

Multifocal angiosarcoma of the stomach and jejunum: A case report

Maher Al Khaldi, Anne-Marie Dufresne, DesGroseilliers S.

ABSTRACT

Introduction: Gastrointestinal angiosarcoma is a very rare soft-tissue malignancy with a poor prognosis. There are only seven cases of angiosarcoma to have ever involved the stomach in the actual literature. **Case Report:** This report describes a unique case of a multifocal angiosarcoma of the stomach and jejunum in a 79-year-old female who displayed non-specific symptoms of abdominal pain, postprandial heaviness and lower left quadrant pain. A computed-tomography scan revealed an ulcerated lesion in the stomach. Upon surgical exploration, two primary lesions were found in the stomach and two similar lesions in the jejunum. After surgical resection of the involved parts, the diagnosis of primary angiosarcoma was made by pathological and immunohistochemical examination. A favorable two month postoperative follow-up was noted. **Conclusion:** This is the first documented case in the English literature of four simultaneous primary angiosarcoma lesions involving the stomach and the small intestine. Given its rare nature, patients with angiosarcoma could be misdiagnosed. Standard treatment is surgery. However, more studies are needed to establish the

role of adjuvant chemotherapy and radiotherapy in the treatment of angiosarcoma.

Keywords: Angiosarcoma, Epigastric discomfort, Jejunum, Stomach

How to cite this article

Al Khaldi M, Anne-Marie D, DesGroseilliers S. Multifocal angiosarcoma of the stomach and jejunum: A case report. J Case Rep Images Surg 2016;2:50–56.

Article ID: 100022Z12MK2016

doi:10.5348/Z12-2016-22-CR-14

INTRODUCTION

Angiosarcoma is a rare, malignant neoplasm. It can occur in any part of the body, more commonly in the skin and soft tissues, and less commonly in the breast, bone, liver and spleen [1]. Primary angiosarcoma of the gastrointestinal tract is even more rare, especially in the stomach. This is a case of a multifocal angiosarcoma of the stomach and jejunum.

CASE REPORT

This is the case of a 79-year-old female who complained of epigastric discomfort with postprandial heaviness. She also described lower left quadrant pain and discomfort. She had no history of hematemesis, hematochezia or melena. Past medical history included breast epithelioma diagnosed at the age of 68 and collagenous colitis. Past surgical history included unilateral partial mastectomy with axillary clearance followed by adjuvant radiotherapy

Maher Al Khaldi¹, Anne-Marie Dufresne¹, DesGroseilliers S.¹

Affiliations: ¹General Surgeon, General Surgery Department, Pierre-Boucher Hospital, Longueuil, Quebec, Canada.

Corresponding Author: Maher Al Khaldi, 2830 Goyer, apartment 112, Montreal, Quebec, Canada, H3S1H4; Email: alk-maher@gmail.com

Received: 08 May 2016
Accepted: 04 June 2016
Published: 15 June 2016

completed at the age of 68, bilateral total hip replacements and abdominal aortic aneurysm repair.

Upon initial investigation by the patient's general practitioner, a barium swallow revealed a 5x4 cm gastric lesion with irregular contour located at the junction between the proximal third and the middle of the stomach. An upper gastrointestinal endoscopy was further performed and revealed two distinct, raised, ulcerated lesions in the stomach. A contrast-enhanced thoracic-abdominal-pelvic computed tomography (CT) scan was performed to rule out metastases. The CT scan revealed no signs of metastases (Figure 1).

Biopsies obtained during endoscopy showed ulcerated gastric mucosa with fibrinous and leukocytic exudate at the surface of the lesions and a poorly differentiated malignant infiltration at deeper layers. Malignant cells with fusiform to epithelial shapes were found around the ulcer. A few foci revealed irregularly-shaped, anastomosing vascular structures lined with hyperchromatic cells.

