Case Report

Primary Mesenteric Angiosarcoma in an Adult Presenting with Peritonitis: A Case Report and Review of Literature

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Abstract

Primary mesenteric angiosarcoma is a rare soft tissue tumour accounting for 1%–2% of soft tissue tumours. Because of the ubiquity of blood vessels and lymphatics, it can occur anywhere with a poor prognosis. To the best extent of our literature search, this type of tumour is sparsely reported in our environment; we hereby report a case of primary mesenteric angiosarcoma in a 32-year-old man who presented with acute abdomen, intraperitoneal haemorrhage, and shock. Abdominal ultrasound revealed intraperitoneal collection, and intraoperative findings revealed haemoperitoneum 1.5l with vascular cystic tumour at the mesentery close to the duodenojejunal junction. Histopathological examination of the resected specimen revealed angiosarcoma. The patient was seen last at follow-up in early 2022.

Keywords: Angiosarcoma, mesentery, peritonitis

Introduction

Angiosarcoma is very rare in adults; it is an aggressive tumour that arises from vascular endothelial cells and which has a poor prognosis. It accounts for 1%–2% of soft tissue sarcomas. They can arise from any location due to the ubiquity of blood vessels and lymphatics, but commonly occurs in the head and neck. There are cutaneous, visceral, and soft tissue types of angiosarcoma with the visceral form accounting for 15%–47% of such neoplasm. Angiosarcomas have been reported in the breast, heart, lung, liver, spleen, adrenal glands, ovaries, and rarely in the gastrointestinal (GI) tract, especially the small intestine, which has only been found in scattered case reports and small series.[1,2]

The presentation of bowel/mesenteric angiosarcoma is nonspecific, including radiologic findings and histologic diagnosis in most cases requiring biomarkers and has a median survival of 18–28 months. The difficulty in diagnosis has led to an underestimation of its true incidence. Less than twenty-five percentage of angiosarcomas are soft tissue angiosarcomas, and of these, <5% are intra-abdominal.[3,4]

We describe a case of mesenteric angiosarcoma in an adult who presented with abdominal pain and intraperitoneal haemorrhage.

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Case Summary

A 32-year-old university student presented to the accident and emergency unit with a one-day history of sudden-onset central abdominal pain radiating to the right iliac fossa with abdominal distension and has an associated history of nausea, but no vomiting. His medical history was unremarkable. On examination, he was afebrile, and had a regular pulse of 92 bpm, but was hypotensive with a blood pressure of 98/60 mmHg. He had a distended abdomen with tenderness in the periumbilical region and the right iliac fossa with peritonism. Initial blood tests were normal apart from the haemoglobin of 8.8 g/dl. An abdominal ultrasound scan showed free fluid in the entire abdomen, and the bowel loops showed poor peristalsis. After resuscitation with intravenous fluids, an urgent exploratory laparotomy was performed. During operation, 1.5 L of blood was found in the abdominal

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cavity, the bleeding originating from a tumour at the root of the mesentery close to the duodenojejunal junction. *En bloc* resection of the tumour was done. Macroscopically, the resected tumour was circumscribed, measures about 8 cm × 7.5 cm × 7 cm and weighs 175 g. The cut surface shows thick grey-brown haemorrhagic areas and grey tissue. Pathological examination of the resected specimen as showed angiosarcoma as shown in [Figures 1-4], and the patient was subsequently referred to the oncologist for chemotherapy.

Discussion

Malignant mesenteric angiosarcomas are extremely rare tumours. Less than one percentage of all sarcomas are angiosarcomas, <25% of angiosarcomas are soft tissue angiosarcomas, and of these, <5% are intra-abdominal. These tumours seem to be squarely distributed between the decades (25–85 years) with a mean age of 68.5 years, and there is no obvious sex difference is apparent, while other authors reported a male preponderance of 1.6: 1.[2,4,5]

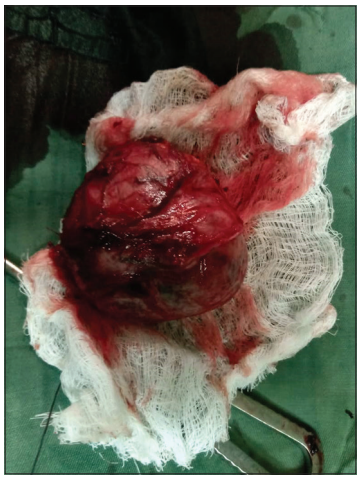

Figure 1: Intraoperative image – The resected specimen

This tumour is known to present with nonspecific clinical and radiologic findings, and also present with abdominal pain, anaemia, GI bleeding, fatigue/weakness, weight loss, dyspnea, abdominal distension/bowel obstruction, anorexia, nausea/vomiting, acute abdomen, and a bowel perforation.[2,4,6-10] The imaging modalities for diagnosis are also nonspecific, including abdominal pelvic ultrasound, abdominopelvic *computed tomography* scan, abdominal *magnetic resonance imaging*, endoscopy, and enteroscopy.[2-4]

The etiology is unknown, but there is an increasing evidence of the role of radiation in the development of soft tissue angiosarcomas, particularly intra-abdominal lesions. They have also been reported with retained foreign bodies like swabs.[8,11,12] Other risk factors include exogenous toxins such as vinyl chloride, arsenic, anabolic steroids, chronic lymphoedema,

