CASE REPORT PEER REVIEWED | OPEN ACCESS

Primary angiosarcoma of the small bowel: A case report

Maria Olim Sousa, Ricardo Cabrita Viveiros, Diana Fernandes, Rómulo Ribeiro, Lídia Ferreira, Ana Filipa Capelinha

ABSTRACT

Introduction: Small bowel angiosarcomas are exceedingly rare neoplasms with unspecific symptomatology which may lead to a delay in the diagnosis and consequently a worst prognosis. Case Report: A 73-year-old male patient presented with nausea, vomiting and abdominal pain. The blood test showed a mild anemia. Computed tomography (CT) scan revealed an ileal tumor. The patient was submitted to an exploratory laparotomy and segmental enterectomy. Pathology findings described an angiosarcoma. The patient had disease progression and died after two months. Conclusion: Angiosarcomas are high grade rapidly progressive neoplasms and have a very poor prognosis with a high mortality rate. The average life expectancy is 2–6 months after diagnosis.

Keywords: Small bowel, Cancer, Primary angiosarcoma, Obstruction

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INTRODUCTION

Angiosarcomas account for 1–2% of soft tissue sarcomas and are preferentially localized to skin and superficial soft tissue. Rare cases are described in heart, liver, spleen and adrenal glands, being the digestive tract localization an exceptionally rare occurrence [1–3].

Malignant tumors of the small intestine make up only 1– 1.6% of all gastrointestinal tract tumors.

The most widely recognized predisposing factors for angiosarcomas of skin and soft tissue are radiation and chronic lymphedema (Stewart-Treves syndrome). There is also a strong association with contact to some chemical agents such as thorium dioxide, arsenic, vinyl chloride or to foreign material introduced iatrogenically like vascular graft material or by trauma such as foreign bodies [1, 4].

Clinically, like other tumors of the small bowel, angiosarcomas have nonspecific symptoms including recurrent gastrointestinal bleeding, abdominal pain and nausea [2, 5–7]. The few cases described in the literature report rapid dissemination and very reserved prognosis with average survival from two to six months after diagnosis [3, 6].

The authors present a clinical case of a primary angiosarcoma of the small bowel.

CASE REPORT

A 73-year-old male, with irrelevant personal history and no known exposure to chemical toxins, surgery, chemotherapy or radiation was admitted to the emergency department with abdominal pain in the right lower quadrant and vomiting for three days. Physical examination revealed a palpable mass in the mesogastric region of the abdomen. Laboratory testing detected a mild anemia (Hb 11.3 mg/dL) with no other analytic abnormalities. The computed tomography demonstrated an abdominal tumor measuring approximately 12.3x11.6x7 cm in the proximal ileum, with regional lymph nodes and a moderate amount of intra-abdominal free fluid (Figure 1).

Laparotomy showed a small bowel obstructing tumor and a minimal amount of bloody ascites. No other lesions were observed. A segmental enterectomy was performed (Figure 2). Pathological analysis revealed an angiosarcoma and immunohistochemistry shows positivity for factor VIII, CD 34, CD31 and negativity for CD 117, S100 and MITF (Figure 3). The tumor was 7 cm in larger axis and had metastasis in 4 of 16 nodes removed.

Due to the rarity of randomized trials and prospective studies, the management guidelines for other soft tissue sarcomas tend to be utilized when dealing with angiosarcoma. According to 7th edition of AJCC Soft-Tissue Sarcoma Staging System, this case was classified as pT2bN1M0. The presence of positive nodes (N1) in M0 tumors is considered stage III.

There were no complications during the postoperative period. Adjuvant chemotherapy was decided by multidisciplinary team discussion.

One month after surgery was readmitted with abdominal pain, vomiting, ascitic abdominal distension and worsening anemia (Hb 9.2 mg/dL). Paracentesis was performed and 1000 mL of serous-hematic fluid was drained. The ascitic fluid was positive for malignant cells. CT scan showed extensive peritoneal carcinomatosis (Figure 4). Patient experienced a progressive abdominal distention, worsening of pain and anemia despite transfusions; loss of weight with severe malnutrition and rapid deterioration with multiorgan failure; he died within two months after the initial diagnosis.

