Lymphangioma of small bowel wall: A rare case report

Dr. Sanchit Jain[1], Dr. Mohit Jain[2], Dr. L.N. Meena[3], Dr. Somendra Bansal[4]

ABSTRACT

Introduction: Lymphangiomas are benign lesions of vascular origin that show lymphatic differentiation. They occur in many anatomic locations and may have a paediatric or adult clinical presentation. **Case presentation:** We herein describe the case of a 15-year old female who presented to the emergency of our hospital with abdominal distension and pain. At laparotomy, a giant multicystic tumor of jejunum was found which was leading to volvulus with gangrene of distal jejunum and whole of ileum. Histologically, the tumor was diagnosed as lymphangioma arising from the wall of jejunum. **Conclusion:** Intra-abdominal lymphangiomas are rare and most commonly are mesenteric in origin but in our case it was arising from the wall of jejunum which is a very rare phenomenon.

KEYWORDS: Benign, Lymphangioma, Multicystic Tumor, Obstruction

[1]Assistant Professor, [2,4]Senior Resident, [3]Associate Professor

[1]Department of General Surgery, RUHS College of Medical Sciences, Jaipur, Rajasthan, India

[2,3,4] Department of General Surgery, SMS Medical College, Jaipur, Rajasthan, India

[1]Corresponding Author mail: sanchit4088@gmail.com

INTRODUCTION

Lymphangiomas are malformations of lymphatic system. These tumors are a rare entity and constitute about 4% of all vascular tumors. These are mostly benign and can occur at any age and any part of the body, but 90% occur in children <2 yrs age and involve head and neck. Lymphangiomas rarely can occur intra-abdominally most commonly arising from mesentery especially small bowel mesentery.

CASE PRESENTATION

Our patient was a 15-year old female who presented with chief complaints of fever for 3 months, pain abdomen for 2 months, vomiting and abdominal distension for last 15 days and constipation for 5 days. There was no significant past or family history.

On general examination, there was hypotension, pallor and edema of hands and feet. On per abdominal examination there was abdominal distension which was more in lower abdomen. Tenderness, guarding and rigidity were present. And

bowel sounds were absent. Routine blood investigations showed Hemoglobin - 6.1 gm/dl and TLC (Total Leucocyte Count) - 20.91 X 1000/µl. Total serum proteins were decreased, Albumin:Globulin ratio was reversed (1:1.3) and serum calcium levels were decreased (7.6 mg/dl).

X-ray flat plate abdomen showed multiple air fluid levels suggestive of small bowel obstruction. On USG whole abdomen, a large collection with multiple septations was noted in the abdomen and pelvic cavity.

Based on history and investigations exploratory laparotomy was planned. On exploration, a large multicystic jejunal mass approximately 15 X 10 cm size was found (Figure 1) which was associated with volvulus of small bowel leading to gangrene of distal part of jejunum and whole of ileum (Figure 2).

