, new immunohistochemical studies on tumor cells were requested, reporting positive reactions with monoclonal antibodies to Cytokeratins AE1/AE3, CD56, and CD10. Negative reactivity was found for Chromogranin, Synaptophysin, SOX-10, and CD117. The Ki 67 Cell proliferation index was 1% to 2%. With these studies, the diagnosis of gastroblastoma was confirmed. Table 1, describes some characteristics of our patient shared with other published cases. Ten months after surgery, the patient is asymptomatic and continues in follow-up.

Table 1: Summary of 20 cases.

Author	Year	Age/ Gender	Mitoses/ 50 HPF	Gastric Location	Size (cm)	Surgery	Immunohistochemistry	Follow-up (months)/ Outcome
Miettinen et al.	2009	19/M	30	Greater curvature	5	Subtotal gastrectomy	CD10, CK AE1/AE3, Keratin 18, Keratin 7	42/ANED
Miettinen et al.	2009	27/F	4	Greater curvature	6	Partial gastrectomy	CD10, CK AE1/AE3, Keratin 18, Keratin 7	60/ANED
Miettinen et sal.	2009	30/M	1	Antrum	15	Antrectomy	CD10, CK AE1/AE3, Keratin 18, Keratin 7	168/ANED
Shin et al.	2010	9/M	1	Antrum	9	Antrectomy	CD10, CD56, CK AE1/AE3, Vimentin	9/ANED
Wey et al.	2012	28/M	35	Antrum	4	Antrectomy	CD10, CD56, CK AE1/AE3, CK7, Vimentin, Chromogranin A	3/ANED
Fernandes et al.	2014	19/F	5	Antrum	10	Antrectomy	CD10, CD56, CK AE1/AE3, Vimentin	20/ANED
Ma et al.	2014	12/M	40	Antrum	6	Antrectomy	CD10, CD56, CK AE1/AE3, CK CAM5.2, Vimentin	8/ANED
Toumi et al.	2017	29/F	21	Cardia	7	Partial gastrectomy	CD10, C D99, Vimentin	6/Deceased
Graham et al.	2017	28/M	3	Antrum	4	Antrectomy	CK AE1/AE3, CD56	Not reported/ Deceased
Graham et al.	2017	27/M	1	Antrum	9	Antrectomy	CK AE1/AE3, SMA, GLI1	12/ANED
Graham et al.	2017	9/M	1	Antrum	9	Antrectomy	CK AE1/AE3, CD10, CD56, Vimentin, GLI1	93/ANED
Graham et al.	2017	56/F	6	Antrum	4	Biopsy only	OSCAR, Vimentin, GLI1	Not reported/ Deceased
Castri et al.	2019	79/M	1	Antrum	9	Partial gastrectomy	CD10, bcl2, CD56, CK AE1/AE3, Vimentin	52/ANED
Centonze et al.	2019	43/F	2	Antrum	5	Partial gastrectomy	CD10, EMA, CK AE1/AE3, CK CAM 5.2, CK7, Vimentin, GLI1	100/ANED
Pinto et al.	2019	53/F	2	Antrum	2	Partial gastrectomy	Vimentin, CD10, CD56, CK AE1/AE3	18/ANED
Reardon et al.	2020	22/F	-	Antrum	7	Antrectomy	CK AE1/AE3, CAM 5.2, GLI1	Not reported
Koo et al.	2021	17/M	1	Fundus	6	Partial gastrectomy	Vimentin, CD56, CD10, CK AE1/AE3, Synaptophysin.	23/ANED
Liu et al.	2022	58/M	5	Greater curvature	2	ESD	Vimentin, CD10, bcl2, CD56, CD100, EMA	Not reported
Sugimoto et al.	2023	28/F	1	Antrum	7	Antrectomy	CD10, Vimentin, CD56, GLI1, PD-L1, CK AE1/ AE3, HDCA2, CAM5.2, CK7	8/ANED
Present case	2023	44/F	1	Antrum	2	Antrectomy	CD10, CD56, CK7, Vimentin	10/ANED