An extensive immunohistochemical study was performed with the following antisera: cytokeratin 20, cytokeratin 17, cytokeratin AE1/AE3, CD3, CD20, CD30, Ki67, vimentin, Melan-A, CD117, CD79A, LCA, cytokeratin 5/6, EMA, GATA3, S100 protein, synaptophysin, SMA, calretinin, CD31, CD138 and CEA. The cells stained positive only for CD31, vimentin and, to a lesser extent, CD34. The expression of the cellular marker for proliferation, Ki67, was close to 100%. All lymphocyte surface markers remained negative, including the human leukocyte common antigen (LCA), which eliminated the possibility of a lymphoproliferative process. All epithelial and melanocytic markers were negative. Given that that the patient had previously been diagnosed with breast cancer, and considering the possibility of an infiltrating metastasis originating from a breast carcinoma, immunohistochemical evaluation of GATA3 expression was performed but was negative. Having ruled out the possibility of a mesothelioma or a leiomyosarcoma with a negative staining for calretinin and SMA respectively, the most probable diagnosis was a high-grade angiosarcoma of the stomach.

During surgical exploration (Figure 2), many lymph nodes were noted in the mesentery of the proximal jejunum. Two gastric lesions were found in the body of the stomach (Figure 3). In addition, the involved segment of jejunum was resected and two lesions were found, very similar in appearance to the ones in the stomach. A subtotal gastrectomy with Roux-en-Y anastomosis was performed following the segmental jejunal resection. The margins of resection were 5 cm, as recommended for a gastric adenocarcinoma. Finally, immediate post-operative course was favorable and the patient has been well 2 months after operation. Close follow-up of the patient is still undergoing.

DISCUSSION

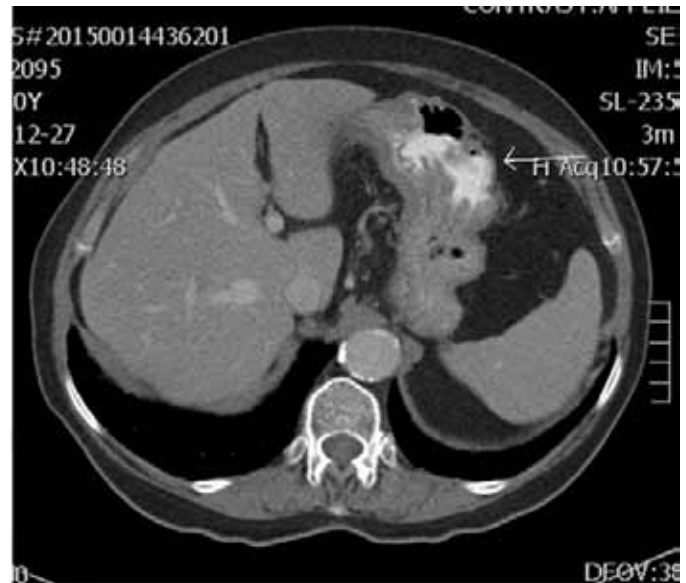


Figure 1: Contrast-enhanced axial computed tomography of the abdomen revealing a cavitory lesion in the stomach filled with contrast (arrow).



Figure 2: Intraoperative presentation of one of the ulcerated lesions of the stomach.

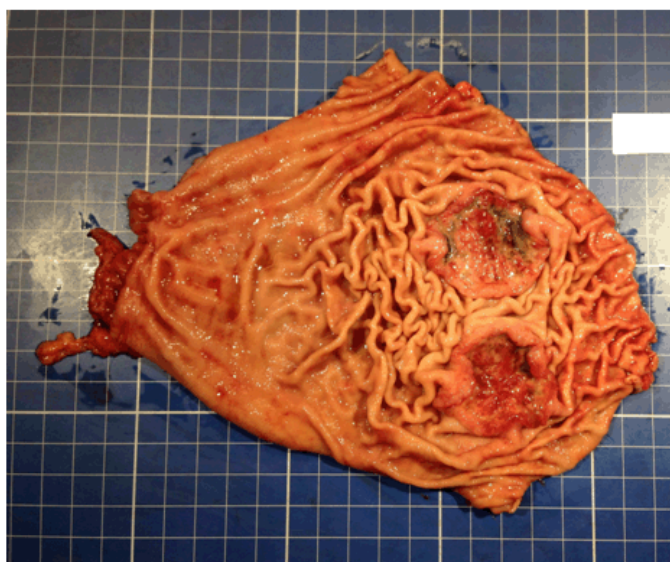


Figure 3: Longitudinally lanced stomach demonstrating two irregularly-shaped submucosal lesions similar in appearance located at the body.