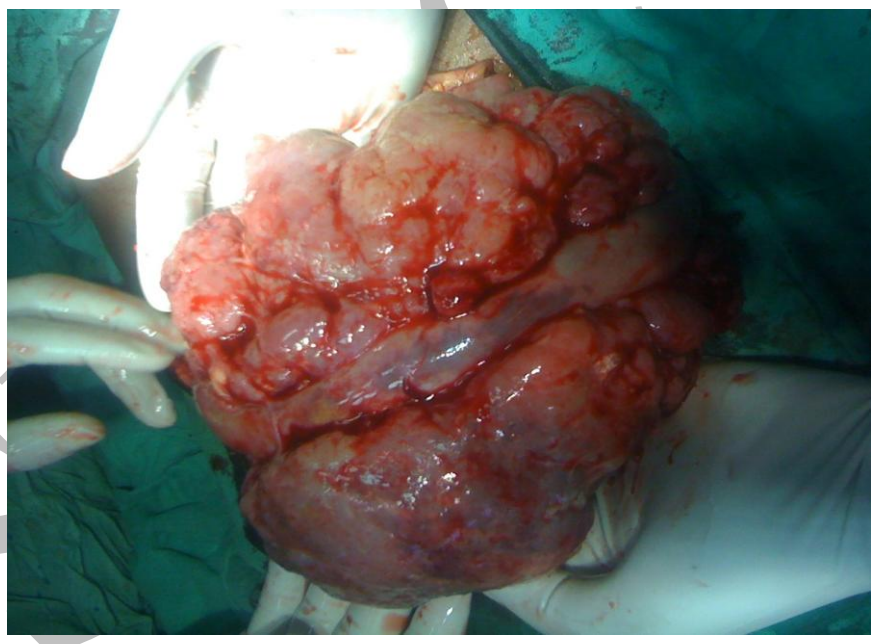

Figure 1 showing multi-cystic Jejunal mass

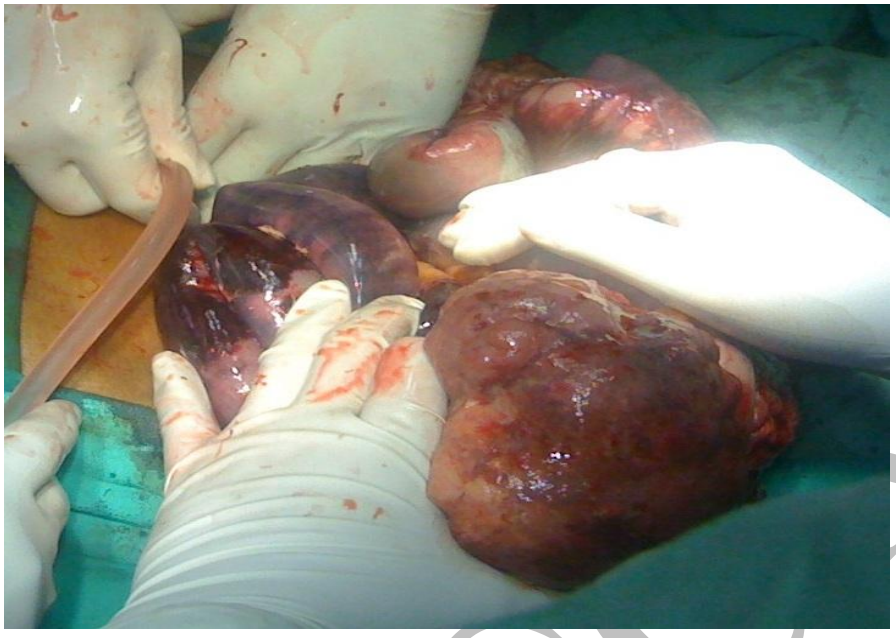

Figure 2 showing Jejunal mass with gangrenous jejunum and ileum loops.

After resection of the mass and gangrenous intestine about 1 feet of proximal jejunum was spared. An anastomosis was made between jejunum and ascending colon. After thorough peritoneal lavage abdomen was closed. The resected specimen was forwarded to the pathologist for examination. Postoperative period was uneventful and patient was discharged on postoperative day 13 with patient taking full oral diet. Patient is in regular follow up since last 1 year with no fresh complaints.

On histological evaluation, on gross examination, segment of small intestine measured 300 cm in length with congestion of external surface. Tumor measuring 16 X 13 X 8 cm was identified, 4 cm away from resected end of the specimen. On cutting, ***the tumor was found to be submucosal in location and was extending from mucosa to serosa.*** Cut surface of tumor showed honeycomb appearance with multiple tiny cyst of size ranging from 0.2 - 2.5 cm. Mucosa of the rest of the segment was found to be flattened and congested.