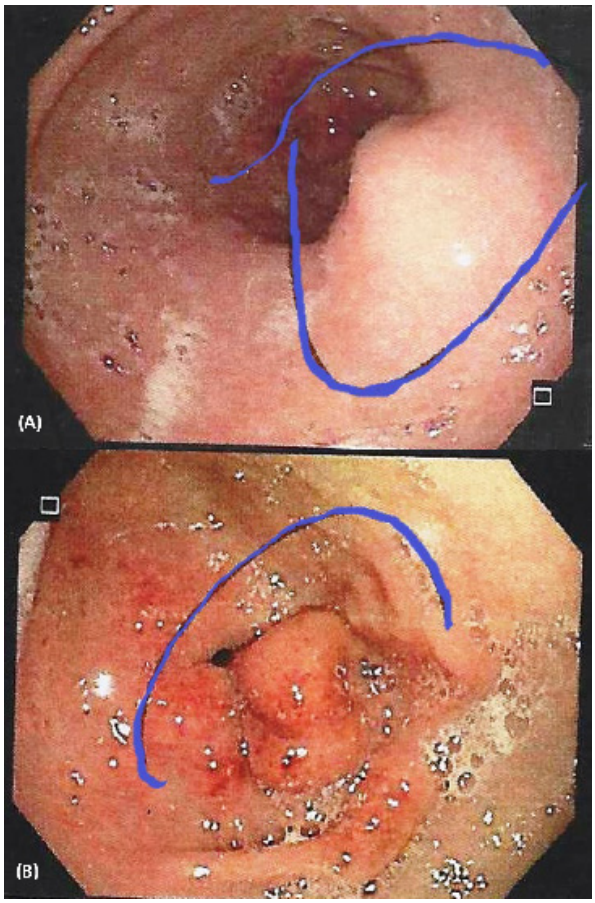

Figure 1: Endoscopic findings.
(A) Submucosal lesion towards the posterior wall of the gastric antrum. **(B)** Eroded submucosal lesion measuring approximately 2 cm.

Discussion

Gastroblastoma is a sporadic epithelioid-mesenchymal biphasic gastric tumor composed of uniform spindle and epithelial cells with an unclear etiopathogenesis, although it is believed that a totipotent cell could be the origin [2,5-7]. Recently, Graham et al, have confirmed that gastroblastoma is a distinct entity, and demonstrated that represent translocation-associated tumors characterized by the presence of a somatic, recurrent, oncogenic MALAT1-GLI1 fusion gene, the presence of which causes over-expression of GLI1 protein and of several of its downstream targets with key roles in tumorigenesis [8].

In most cases, this tumor has been reported in children or young adult males with unspecific symptoms [1,3,5,7,8,11]. Only some cases have been reported in older patients over 40-years-old [9,10]. Our patient was an adult female with vague symptomatology. Clinically, the symptoms are dominated by epigastric pain, impaired general condition, gastrointestinal bleeding, and palpable mass if the tumor is large enough [1,3,5,12]. On upper endoscopy, this tumor appears as a submucous mass, and the overlying gastric mucosa may be normal or ulcerated, as with our patient [7]. In other patients, this tumor is exophytic and not visible at endoscopy. In such cases, endosonography may be used to perform biopsies [4,5,10,11]. In most patients, the tumor was located at the antrum, as it was in our patient. Other locations are the greater curvature and under the cardias [1,5,12]. Gastroblastoma had mesenchymal component and epithelial elements; immunohistochemical markers such as Vimentin, CD10, CD56, and cytokeratins are differently expressed in the two tumor components. A low-grade malignant potential is suspected based on very few atypia, scarce mitosis, low ki-67 index, local growth pattern, and indolent clinical course [7]. Although, malignant behavior has been documented [4,5,6,8].

Radiologic studies such as and abdominal CT show the presence of a solid tumor with some cystic components arising from the gastric wall with exophytic or endophytic growth and some foci of calcification [5,6]. Tomographic characteristics are similar to GIST, which is the tumor most often confused with gastroblastoma. In our case, radiologists also confounded the gastroblastoma with a GIST, which led us to the local resection that we performed. Other studies such as magnetic resonance might confirm the tomographic findings, and better display the cystic components compared with CT [6,7].