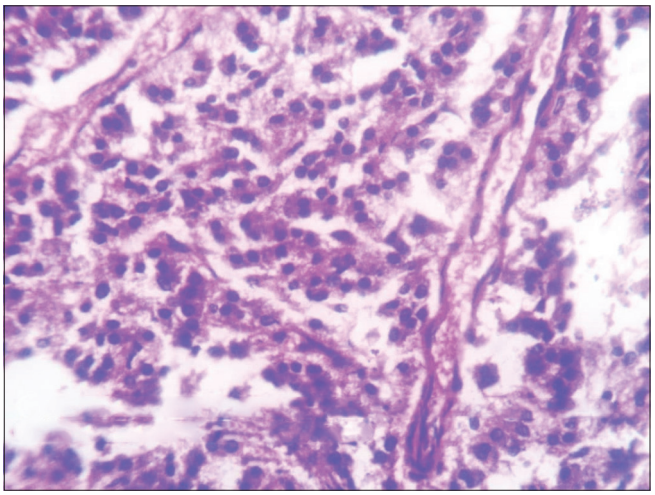

Figure 2: Histology of the resected tissue (angiosarcoma of the mesentery) – H and E, ×40 showing an invasive neoplasm with tumour cells interspersed by proliferating connecting vascular channels

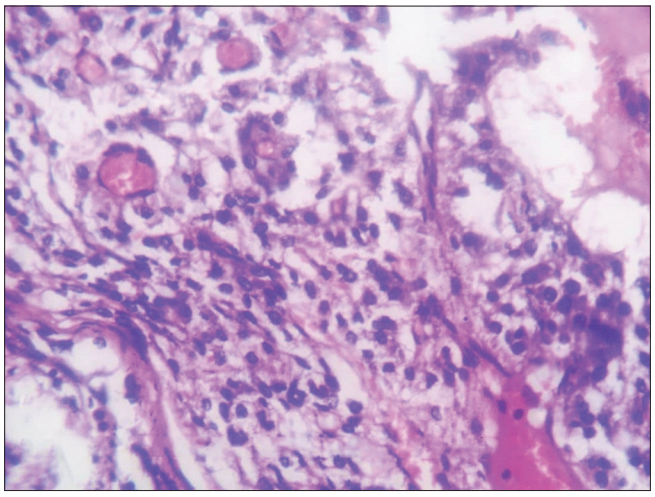

Figure 3: Histology of the resected tissue (angiosarcoma of the mesentery) – H and E, ×40 showing proliferating thin-walled vascular channels

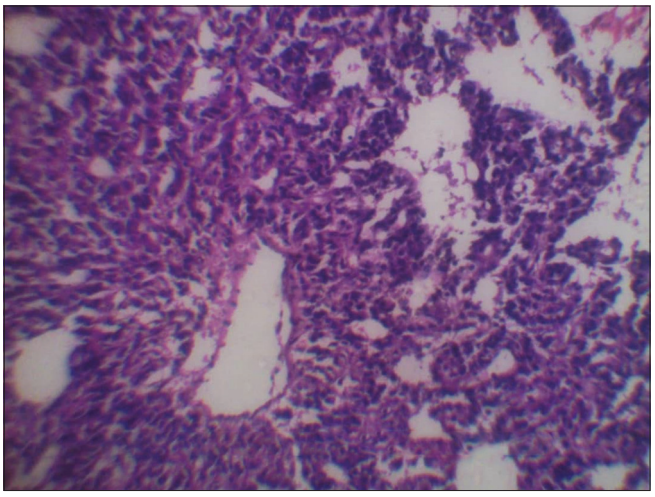

Figure 4: Histology of the resected tissue (angiosarcoma of the mesentery) – H and E, ×20 showing epithelioid tumour cells with hyperchromatic nuclei

familial syndrome – Neurofibromatosis 1, BRACA, Maffucci syndrome, and Klippel–Trenaunaysyndrome.[2,4,13]

The tumour spreads to lymph nodes, liver, lung, and spleen. Cutaneous angiosarcomas can metastatise to the bowel; although in our case, there was no evidence of metastasis, the anatomic distribution is as follows: ileum 29.8%, jejunum 27.7%, and unspecified 23.4%.[2,4] Primary malignant tumours of the mesentery are similar to those described for the omentum. Liposarcomas, leiomyosarcomas, malignant fibrous histiocytomas, lipoblastomas, and lymphangiosarcomas have all been described.[2-4,14]

The tumour cells are strongly positive for the vascular markers CD31 and CD34 and the lymphatic marker D2-40. Ki67 immunostain shows high cell proliferative activity throughout the tumour. The tumour cells are negative for S100, MART1, smooth muscle actin, pan-cytokeratin AE1/AE3, and desmin. Other makers include endothelin 1, vascular endothelial growth factor receptor, ERG, and Ulex europaeus agglutinin-1.[2-4,8,12,15]

Treatment of mesenteric malignancies involves a multidisciplinary approach involving the surgeon primarily, the pathologist radiation, and the medical oncologist. Because of Proximity to the messenteric vessels wide resection of the masss may be technically unfeasable because of the risk of substantial loss of the lenght of the small bowel.[2,4] Other treatment options include paclitaxel-based chemotherapy, intraperitoneal chemotherapy, radiotherapy, and biologic therapy with bevacizumab, sorafenib, and sunitinib.[2,4,9,16]

The outcome of treatment and prognosis will depend on the stage of the disease and the treatment modalities. It has been shown that a combination of surgery and chemotherapy improves survival which our patient had.[2,4]

Conclusion

A high index of suspicion is required for early diagnosis and prompt treatment to prevent morbidity and mortality; similarly, combination therapy offers a better survival opportunity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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