Angiosarcoma is a very rare, malignant neoplasm that is characterized by invasive anaplastic cells derived from blood vessels [2]. It accounts for less than 1% of soft tissue sarcomas. They may occur in many regions of the body, most frequently in the skin and soft tissues, and very rarely in the gastrointestinal tract [3]. Its aggressive characteristics and rarity make it an often misdiagnosed malignancy. Prognosis is usually poor and the mortality rate is high, partly due to a late diagnosis [4].

There have been only seven published cases of angiosarcoma with gastric involvement, four of which involved primary lesions (Table 1) [5–10]. Five patients were men and two were women. Their ages ranged from 21–86 years with a median of 55 years. Three cases were multifocal (involving the stomach and the small intestine), three cases involved a single, isolated gastric angiosarcoma and one case involved three angiosarcomas of gastric origin. Clinical manifestations included abdominal pain, nausea, vomiting and melena. One patient however was asymptomatic. Surgical resection was the most common treatment. The outcome was generally poor. Four patients died several months after diagnosis or treatment. No information was gathered regarding the longest survival period.

Angiosarcoma tends to be multicentric in occurrence [1], it is sometimes difficult to distinguish between multicentricity and metastasis because of its aggressive nature. What makes this case unique is the simultaneous presence of multicentric and multifocal primary lesions rather than metastases.

The carcinogenic mechanism of angiosarcoma remains unclear. However, several risk factors associated with skin and soft tissue angiosarcomas have been documented in the past, including toxic

exposure to environmental carcinogens, radiation therapy, chronic lymphedema, foreign materials and certain familial syndromes such as neurofibromatosis and BRCA1/BRCA2 mutations [2]. However, no risk factors associated to gastric angiosarcomas have been documented, with the exception of a possible association with long-term hemodialysis [1]. In the present case, the patient underwent post-mastectomy therapeutic irradiation. However, radiation-induced angiosarcomas usually occur in areas of previous radiation. Indeed, the most common sites for sarcomas in previously irradiated patients are the female breasts, followed by female genital organs [11]. Several documented cases of intestinal angiosarcoma have occurred following radiation therapy of gynaecological malignancies [1]. However, no cases ever mentioned a link between breast irradiation and angiosarcoma of gastric and intestinal involvement.

Symptoms associated with gastrointestinal angiosarcoma are non-specific. As documented in previous case reports (Table 1), symptoms could include vague abdominal pain, nausea, post-prandial heaviness or melena. Gastrointestinal bleeding and anemia are usually associated with advanced disease [12].

Angiosarcoma tends to be poorly differentiated. Thus, immunohistochemical staining is a very important step in confirming the diagnosis. In this case, positive staining for CD31, CD34 and vimentin helped confirm the vascular origin of the lesions. Similar staining has been confirmed in previous case reports. Staining for factor VIII-related antigen could also be positive in angiosarcoma as reported in several previous cases [1, 7, 8]. In one case of an isolated gastric angiosarcoma lesion, cytokeratin (AE1/AE3) expression was present [9].

Histological evidence in previous cases demonstrated angiosarcomas epithelioid in morphology. In fact, angiosarcomas of deep soft tissues tend to be epithelioid in morphology. On the contrary, lesions in the skin and subcutaneous tissues are mostly nonepithelioid [12].

Treatment of gastric angiosarcoma consists of surgical resection. Chemotherapy and radiation therapy have been used in several cases of gastrointestinal angiosarcoma [4]. However, the role of adjuvant therapy remains unclear.

