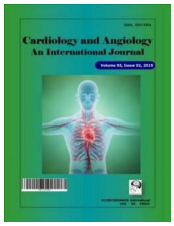

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Angiosarcoma of Large Intestine: A Case Report

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Authors' contributions

This work was carried out in collaboration between all authors. Author SMS designed the study, wrote the protocol and wrote the first draft of the manuscript. Authors NA and KA performed and managed the statistical analysis of the study. Authors UM, LTO, KOE and BMB managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Angiosarcomas are collectively one of the rare forms of soft tissue neoplasms. That of the large intestine is much rarer. We report a case of large intestinal angiosarcoma. This is a case of a 30-year-old man who presented with intestinal obstruction, and after resection, it was histologically diagnosed as Angiosarcoma of large bowel.

Keywords: Angiosarcoma; rare; large intestine.

1. INTRODUCTION

Angiosarcoma (AS) refers to a malignant epithelial neoplasm arising from endothelial cells of blood vessels. It is usually seen in adults and the elderly, but it can also occur in children. The most common locations are the skin, soft tissue, breast, bone, liver and spleen [1,2]. They can occur in any region of the body, but 60% arise in skin or superficial soft tissue. Fifty percent of cutaneous AS occurs in the head and neck [3].

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Angiosarcomas are poorly differentiated neoplasms composed of neoplastic malignant endothelial cells and have a very narrow lumen filled with blood cells. A review and analysis of 366 cases of angiosarcomas at the Armed Forces Institute of Pathology (AFIP) during a 10-year period showed absence of large intestinal angiosarcoma among them [1].

Angiosarcomas are commonly detected during the initial six months of life since they usually involve the soft tissues of the head and skin. Angiosarcomas are frequently seen in the liver in children [4]. In adults, angiosarcomas are very rare findings, being mostly described in the liver, lungs, brain and bones [5]. Angiosarcomas of the intestine are extremely rare in adults [6]. Immunohistochemical demonstration of CD31, and less consistently the von Willebrand factor is diagnostically used.

2. PRESENTATION OF CASE

We report a case of a 30-year-old man who presented with a 5-day history of abdominal pain and inability to pass stool with associated progressive abdominal distension. The patient had an appendicectomy two months prior to presentation in a peripheral hospital on account of recurrent right lower quadrant abdominal pain. Also, he had left inguinal herniorrhaphy seven months before the presentation.

Physical examination revealed a pale middle aged man, who was anicteric, acyanosed, mildly dehydrated no peripheral stigmata of chronic liver disease and no splenomegaly. Ascites was demonstrable by shifting dullness. Abdominal girth measures 185 cm.

Haematological tests showed a microcytic hypochromic anemia with packed cell volume of 27%. The electrolyte, urea and creatinine were elevated, serologic test for HIV-1 and HIV-2 were non-reactive. Ultrasonography showed moderate ascites with multiple solid masses involving large bowel. The liver, spleen, urinary bladder and kidneys were within normal limits.

The patient was resuscitated and prepared for exploratory laporotomy. Findings included ascitic fluid of 2.8L of serosanguinous fluid with the tumour involving the caecum, ascending colon and hepatic flexure. Resections and anastomosis were done. A segment of large bowel was submitted to the laboratory for histological examination, variably sized firm to hard tumour nodules were received with a tangled mass. In one area, there was a friable fungating tumour. The whole specimen measures 23x20x11 cm and weighs 2.5 kg. Cut surfaces show fleshy masses of pale, gray-white, soft tissue and greenish mucosa in areas. Representative sections were taken and routinely stained with H & E, then examined histologically.

Histologic sections showed a vasoformative lesion composed of anastomosing vascular channels lined by atypical endothelial cells exhibiting moderate pleomorphism and hyperchromasia. The cells ranged from elongated to multinucleated forms with numerous hyaline globules seen. The tumour infiltrated and destroyed serosa and muscularis propria with foci of necrosis noted. Immunohistochemical stain for CD31 was positive. A diagnosis of AS was made (Figs. 1, 2 and 3).

