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Case Report

Primary Gastric Sarcoma: An Uncommon but Possible Diagnosis

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Abstract

Gastric cancer is a neoplastic disease of undeniable importance, which corresponds to adenocarcinoma in 90% of the cases. The remaining 10% are almost entirely lymphomas and Gastrointestinal Stromal Tumors (GIST); however, sarcomas remain a likely differential diagnosis to keep in mind and constitute a neoplastic pathology that is treated fundamentally with surgery. Because of their rarity, we present this case report and literature review.

Keywords: Leiomyosarcoma; Stomach Neoplasm

Introduction

A In a classic article published in 1948 in JAMA, Drs. Bassler and Peters [1] mention the clinical differences between sarcomas and gastric carcinomas and invite us to have in mind the diagnostic possibility of sarcoma in the clinical setting of malignant gastric neoplasia. Some of the relevant features of sarcomas mentioned in that paper, are the early age of presentation, with an average of 38 years; more frequent in men, prolonged clinical pictures with less cachexia than in carcinomas, and abdominal pain as the main symptom frequently associates with bleeding. Since then, our knowledge about sarcomas has grown, and its classification has been reviewed many times. We present the case of a young man with a family history of gastric adenocarcinoma, who debuted with pain and digestive bleeding, secondary to a primary gastric leiomyosarcoma and review the literature.

Case Presentation

We report the case of a 42-year-old man with a 3-month history of abdominal pain and upper gastrointestinal bleeding. A family history of grandfather and father with gastric adenocarcinoma, at 70 and 53 years respectively. Upper endoscopy describes a subcardial, ulcerated mass in greater curvature. The biopsy refers to a hyperplastic polyp negative for malignancy. Upon arrival at the National Cancer Institute, a new endoscopy and biopsy are performed. The findings are ulcerated polypoid lesions, 6x8cm subcardial lesions with a 3cm implantation base, lobed and firm. The pathology corresponds to a fusocellular mesenchymal lesion that, after the immune histochemical study, is classified as a leiomyoma. Meanwhile the patient has presented two episodes of digestive bleeding that have required transfusion and persists with difficult abdominal pain.

Extension studies with contrast-enhanced tomography of the thorax and abdomen that demonstrate the mass and exclude metastatic lesions (Figures 1 and 2) are performed and the total gastrectomy with Roux-en-Y reconstruction is performed (Figure 3). Postoperative evolution is favorable and is discharged at 7 days. The definitive pathological diagnosis is a 14x12cm moderately differentiated leiomyosarcoma with 50 mitoses in 50 high power fields, 40% Ki-67; focal necrosis <50%. Tumor free margins and 15 tumor free lymph nodes (Figures 4, 5 and 6). Follow-up at 17 months has not shown tumor recurrence and the patient's overall condition is good. Digestive pain and bleeding completely remitted after surgery.

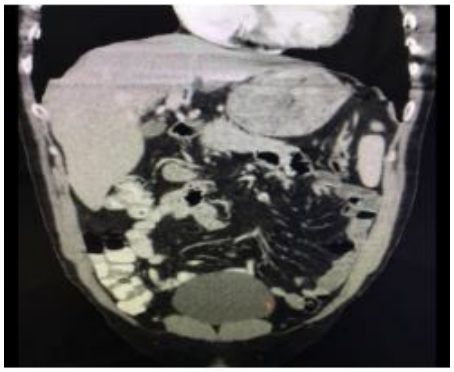

Figure 1: Abdominal CT that Shows the Gastric Mass. Coronal Section.

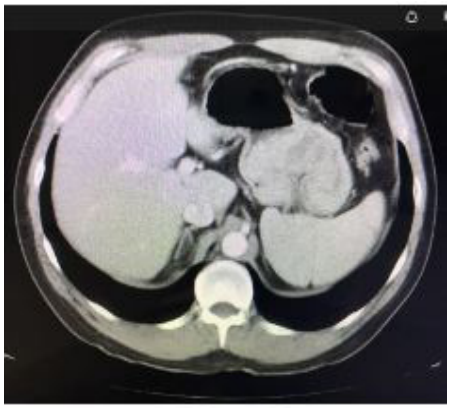

Figure 2: Abdominal CT Scan Showing Gastric Mass. Axial Section.

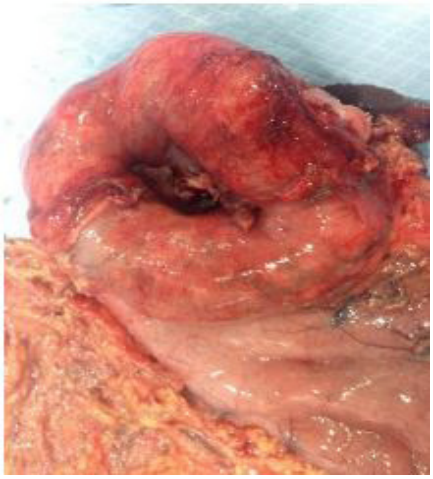

Figure 3: Surgical Piece of Gastrectomy with the Lesion.