Surgical resection with clear margins and lymphatic dissection might the treatment of choice. Lymph node dissection should be advocated because these tumors may spread via lymphatic vessels, and metastases to the liver and lymph nodes have been reported [5,6,8]. In one case, the outcome was the patient's demise due to metastatic dissemination [5]. However, gastroblastoma seem to have a low and uncertain malignant potential despite some reports on this tumor malignant behavior with ominous outcomes [10]. The issue of whether the gastroblastoma should be submitted to a second look surgery with lymphatic resection after primary economic resection is perhaps the current controversial subject of its treatment. The laparoscopic approach or local resection might be suitable for tumors less than 5 cm away from the gastroesophageal junction [1,2,9,10].

Macroscopically the gastroblastoma varies in size from a few centimeters to 15 cm or more [1,5,8]. It can appear as ulcerated endophytic masses, as polypoid tumors, intramural bulgings, or exophytic excrescences [1,5]. Grossly these tumors are described as multinodular or lobulated, and the cut surfaces varied from hemorrhagic to mottled surface [1,3,6]. The tumor originates in the muscular gastric wall layer [9,10]. The histology of gastroblastoma is biphasic, showing cell proliferation involving mesenchymal and spindle to ovoid cells, which are dominant component [1,5,7,9]. The epithelial component is organized mainly in sheets, nests, cords and tubules composed of glands, carpeted in places by the cylindrical cells, other characteristics are the hyperchromatic nuclei and the slightly eosinophilic cytoplasm, and discrete nucleoli [1,3,9]. In some cells, the nucleus and cytoplasm are condensed and clearly bordered by separate cells [5,7]. The nuclei of glandular structures are dark and elongated [1,5]. The mesenchymal cells are arranged in short fascicles or layers formed by oval cells with scant cytoplasm with inconspicuous nucleoli and regular nuclei. No signs of structural differentiation such as fences or nuclear vacuolation are noted [3]. Both components showed blastemal immature appearance. These tumors had consistent clinicpathological features; occurrence in young adults, relatively large tumor size, low-grade features with relatively low-mitotic activity and low overall atypia, and lack of overt pleomorphism [1,9]; characteristics shared with our patient.

Immunohistochemical studies show vimentin and CD10 reac-

tivity without any expression of muscle markers, CD34 or CD117 in the mesenchymal elements [1,8]. CD10 reactivity reflects fibroblastic phenotype [1]. Other commonly reactive markers in these tumors are CK AE1/AE3, CD56, epithelial membrane antigen, and CD117 in the epithelial components [3,7,8,10]. Neuroendocrine differentiation is absent in gastroblastoma [1]. It has been suggested the use of the GLI1 immunochemical stain to diagnose limited samples taken by cytologic aspiration by endosonography [11].

Several types of biphasic epitheliomesenchymal tumors are known to occur in the stomach, the differential diagnosis includes inflammatory myofibroblastic tumors, teratoma, gastrointestinal stromal tumor, biphasic synovial sarcoma, and carcinosarcomas [1,7]. Lymph node metastases have been reported in some cases, as well as liver metastases and peritoneal carcinomatosis [4,5,8]. No standard therapy has been established for this tumor. The intra-peritoneal chemotherapy could reduce the loco-regional recurrence and peritoneal dissemination [5]. Some reported cases received radiotherapy or chemotherapy with no response [1,4]. Gastroblastoma appears to have low malignant potential as recurrence after curative resection has seldom been reported [5]. The prognosis depends on several parameters including the size of the tumor, the degree of parietal invasion, mitotic index, and lymph node invasion; however, gastroblastoma seem to have a low malignant potential. Probably long-term follow-up should be advocated to avoid missing early and late recurrence.

Conclusion

Gastroblastoma is a distinct clinicopathological entity due to its clinical, radiological, histopathological, and immunohistochemical characteristics. Our case is the first reported case in Chile. To the best of our knowledge, only 19 other cases have been reported in the world literature. Despite the development of diagnostic, morphological, immune-histochemical, and anatomopathological techniques, diagnosis is often difficult. Gastroblastoma malignant potential should be considered, and lymphatic dissection should be performed at the surgery. In the case it was not performed, long-term followup is important to avoid missing early or late recurrence.

Declarations

Conflict of interest: None.

Patient consent: Was obtained for publication of the manuscript and all the images included.

Institutional Ethics Committee registration: 7C/004-23.

Funding: The authors declare that no funds, grants, or other support were received during the preparation of this manuscript.

Authors contribution: All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by Marcelo A. Beltrán, and Constanza Dictter. The first draft of the manuscript was written by Marcelo A. Beltrán and all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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