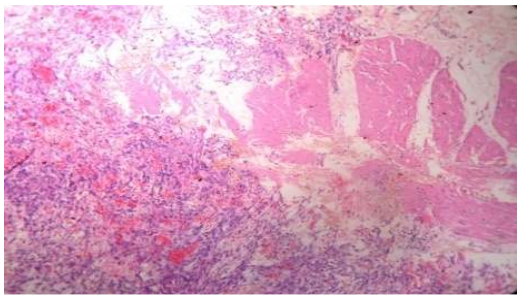

Fig. 1. Photomirograph showing a vasoformative lesion composed of anastomosing vascular channels lined by atypical endothelial cells exhibiting moderate pleomorphism and hyperchromasia infiltrating the muscularis propria (H&E mag.x100)

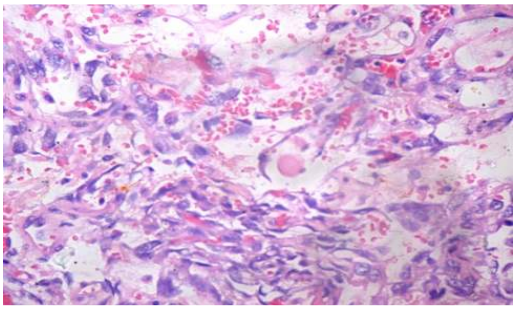

Fig. 2. Photomicrograph of tumor cells at higher power showing tumor cells that ranged from elongated to multinucleated forms with numerous hyaline globules (H&E mag.x400)

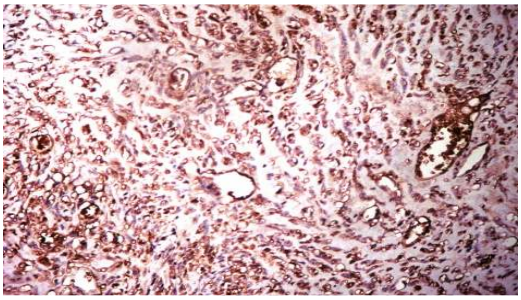

Fig. 3. Immunohistochemical stain showing tumor cells expressing positivity for CD31 (mag.x200)

3. DISCUSSION

ASs are collectively one of the rarest forms of soft tissue neoplasm. They account for the small proportion of all vascular tumour, and they comprise less than 1% of all sarcomas. Although they occur at any location in the body, they rarely arise from major vessels and the occurrences in large bowel are rarer [7].

ASs are rare neoplasms characterized by proliferation of tumor cells with vascular endothelial features, accounting for only 1–2% of all soft tissue sarcomas [5,8]. They occur most commonly in the scalp, skin and soft tissues of the head and neck region in elderly men.

Intra-abdominal ASs are very rare neoplasms, which usually arise in the liver or spleen and extremely rarely in the gastrointestinal tract. From 1965 till 2003, only 13 cases of small intestine ASs have been reported [5,8–13] in the English literature. Of these, none have been reported to occur at the large intestine.

The pathogenesis of intestinal AS is unclear, several etiologic factors have been suggested such as previous irradiation, chronic lymphedema, exogenous toxin like thorostat, vinyl chloride and arsenic, long term peritoneal dialysis, intra-abdominal foreign body. However, none of this factors can be linked to this case [5,8,14,15,16].

ASs have a similar distribution between both sexes. They can develop at any age, being commoner in older patients as opposed to the index case who is 30 years old. The usual presentation of large bowel AS include abdominal pain, nausea, vomiting and abdominal distension, all of which were present in this patient [17].

The clinical presentations as highlighted above can also be caused by the following tumours - gastrointestinal stromal tumour (GIST), leiomyosarcoma, a metastatic melanoma, lymphoma, neuroendocrine tumour, Crohns disease; and mesothelioma [3,17].

Histopathologically, these tumours are highly vascularised with abundance of endothelial cells and areas of solid and spindle cell tumour with an infiltrative and destructive growth pattern typical of AS. Immunohistochemically, ASs of large intestine/bowel express endothelial markers including von Willibrand factor, CD34 and CD31, which is the same in our own case that expressed CD31 [18,19].

The treatment of choice is radical surgery with wide margins of resection because of invasive and often multifocal nature of AS but this is difficult to achieve. The role of adjuvant therapy is unclear, but the survival benefit is limited [1-3].

4. CONCLUSION

AS of the large bowel is a rare malignancy, presentation clinically can mimic that other diseases. It therefore means that AS should be included in differential diagnosis of clinician dealing with gastrointestinal tumour or diseases.

CONSENT

All authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this paper and accompanying images.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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