DISCUSSION

The primary angiosarcoma of the small bowel like other neoplasms on this location present with nonspecific symptomatology that may include abdominal pain, nausea, vomiting, intestinal obstruction, gastrointestinal bleeding and anemia [2, 4].

The patient, in our case report, presented with nonspecific complaints of pain and vomiting, and had no identifiable risk factors described in the medical literature [1]. This may have been the reason for delayed diagnosis and consequent poor prognosis.

Magnetic resonance imaging (MRI) scan, computed tomography (CT) scan, abdominal X-rays and ultrasound can be used for diagnosis, but all of them have limited diagnostic utility [5, 6].

Immunohistochemistry is the only method to confirm the diagnosis [7]. Immunohistologically, intestinal angiosarcomas are positive for endothelial markers as CD31, CD34, Von Willebrand factor and vascular endothelial growth factor and negative for epithelial,

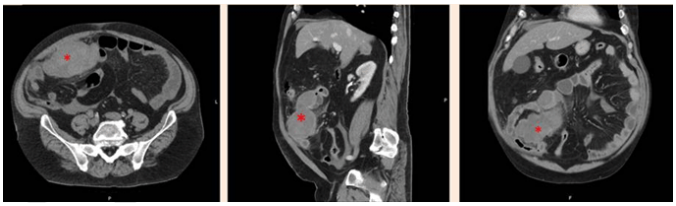

Figure 1: Computed tomography scan showing an abdominal tumor.

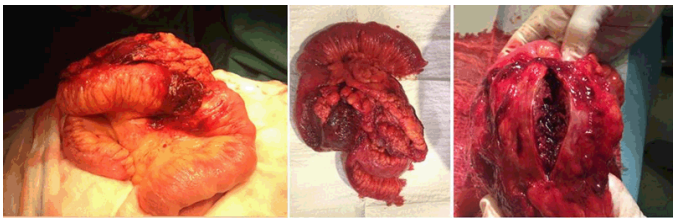

Figure 2: Laparotomy findings: small bowel obstructive tumor. A segmental enterectomy was performed.

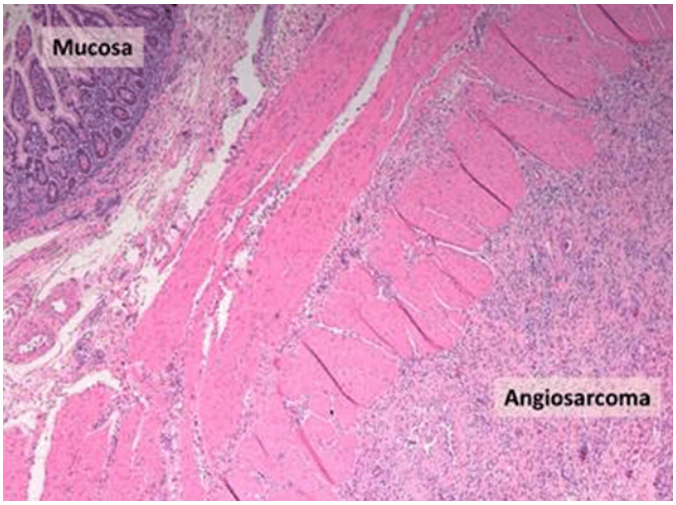

Figure 3: Histological sections of intestinal tissue observed through an optical microscope (hematoxylin eosin). Specimen showed an angiosarcoma. Immunohistologically: Factor VIII+, CD34+, CD31+, CD117-, S100- and MITF-.