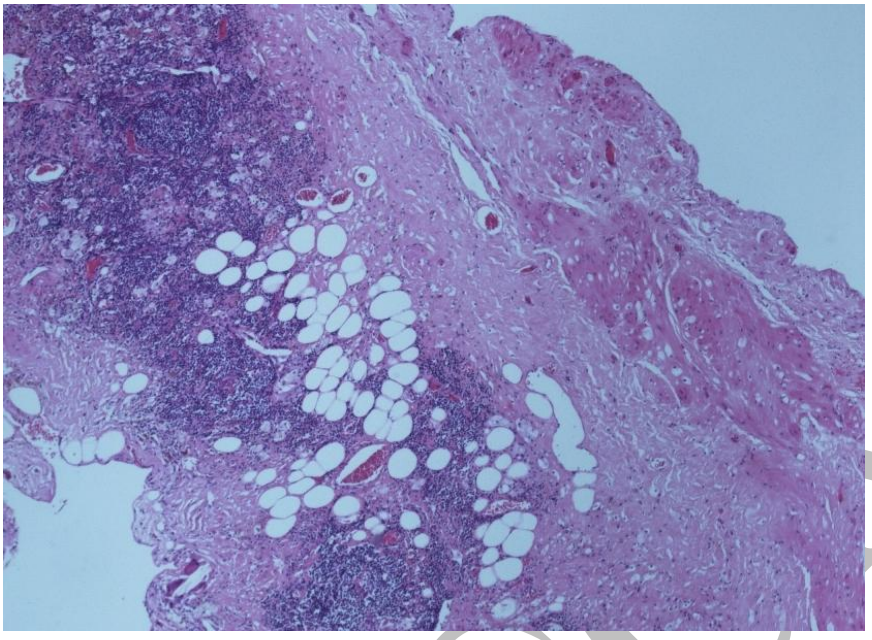

Figure 3 Histo-pathological Features.

Microscopic examination of the tumor revealed dilated lympho-vascular channels, edema and serosa showed infiltration by acute and chronic inflammatory cells along with thrombosis of mesenteric vessels (Figure 3). Both the proximal and distal resected ends were found to be unremarkable. 12 lymph nodes were resected out of which 4 showed hemorrhage, necrosis and infiltration by acute and chronic inflammatory cells. Examination of remaining lymph nodes was unremarkable. Based on gross and microscopic examination diagnosis was suggested to be ***lymphangioma of small intestine with the tumor being submucosal in location and involving the whole thickness of wall of small intestine.***

DISCUSSION

Lymphangiomas are benign lesions of vascular origin that show lymphatic differentiation. Most (95%) occur in the head and neck and axillary regions, the remaining being located in the mesentery, retro-peritoneum, abdominal viscera, lung, and mediastinum[1]. Traditionally, lymphangiomas are classified as simple, cavernous or cystic. Simple type is usually situated superficially in the skin and composed of small thin-walled lymphatic vessels. Cavernous type is composed of dilated lymphatic vessels and lymphoid stroma, and has a connection with various normal adjacent lymphatics while the cystic type consists of lymphatic spaces of various sizes that contain fascicles of

smooth muscle and collagen bundles, but has no connection with adjacent normal lymphatics[2].

Intra-abdominally lymphangiomas arise most commonly from the mesentery; it is very rare to find these lesions arising from wall of the bowel. Infrequently they may be found in the esophagus, stomach, small intestine and colo-rectum.

The aetiology is uncertain but, the 'Blind Sac' hypothesis, postulates that a lack of lymphatic connections causes them to proliferate and dilate[3]. Intramural lymphatic obstruction, disturbed endothelial permeability, inflammation, congenital absence of lymphatics and aging of the bowel wall have also been suggested as causes for the development of intestinal lymphangiomas.

These tumors are usually asymptomatic. Abdominal pain and distention are the most common symptoms, but the clinical presentation varies[4]. Although benign in nature, mesenteric lymphangiomas may cause significant morbidity or mortality due to their large size and critical location, when they compress the adjacent structures. The most common mode of acute presentation in children is small bowel obstruction, sometimes associated with volvulus and gangrene[5].

Most intestinal lymphangiomas are mural masses discovered incidentally at endoscopy or on radiologic studies performed for other reasons[6]. The imaging studies are the revealing diagnostic tool; the abdominal ultrasonography being the procedure of choice for establishing the diagnosis, even during the antenatal stage[7]. It is also the test of choice for postoperative follow up. Preoperative imaging may provide some clinical information; however, the diagnosis can only be conclusively confirmed after surgery. A CT scan or MRI is the best means of identifying the solid components of the lymphangioma and providing information needed for surgical planning[8].

The optimal treatment is radical excision, even when asymptomatic. However, mesenteric lymphangiomas may cause complications such as infiltration of the intestine, or involvement of the main branch of mesenteric arteries or adjacent organs that necessitate segmental resection of the intestine. Sometimes radical resection might be technically impossible[9].

CONCLUSION

Intra-abdominal lymphangiomas are rare benign lesions. The clinical symptomatology is not specific. The diagnosis is usually suggested by the

imaging studies, but it still requires histopathologic confirmation. The treatment of choice is surgical and consists of a full resection of the lesion. Although very rare they can present as acute abdomen requiring an emergent surgery.

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