International Surgery Journal
Aziz Md T et al. Int Surg J. 2016 May;3(2):956-958
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902
DOI: http://dx.doi.org/10.18203/2349-2902.isj20161175

Case Report

Angiosarcoma of small intestine presenting with intestinal obstruction

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Received: 27 December 2015
Accepted: 04 February 2016

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ABSTRACT

Angiosarcomas are rare high-grade vascular tumors that mostly have poor prognosis. The pathogenesis of an intestinal Angiosarcoma is not clear and many etiologic factors have been suggested. In this report, we reviewed the English-written literature about primary intestinal Angiosarcomas and analyzed a case of 50-year-old male patient with intestinal Angiosarcoma presenting with intestinal obstruction and acute abdominal pain and absolute constipation. We report a unique case of Angiosarcoma of small intestine presented with intestinal obstruction. A 50-year-old male patient came with complaint of pain, vomiting, abdominal distension and absolute constipation. Investigations suggested he was anemic. X-ray abdomen erect suggested multiple air fluid level. USG revealed intestinal obstruction. Emergency Exploratory Laparotomy was performed; there was Intra-abdominal tumor present over the serosal surface of ileum with torsion causing Volvulus of small bowel leading to obstruction. Histopathology and Immunohistochemistry confirmed the tumor to be Angiosarcoma arising from small intestine. Literature review has suggested exploratory laparotomy is the definitive diagnosis. Prognosis tends to be favorable after resection.

Keywords: Primary angiosarcoma, Small intestine, Intestinal obstruction, Abdominal pain, Intestinal tumour

INTRODUCTION

Primary Angiosarcomas are very rare and Angiosarcoma of small intestine is even rarer, with poor prognosis.[1] Angiosarcomas which account for only 1% to 2% of all soft tissue sarcomas are rare malignant tumors of endothelial origin.[2] These tumors have predilection for skin and superficial soft tissue, breast, bone, liver; they are rarely seen in deep soft tissue.[3] They are associated with chronic lymphedema, previous radiotherapy, and arterio-venous fistulas in renal transplant patients.[1,4] In our case report, we present a 50-year-old male presented with pain, vomiting, abdominal distension and absolute constipation.

CASE REPORT

A 50-years male came to Casualty with complaint of severe and continuous abdominal pain especially near umbilical region and absolute constipation since 2 days. Pallor was present. Per Abdomen Inspection revealed fullness over whole abdomen. Abdominal examination revealed tenderness over abdomen especially over umbilical region and distension of abdomen. He was anemic with hemoglobin 5.2gm/dl with ESR raised to 97mm/hr. X-ray Abdomen Erect suggested dilated small bowel loops with multiple airs-fluids level seen (Figure 1).

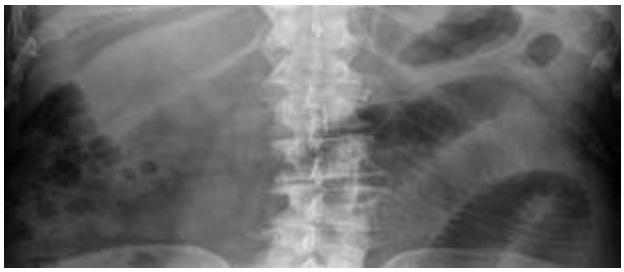

Figure 1: X-ray abdomen erect dilated Small bowel loops and multiple air fluid level.

Ultrasound of Abdomen and pelvis revealed Intestinal obstruction with Excessive bowel gas.

On the basis of clinical findings, X-ray and USG an emergency exploratory Laparotomy was performed. Intra-abdominal tumor was present over the serosal surface of ileum with torsion causing Volvulus of small bowel leading to obstruction (Figure 2).

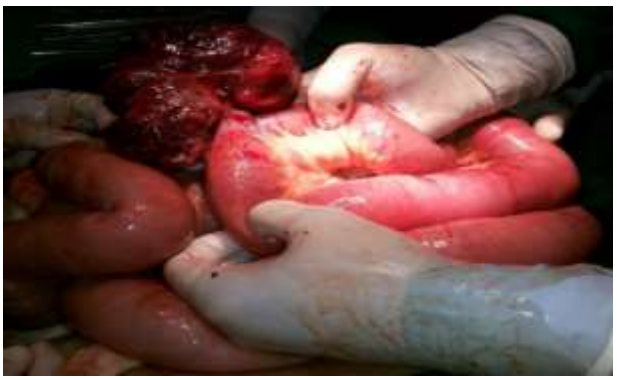

Figure 2: Intra-operative tumor attached to small intestine

The tumor along with portion from ileum was resected, obstruction released and end to end anastomosis was performed (Figure 3).