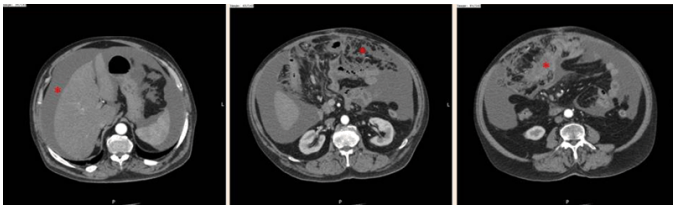

Figure 4: Computed tomography scan evidencing extensive peritoneal carcinomatosis.

neuronal and melanocytic markers as Keratins, S-100 and HMBE-45 [1, 4, 7].

Despite its aggressive behavior, angiosarcoma of the small bowel is an extremely rare entity [3]. In a literature review only 22 cases of truly primary angiosarcoma of small intestine cases were described (Table 1) [8].

There is no established standard treatment, and treatment itself becomes difficult owing to late detection due to inaccessible localization and the nonspecific symptoms.

The attempt to treat these patients requires a multidisciplinary team that may include radiologists,

Table 1: Cases of confirmed primary angiosarcoma involving the small intestine reported in English literature

Authors	Sex/age (years)	Site	Immuno-histochemical staining	History of prior radiation or other predisposing factor	Presentation	Treatment	Follow-up
Siderits et al.	M/79	Small bowel	Strongly positive for CD31	None	Obstruction	Resection	Unknown
Taxy and Battifora	M/64	Small bowel	Positive for Factor VIII, collagen type IV and vimentin	Not available	Gastrointestinal bleeding	Resection	Died 1 year after the initial diagnosis
Taxy and Battifora	F/57	Small bowel	Positive for Factor VIII, collagen type IV	Not available	Not available	Resection	Died shortly after surgery
Chami et al.	M/59	Small bowel	Weakly positive for factor VIII-related antigen, Ulex europaeus I antigen and cytokeratin	None	Gastrointestinal bleeding, bowel obstruction, anorexia and weight loss	Resection and transfusions	Died on the 11th day after surgery
Ordonez et al.	M/80	Small bowel	Positive immunoreaction for FVIII-RAG	None	Anemia, undue tiredness and weakness	Resection	Died on the 20th postoperative day
Hwang et al.	F/60	Small bowel	Positive for Ulex europaeus agglutinin 1	History of radiotherapy	Diffuse abdominal pain	Resection	Died 2 months after discharge
Mohammed et al.	F/25	Small bowel	Not available	None	Intermittent abdominal pain, weight loss, abdominal distension, hematemesis and malaena	Resection	Died on the 11th day after surgery
Fraiman et al.	M/85	Small bowel	Strong positivity for vimentin and CD31; focal positivity for factor VIII and CD34	None	Weight loss, anemia, weakness and abdominal pain	Resection and thalidomide	Not available
Selk et al.	M/57	Small bowel	Not available	History of radiation therapy	Progressive abdominal distention and shortness of breath	Resection	Died 4 months after surgery
Berry et al.	M/51	Small bowel	Positive for Ulex europaeus and vimentin	History of 3-year irradiation	Peritonitis	Resection, adriamycin and dacarbazine	Died 5 months after initial presentation