Patients diagnosed with angiosarcoma with gastrointestinal involvement usually have poor prognoses. Metastases to lymph nodes and other organs are not uncommon and even expected given the aggressive nature of angiosarcomas. Indeed, many cases progress rapidly with a median survival of only two months after diagnosis [13].

CONCLUSION

In conclusion, a few cases of angiosarcomas with gastric involvement have ever been reported in English literature. Although gastrointestinal angiosarcoma remains rare, it is important to consider it as a rare differential diagnosis in patients complaining of vague

Table 1: Cases of primary angiosarcoma involving the stomach reported in English literature

Author	Gender and age (yr)	Site(s) of gastro-intestinal involvement	Histology	Positive IHC* staining	Possible risk factor	Presenting symptoms	Metastases	Treatment	Outcome
Multicentric angiosarcoma in the gastrointestinal tract including the stomach									
Vrind [5]	M, 46	Stomach (location not specified), duodenum, ileum	Epithelioid	NA**	NA	Abdominal pain, melena	Lymph nodes, pancreas, liver	Surgical resection	Died after six months
Usuda [1]	M, 45	Stomach (location not specified), duodenum, jejunum, ileum, caecum	Epithelioid	Vimentin, Factor VIII-related antigen, Collagen type IV, CD31, CD34	Long-term dialysis	Melena	Lymph nodes, lungs, bones, liver, gallbladder	Surgical resection	Died of respiratory failure eight months after initial presentation
Amy [6]	M, 70	Stomach (location not specified), secondary intestinal involvement (location not specified)	Epithelioid (the diagnosis of poorly differentiated adenocarcinoma was initially made on the biopsy)	Cytokeratin, Vascular antigens	NA	NA	NA	Surgical resection, chemotherapy	Died eight months after chemotherapy
Isolated gastric angiosarcoma									
Seki [10]	F, 21	Stomach	NA	NA	NA	Melena	Liver, peritoneal omentum	NA	NA
Taxy [8]	M, 86	Stomach (one lesion in the body)	Malignant cells with fusiform to epithelial shapes	Vimentin, Factor VIII-related antigen, Collagen type IV	None	Epigastric pain and discomfort, occasional vomiting	NA	None	Patient was alive four months after biopsy
Tai [9]	M, 55	Stomach (one lesion in the body)	Mixed: moderately differentiated and epithelioid cells	CD34, Vimentin, Cytokeratin (AE1/AE3)	NA	Asymptomatic	NA	Surgical resection	Patient has been well during the 11-month postoperative follow-up
Multifocal isolated gastric angiosarcoma									
Park [7]	F, 65	Stomach (three lesions in the body and antrum)	Epithelioid	Factor VIII-related antigen, CD31, CD34	NA	Epigastric pain, nausea, vomiting	Left thalamus	Surgical resection	Died one month after the initial diagnosis

Table 1:(Continued)

Author	Gender and age (yr)	Site(s) of gastro-intestinal involvement	Histology	Positive IHC* staining	Possible risk factor	Presenting symptoms	Metastases	Treatment	Outcome
Multicentric and multifocal angiosarcoma (including stomach)									
Present case	F, 79	Stomach (2 lesions in body and fundus), jejunum (2 lesions)	Malignant cells with fusiform to epithelial shapes	CD31 CD34 Vimentin	Radiation therapy (although area of irradiation was relatively far from stomach and small intestine)	Abdominal pain, postprandial heaviness	None	Surgical resection	Patient has been well during the 2-month postoperative follow-up

*IHC: immunohistochemical

**NA: Not Applicable

gastrointestinal symptoms and when dealing with previously irradiated patients. The mainstay of treatment is surgery. Further studies are needed to clarify the role of adjuvant radiation therapy and chemotherapy in the treatment of gastrointestinal angiosarcoma given its very aggressive nature.

Acknowledgements

We are thankful to Mr. Majd Al Khaldi. Dr. Pierre Bergeron and Dr. Luc Larivière for contributing in the preparation of this article

Author Contributions

Maher Al Khaldi – 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

Anne-Marie Dufresne – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

DesGroseilliers S. – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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