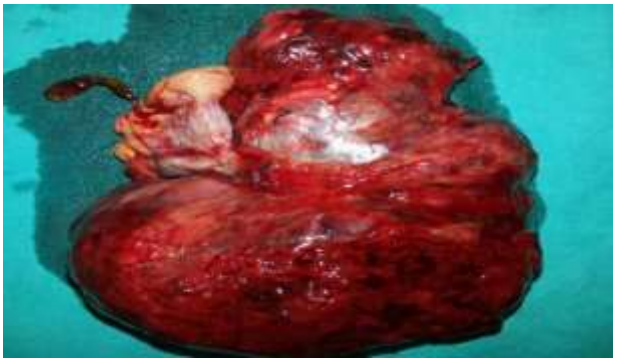

Figure 3: Resected tumor mass from small intestine.

Resected specimen was sent for histopathology. The specimen consisted of segment of small intestine with tumor mass with large area of necrosis. Mass was continuous with the serosal layer believed to be vascular tumor? Tumor cells were arranged mainly around thick wall vessels and were spindle to oval in shape with moderately pleomorphic. There were no eosinophilic hyaline bodies identified, Histopathology report was suggestive of Angiosarcoma of small bowel arising Serosa.

Immunohistochemistry revealed tumor cells were diffusely positive for CD 31 and vimentin, focally positive for CD 34. They were negative for cytokeratin and HMBE-45, S-100. This confirmed our diagnosis.

Patient recovered well post operatively and was discharged with instruction to follow-up in Department of Oncology.

DISCUSSION

Primary Angiosarcomas of the small intestine are rare high grade vascular neoplasms.[2] Diagnosis is challenging due to non-specific clinical, radiological and histopathological findings.[2] The clinical findings of intestinal Angiosarcoma are nonspecific. Symptoms include abdominal pain, nausea, vomiting, intestinal obstruction, anemia, gastrointestinal bleeding.[3,6] The pathogenesis is unclear; several etiologic factors have been suggested such as previous radiation, chronic lymphedema, exogenous toxins like thorotrast, vinyl chloride and arsenic, long-term peritoneal dialysis, intra-abdominal foreign body, visceral metastasis from Kaposi's sarcoma and familial syndromes such as neurofibromatosis Type 1, mutated BRCA1 and BRCA2, Mafucci syndrome and Klippel-Trenaunay syndrome.[1,3,4] Magnetic resonance imaging (MRI), computerized tomography (CT), abdominal x-rays and ultrasound can be used for diagnosis but all of them have limited diagnostic utility.[2,7] Conventional endoscopy is useful for tumors located in the stomach, duodenum and colon but not for jejunum and ileum. Although capsule endoscopy and barium can be used, they have limited diagnostic success.[8] Exploratory Laparotomy is the definitive diagnosis.[3] Angiosarcoma may be similar to a GISTs, leiomyosarcoma, metastatic melanoma, lymphoma, poorly differentiated carcinoma, neuroendocrine tumor, Crohns disease, and mesothelioma.[1,2,5] Microscopically anastomosing, delicate vascular channels lined by atypical endothelial cells mixed with solid sheets of spindled, epithelioid or undifferentiated cells are seen. Cystic degeneration, cellular necrosis may be found.[2] 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.[1,2]

CONCLUSION

Literature review has suggested exploratory laparotomy is the definitive for diagnosis and Immunohistochemistry is the best to confirm the diagnosis. Prognosis tends to depends after resection.

ACKNOWLEDGEMENTS

It is acknowledgements to whom so ever concerned this case study has been done on individual basis and has not been supported by any grant from any source.

Funding: None
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Aziz MT, Tabrez MO. Angiosarcoma of Small Intestine presenting with Intestinal Obstruction. Int Surg J 2016;3:956-8.