Khalil et al.	M/68	Small bowel	Strongly positive for CD31, CD34 and vimentin	30 year history of heavy occupational exposure to radiation and polyvinyl chloride	Gastrointestinal bleeding and melaena	Resection	Died 6 months after initial presentation
Suzuki et al.	F/61	Ileum	Positive for factor VIII-related antigen and Ulex europaeus agglutinin 1	20 year history of radiotherapy	Abdominal pain	Resection and intraabdominal cisplatin	Died 1 year after initial presentation
Delvaux et al.	M/67	Small bowel	Positive for CD 31, CD 34, factor VIII-related antigen and keratin	Not available	Weight loss, intermittent severe abdominal pain and melaena	Resection	Died 3 months after diagnosis
Policarpio-Nicolas et al.	F/51	Small bowel	Positive for CD 31, CD 34 and factor VIII-related antigen	History of irradiation	Abdominal pain	Resection	Died 10 months after laparotomy
Hansen et al.	F/76	Small bowel	Positive for factor VIII and vimentin	History of irradiation	Watery diarrhea, vomiting, weight loss and abdominal pain	Resection	Died 5 months after operation
Aitola et al.	F/50	Small bowel	Positive for CD 31, CD 34 and factor VIII-related antigen	≥10 year history of radiotherapy	Intestinal obstruction	Resection followed by combination chemotherapy with doxorubicin	1 year and 9 months after diagnosis, she was alive
Ogawa et al.	M/36	Small bowel	Positive for factor VIII-related antigen	Not available	Abdominal pain and nausea	Resection	Not available
Liu et al.	F/39	Terminal ileum	Positive for CD31 and CD34	None	Increasing right iliac fossa pain, abdominal bloating and vomiting	Resection and chemotherapy	Not available
Kelemen et al.	M/76	Small bowel	Positive for CD31	None	Abdominal pain and fatigue	Resection	Died of cardiac arrest on the 9th day after surgery
Fohrding et al.	M/84	Small bowel	Positive for CD31, cytokeratin and vimentin; slightly weaker for CD34; Focally positive for factor VIII	Not available	Gastrointestinal bleeding	Resection, adjuvant chemotherapy with paclitaxel and transfusion	Not available
Grewal et al.	M/73	Small bowel	Positive for CD31	None	Gastrointestinal bleeding, weakness and melaena	Resection	Died within 4 months of the diagnosis
Qingquianq Ni et al.	M/33	Small bowel Liver metastasis	Positive for CD31 and vimentin	None	Abdominal pain, vomiting, weight loss, fatigue, fever	Resection, adjuvant chemotherapy	Died on 27th postoperative day

pathologists, surgical oncologists, medical oncologists and radiation oncologists. Although the cornerstone of the treatment is surgical complete resection when possible to achieve maximal locoregional control, treatment often includes palliative resection of the bleeding or obstructing lesions, chemotherapy, radiotherapy and best supportive care which may include massive blood transfusions [3].

In 1999, Aitola et al. reported a case of a small bowel tumor resection followed by combination chemotherapy with doxorubicin which survived one year and nine months after diagnosis [9]. The patient described had 14 years previously undergone total hysterectomy and salpingo-oophorectomy for a stage I adenocarcinoma of the uterine corpus and received 55.6 Gy external radiation therapy to the lower pelvis. In May 1997, at the age of 50 years, she was again admitted to hospital due to repeated symptoms of intestinal obstruction. Complementary study demonstrated a constant 5-cm-long stricture at the terminal ileum. Laparotomy revealed a 20 cm long segment of thickened terminal ileum, an extended ileocecal resection was performed. The patient received 6 adjuvant doses of doxorubicin (110 mg). Relaparotomy was undertaken one year and nine months after diagnosis of the angiosarcoma from the operative specimen, and this showed wide intra-abdominal spread and retroperitoneal recurrence. This case is the one with the highest survival described in literature in patients with this disease. Most patients die within a few months after diagnosis with an average survival from two to six months after diagnosis secondary to refractory bleeding and disease progression [2, 3, 6, 10–13].

CONCLUSION

Angiosarcoma of the small bowel is an exceptionally aggressive and rare entity with very poor prognosis. Early diagnosis is a challenge due to the nonspecific symptoms. Imaging studies can be extremely important in timely finding these lesions, but high clinical suspicion based on clinical history is necessary for diagnosis. There are no defined guidelines or demonstrated efficacy of adjuvant treatment due to the low incidence of this pathology, thus, the multidisciplinary approach of these patients is of utmost importance.

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Author Contributions
Maria Olim Sousa – 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
Ricardo Cabrita Viveiros – 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
Diana Fernandes – 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
Rómulo Ribeiro – 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
Lídia Ferreira – 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
Ana Filipa Capelinha –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

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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