Intra-Abdominal Lymphangioma
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1. Abstract
Lymphangiomas are rare slowly growing benign tumors derived from lymphatic vessels. They occur in various anatomical locations of body like head, neck and abdomen. Abdominal cystic Lymphangiomas are rare intra-abdominal masses affecting the infants and young children, these may be detected incidentally on radiological investigations or present with abdominal distension, vague abdominal pain and life threatening emergencies like bowel obstruction and torsion of cystic masses. We are discussing the case of an intra- abdominal lymphangioma in a 5 years old girl who presented to the pediatrics outpatient department with complaints of abdominal distention and pain. Her ultrasound and CT scan abdomen with contrast were done. She later underwent surgery and histopathology was sent.

2. Introduction
Abdominal cystic lymphangiomas are very rare congenital benign tumor arising from the lymphatic vessels [1]. These usually occur in pediatric age groups and may be asymptomatic or present with nonspecific symptoms like vague abdominal pain and distention [2, 3]. There are two types of lymphangiomas includes congenital and acquired. The exact cause of these lymphangiomas is unknown but a well-known theory, “blind sac” hypothesis stated that lack of lymphatic connections causes them to proliferate and dilate. They can be developed in any organ of the body, however 95% occur in neck and axillary regions, rest of the 5% occur in the mesentery, retro peritoneum, abdominal viscera, lungs and mediastinum [4]. There are various complications associated with intra- abdominal lymphangiomas including torsion, infection, bleeding and rupture of cystic masses in lymphangiomas. The definite management of lymphangiomas is surgery, although it can be challenging in some cases. Recurrence can be seen in some cases, this is depending on the location, size, and complexity of lesions. Spontaneous regression is less common but can be seen in recurrent lymphangiomas [2]. CT scan and ultrasound are commonly used for the diagnosis of lymphangiomas. The exact and appropriate diagnosis is usually made after surgery on histopathological evaluation 4. Here we are discussing a biopsy proven case of intra-lymphangioma in a 5 years old girl and the characteristic radiological features on CT scan and ultrasound.

3. Case Report
A 5 years old girl with no known co-morbid and insignificant birth history came to the pediatric outpatient department with complaints of abdominal pain and distension. Pediatrician performed an abdominal examination and revealed soft mass in abdomen extending from epigastric region up to the pelvis with mild tenderness on palpation. The initial baseline laboratory investigations were noncontributory. Her past medical and surgical history were unremarkable. She was sent to the radiology department for ultrasound abdomen with query of abdominal mass and intestinal obstruction. Her ultrasound abdomen scan with contrast were done. She later underwent surgery and histopathology was sent.

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the epigastric region up to the pelvis, bulk of the mass is seen within the pelvis and in mid abdomen. There is no evidence of calcification, fat, or solid component. The masses are abutting and displacing the adjacent small and large bowel loops without evidence of intra-luminal extension. There is no evidence of bowel obstruction or dilation (figure 2-4). This was reported as an intra-Abdominal Lymphangioma. She was taken to the operation theater after baseline investigations and general anesthesia fitness. Exploratory laparotomy was performed. Her operative findings showed a large multi-loculated cystic mass extending from epigastric region up to the pelvis adherent to the mesentery. There was no evidence involvement of small or large bowel loops. It was full of fluid which was aspirated to get access to the peritoneum and complete excision of tumor was performed. (Not shown here). The sample was sent for the histopathological examination. Her histopathology shows numerous irregular cystic dilated channels lined by flat attenuated lining. Scattered lymphoid aggregates and mild to moderate inflammatory infiltrates are present in the stroma. No atypia is noted. The cells lining the cystic spaces are positive for immune-histochemical stain CD31 and negative for Cytokeratin and Calretinin. The diagnosis is of a benign lymphatic malformation compatible with lymphangioma (figure 5-7).

Patient was followed in outpatient department of paediatric in our hospital, with no any symptoms, and her post-operative ultrasonography showed no evidence of any mass lesion.

Figure 1: Grey scale ultrasound images of abdomen show multi-loculated cystic mass

Figure 2: Contrast enhanced axial CT scan abdomen show multi-loculated cystic masses extending from epigastric region up to the pelvis.

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Figure 3: Contrast enhanced sagittal CT scan abdomen images show cystic masses in abdomen and pelvis

Figure 4: Contrast enhanced coronal CT scan abdomen images show cystic masses in abdomen and pelvis

Figure 5: Low power view of multiple cystic spaces
4. Discussion

Lymphangiomas are benign tumors of lymphatic origin first described in 1828 by Radenbacker [2]. Incidence of these tumors is 1:20,000 to 1:250,000 with mean age of presentation being 2 years and a male predominance [2, 3]. Although the exact etiology is still unknown according to Godart’s theory these tumors arise due to anomalous sequestration of lymphoid tissues that fail to communicate with the flow channels or due to obstruction in the passage of flow of lymphatic resulting in blind ended cystic spaces containing lymphoid secretions [2, 3, 5]. Secondary causes include inflammatory process, infections, lymphatic obstruction, abdominal trauma, surgery and radiation therapy may be associated with the development of such tumors [1, 4].

Clinical features of these tumors are non-specific and often misleading however the various symptoms that patients may present with include nausea, vomiting, weight loss, pain and other symptoms of bowel obstruction [1-7]. Our patient also presented with complaints of abdominal pain and distension. On examination in approximately 58% of these patients mostly have a painless mass that is soft and mobile 3, as seen in our case.

Imaging plays an important role in the evaluation and diagnosis of these patients with ultrasonography being the primary investigation, often sufficient to delineate the basic features of the palpable masses their location, size, content and relation to adjacent viscera’s. However, a contrast enhanced CT scan is the imaging modality of choice. These appear as homogenous cystic masses with wall enhancement, CT scan can more clearly demonstrate the location, extension and degree of vascular involvement [1-7]. In our case there was a large non enhancing multi-loculated cystic masses extending from the epigastric region up to the pelvis, bulk of the mass was seen within the pelvis and in mid abdomen. There was no evidence of calcification, fat, or solid component. The mass was abutting and displacing the adjacent small and large bowel loops without evidence of intra-luminal extension. There was no evidence of bowel obstruction or dilation.

The ultimate diagnosis is dependent on the histopathology. Surgical resection of the mesenteric lymphangiomas is the ultimate treatment in order to avoid the complications that may occur due to the tumor [1-7].
5. Conclusion
Mesenteric lymphangiomas are a rare presentation, but may result in serious complications including torsion, infection, bleeding and rupture of cystic masses. It is therefore important for the clinicians, radiologists and the histo-pathologist to be aware of this rarely occurring tumor and its basic features.

6. Ethical Review
This case report was written and images of patient’s CT scan, Ultrasoundography, and Histopathology were added after taking consent from patient’s parents and patient’s identity is not shown here and will be kept confidential.

References