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,
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>
<thead>
<tr>
<th>Authors</th>
<th>Sex/age (years)</th>
<th>Site</th>
<th>Immunohistochemical staining</th>
<th>History of prior radiation or other predisposing factor</th>
<th>Presentation</th>
<th>Treatment</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Siderits et al.</td>
<td>M/79</td>
<td>Small bowel</td>
<td>Strongly positive for CD31</td>
<td>None</td>
<td>Obstruction</td>
<td>Resection</td>
<td>Unknown</td>
</tr>
<tr>
<td>Taxy and Battifora</td>
<td>M/64</td>
<td>Small bowel</td>
<td>Positive for Factor VIII, collagen type IV and vimentin</td>
<td>Not available</td>
<td>Gastrointestinal bleeding</td>
<td>Resection</td>
<td>Died 1 year after the initial diagnosis</td>
</tr>
<tr>
<td>Taxy and Battifora</td>
<td>F/57</td>
<td>Small bowel</td>
<td>Positive for Factor VIII, collagen type IV</td>
<td>Not available</td>
<td>Not available</td>
<td>Resection</td>
<td>Died shortly after surgery</td>
</tr>
<tr>
<td>Chami et al.</td>
<td>M/59</td>
<td>Small bowel</td>
<td>Weakly positive for factor VIII-related antigen, Ulex europaeus I antigen and cytokeratin</td>
<td>None</td>
<td>Gastrointestinal bleeding, bowel obstruction, anorexia and weight loss</td>
<td>Resection and transfusions</td>
<td>Died on the 11th day after surgery</td>
</tr>
<tr>
<td>Ordonez et al.</td>
<td>M/80</td>
<td>Small bowel</td>
<td>Positive immunoreaction for FVIII-RAG</td>
<td>None</td>
<td>Anemia, undue tiredness and weakness</td>
<td>Resection</td>
<td>Died on the 20th postoperative day</td>
</tr>
<tr>
<td>Hwang et al.</td>
<td>F/60</td>
<td>Small bowel</td>
<td>Positive for Ulex europaeus agglutinin I</td>
<td>History of radiotherapy</td>
<td>Diffuse abdominal pain</td>
<td>Resection</td>
<td>Died 2 months after discharge</td>
</tr>
<tr>
<td>Mohammed et al.</td>
<td>F/25</td>
<td>Small bowel</td>
<td>Not available</td>
<td>None</td>
<td>Intermittent abdominal pain, weight loss, abdominal distension, hematemesis and malaena</td>
<td>Resection</td>
<td>Died on the 11th day after surgery</td>
</tr>
<tr>
<td>Fraiman et al.</td>
<td>M/85</td>
<td>Small bowel</td>
<td>Strong positivity for vimentin and CD31; focal positivity for factor VIII and CD34</td>
<td>None</td>
<td>Weight loss, anemia, weakness and abdominal pain</td>
<td>Resection and thalidomide</td>
<td>Not available</td>
</tr>
<tr>
<td>Selk et al.</td>
<td>M/57</td>
<td>Small bowel</td>
<td>Not available</td>
<td>History of radiation therapy</td>
<td>Progressive abdominal distention and shortness of breath</td>
<td>Resection</td>
<td>Died 4 months after surgery</td>
</tr>
<tr>
<td>Berry et al.</td>
<td>M/51</td>
<td>Small bowel</td>
<td>Positive for Ulex europaeus and vimentin</td>
<td>History of 3-year irradiation</td>
<td>Peritonitis</td>
<td>Resection, adriamycin and dacarbazine</td>
<td>Died 5 months after initial presentation</td>
</tr>
<tr>
<td>Authors</td>
<td>Gender</td>
<td>Site</td>
<td>Immunohistochemistry</td>
<td>Clinical History</td>
<td>Symptoms</td>
<td>Treatment/Chemotherapy</td>
<td>Outcome</td>
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<tr>
<td>Khalil et al.</td>
<td>M/68</td>
<td>Small bowel</td>
<td>Strongly positive for CD31, CD34 and vimentin</td>
<td>30 year history of heavy occupational exposure to radiation and polyvinyl chloride</td>
<td>Gastrointestinal bleeding and melaena</td>
<td>Resection</td>