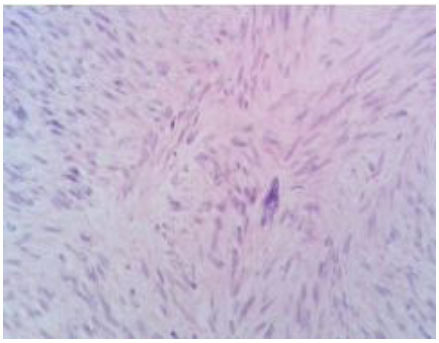

Figure 4: Hematoxylin-Eosin Staining. Spindle cells with Atypical Nucleus.

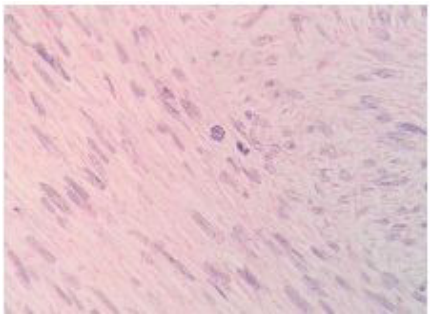

Figure 5: Hematoxylin-Eosin Staining. Atypical Mitosis.

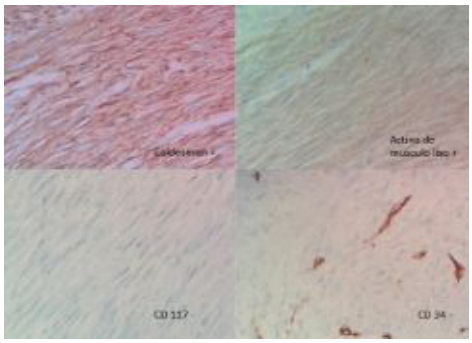

Figure 6: Positive immunohistochemistry for Smooth Muscle Actin and Caldesmon, Negative for CD 117 and CD34.

Discussion

Sarcomas make up 1 to 2% of gastrointestinal malignancies, most of which correspond to leiomyosarcomas and the most frequent location is the stomach [2]. Other types of sarcomas in the gastrointestinal tract, such as synovial, granulocytic and Ewing's sarcoma, have been reported in isolation and are true rarities [3-5].

Mitotic activity has been shown to be the single most important indicator of disease prognosis and surgery remains the mainstay of treatment [6-8]. Likewise, high tumor grade and incomplete surgical resection are independent factors of poor prognosis [9]. The nature of these neoplasms and the peculiarities of their biological behavior imply that the surgical resection en bloc, even with neighboring organs, is fundamental in reducing the possibility of relapse. Unlike adenocarcinomas, sarcomas grow locally and invade by contiguity, with a low tendency to lymph node metastases [10]; however, the extent of the indicated nodal dissection is a reason for discussion as there are reports of ganglionic involvement in up to 15% Of cases [11]. Adjuvant treatments such as chemotherapy and radiotherapy play a secondary role and are indicated almost exclusively for palliative purposes in case of unresectable or relapsed tumors [12].

In reviewing the classification of sarcomas, the place of leiomyosarcomas has been relatively stable, differing fundamentally from GISTs by immune histochemical techniques [13]. Because they constitute intramural, extra mucosal lesions, diagnosis by endoscopy and biopsy is not always easy. Clinical suspicion, leading to in-depth biopsies and imaging techniques are key in establishing preoperative diagnosis [14]. Differentiation between leiomyoma and leiomyosarcoma is also difficult to establish on biopsies and the surgical specimen is often required to establish the diagnosis of certainty, since the presence of cellular atypia and mitosis is the key to differential diagnosis [15].

In the literature, there are many classifications for sarcomas in general and specifically for soft tissue sarcomas. The Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) has proposed a classification system for visceral and soft tissue sarcomas, based on three aspects: degree of differentiation, ulceration and mitotic rate; This system has been validated and widely used [16]. The present case is a high-grade sarcoma, according to the classification of FNCLCC with total score 7, for differentiation (score 2), mitosis (score 2) and necrosis (score 2).

Parameter	Criterion
Differentiation	
Score 1	Well differentiated, structure reminiscent of mature tissue.
Score 2	Moderately differentiated; Defined histological type.
Score 3	Undifferentiated, structure reminiscent of embryonic tissue.
Mitosis Count	
Score 1	0-9 in 10 High power field
Score 2	10-19 in 10 High power field
Score 3	>20 in 10 High power field
Tumor Necrosis	
Score 1	No necrosis
Score 2	≤50% necrosis
Score 3	>50% necrosis
Histologic Grade	
Grade 1	Total score 2-3
Grade 2	Total score 4-5
Grade 3	Total score 6-8

Table 1: Adapted from: Baig M (2015) Evaluation of performance of various histological grading systems of soft tissue sarcomas and the prognosis (metastatic risk and survival rate). International Journal of Research in Medical Sciences 3: 2394-2401 [16].

Other factors, such as tumor size more than 5 cm and the presence of metastases at the time of diagnosis, are important in prognosis and are included in the North American Joint Committee on Cancer (AJCC) TNM staging system [14].

The overall 5-year survival rate for gastric sarcomas is variable and has been reported between 16 and 56% depending on tumor grade and success in complete surgical resection; Recurrences usually occur within two years of resection and range from 36% to 60% [12].

In conclusion, we present a case of primary gastric leiomyosarcoma in a young man and we review the peculiarities of this rare disease.

Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

Authors' Contributions

All authors equally contributed to this study.

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