<td>Died 6 months after initial presentation</td>
</tr>
<tr>
<td>Suzuki et al.</td>
<td>F/61</td>
<td>Ileum</td>
<td>Positive for factor VIII-related antigen and Ulex europaeus agglutinin 1</td>
<td>20 year history of radiotherapy</td>
<td>Abdominal pain</td>
<td>Resection and intraabdominal cisplatin</td>
<td>Died 1 year after initial presentation</td>
</tr>
<tr>
<td>Delvaux et al.</td>
<td>M/67</td>
<td>Small bowel</td>
<td>Positive for CD31, CD34, factor VIII-related antigen and keratin</td>
<td>Not available</td>
<td>Weight loss, intermittent severe abdominal pain and melaena</td>
<td>Resection</td>
<td>Died 3 months after diagnosis</td>
</tr>
<tr>
<td>Policarpio-Nicolas et al.</td>
<td>F/51</td>
<td>Small bowel</td>
<td>Positive for CD31, CD34 and factor VIII-related antigen</td>
<td>History of irradiation</td>
<td>Abdominal pain</td>
<td>Resection</td>
<td>Died 10 months after laparotomy</td>
</tr>
<tr>
<td>Hansen et al.</td>
<td>F/76</td>
<td>Small bowel</td>
<td>Positive for factor VIII and vimentin</td>
<td>History of irradiation</td>
<td>Watery diarrhea, vomiting, weight loss and abdominal pain</td>
<td>Resection</td>
<td>Died 5 months after operation</td>
</tr>
<tr>
<td>Aitola et al.</td>
<td>F/50</td>
<td>Small bowel</td>
<td>Positive for CD31, CD34 and factor VIII-related antigen</td>
<td>≥10 year history of radiotherapy</td>
<td>Intestinal obstruction</td>
<td>Resection followed by combination chemotherapy with doxorubicin</td>
<td>1 year and 9 months after diagnosis, she was alive</td>
</tr>
<tr>
<td>Ogawa et al.</td>
<td>M/36</td>
<td>Small bowel</td>
<td>Positive for factor VIII-related antigen</td>
<td>Not available</td>
<td>Abdominal pain and nausea</td>
<td>Resection</td>
<td>Not available</td>
</tr>
<tr>
<td>Liu et al.</td>
<td>F/39</td>
<td>Terminal ileum</td>
<td>Positive for CD31 and CD34</td>
<td>None</td>
<td>Increasing right iliac fossa pain, abdominal bloating and vomiting</td>
<td>Resection and chemotherapy</td>
<td>Not available</td>
</tr>
<tr>
<td>Kelemen et al.</td>
<td>M/76</td>
<td>Small bowel</td>
<td>Positive for CD31</td>
<td>None</td>
<td>Abdominal pain and fatigue</td>
<td>Resection</td>
<td>Died of cardiac arrest on the 9th day after surgery</td>
</tr>
<tr>
<td>Fohrding et al.</td>
<td>M/84</td>
<td>Small bowel</td>
<td>Positive for CD31, cytokeratin and vimentin; slightly weaker for CD34; focally positive for factor VIII</td>
<td>Not available</td>
<td>Gastrointestinal bleeding</td>
<td>Resection, adjuvant chemotherapy with paclitaxel and transfusion</td>
<td>Not available</td>
</tr>
<tr>
<td>Grewal et al.</td>
<td>M/73</td>
<td>Small bowel</td>
<td>Positive for CD31</td>
<td>None</td>
<td>Gastrointestinal bleeding, weakness and melaena</td>
<td>Resection</td>
<td>Died within 4 months of the diagnosis</td>
</tr>
<tr>
<td>Qingqianq Ni et al.</td>
<td>M/33</td>
<td>Small bowel</td>
<td>Positive for CD31 and vimentin</td>
<td>None</td>
<td>Abdominal pain, vomiting, weight loss, fatigue, fever</td>
<td>Resection, adjuvant chemotherapy</td>
<td>Died on 27th postoperative day</td>
</tr>
</tbody>
</table>
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

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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

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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

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Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES