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Case report of Giant Mesenteric Macrocystic Lymphatic Malformation in an adult.

Radiology

KEYWORDS: Lymphatic malformation, Abdomen, mesentery.

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ABSTRACT

Lymphatic malformations are subset of congenital vascular malformations with lymphatic differentiation. The most common locations of lymphatic malformations are neck and axilla, they can also occur at several other locations including abdomen. The abdominal lymphatic malformations are rare and accounts for approximately 5% of all lymphatic malformations. Clinical presentation includes abdominal lump and vague abdominal discomfort. They can present with broad spectrum of radiological manifestation ranging from small simple cysts to large multi-cystic masses. Differentiating cystic lymphangiomas from other cystic lesions by imaging studies alone is often very difficult and surgery is frequently required for definitive diagnosis. In the present case report, we describe an interesting and rare case of a mesenteric lymphangioma presenting in an adult patient, which was removed surgically and was histo-pathologically proven to be a lympho-vascular malformation.

Introduction:

Lymphatic malformations are a subset of congenital vascular malformations which are slow-growing benign lesions characterized by proliferating lymphatics (Lowe 2012, Hyodoh 2005). The abdominal lymphatic malformations are rare and accounts for approximately 5% of all lymphatic malformations (Lal 2016). Within the abdominal cavity, they can occur in mesentery, retroperitoneum, solid organs (liver, spleen and pancreas) and intestinal tract (Lal 2016). The majority of abdominal lymphatic malformations are diagnosed in children. Rarely, they present in adults. Mesenteric lymphangiomas can present as a palpable abdominal mass or symptoms related to mass effect or secondary complications including intra-cystic hemorrhage, rupture, volvulus or infection (Lal 2016). Surgery remains the treatment of choice. The less invasive forms of treatment including per cutaneous sclerotherapy can also be done, but these are found to have greater recurrence rate (Allen 2006).

Case Report:

A 16-year-old male presented with vague abdominal pain for past 3 months. Ultrasound of patient revealed multi-loculated cystic lesion and few well defined echogenic foci with post-acoustic shadow suggestive of phleboliths.

Contrast-enhanced computed tomography (CECT) of abdomen and pelvis was done after adequate preparation. The scan revealed a large, multi-loculated, non-enhancing cystic lesion with multiple thin enhancing septae in right half of the abdomen (sub-hepatic, right lumbar, right iliac fossa and pelvic region and umbilical region). Multiple calcific foci were seen in lesion, suggestive of phleboliths. The lesion was encasing distal ileal loops and mesenteric vessels with associated displacement of other small and large bowel loops to left side of abdomen. There was no ascites, enlarged nodes or bowel obstruction. On basis of these findings, differentials of ? Lympho-vascular malformation. / ?? Mucinous carcinoma of bowel were given.

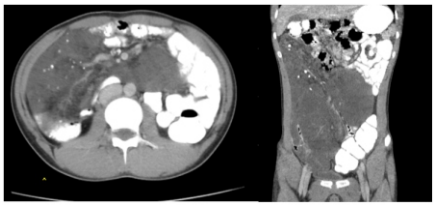


Figure 1 and 2: Computerized Tomography (CT) abdomen axial and coronal images show multi-loculated cystic lesion in mesentery with encased small bowel loops. Few hyperdense calcific foci are seen in lesion, suggestive of phleboliths.

Exploratory laparotomy with resection of mesenteric cystic mass and adjacent ileal loops was done. Histopathological report confirmed diagnosis of Cystic mesenteric lymphangioma.



Figure 3: Histopathological examination: Multiple cystic lesions of varying size and shape which are lined by endothelium and contain eosinophilic material in lumen with interspersed muscle fibers are seen. The cystic spaces invade serosa, muscularis propria, submucosa and reaches upto mucosal. Discussion: The International Society for Study of Vascular Anomalies (ISSVA) has divided vascular anomalies based on endothelial cell turnover and cellular diffentiation into two biological categories: (1) Vasoproliferative/ vascular neoplasms and (2) Vascular malformations (Lowe 2012). Lymphangiomas are major group of vascular malformations, which result from failure in evolutionary development of the vascular system, including lymphatic and/or arteries and veins. These lymphatic tumors are divided in: 1) simple, with capillary lymphatic channels; 2) cavernous, with dilated lymphatics and capsule; and 3) macrocystic malformations, clinically termed "cystic hygroma" (Reis et al 2014). Lymphatic malformations can affect any area of the body except the brain, most common sites are the head and neck while other locations include the mesentery, retroperitoneum, abdominal viscera, lung and mediastinum (Nosher 2014). Patient is usually an infant or child, sometimes can be an adolescent or adult (Lal 2016). When they get complicated (secondary infection/ intralesional haemorrhage), they are detected by pain, tenderness or discoloration over the swelling (Allen 2006).

Plain X-ray is of limited use as it can only detect soft tissue swelling over the part or sometimes punctate calcifications due to the phleboliths. Ultrasound is suitable modality to depict the exact location, uni or multilocularity and cystic character of lesion. Colour Doppler may demonstrate peripheral/ intraseptal vascularity. The presence of hemorrhage or infection in the lesion gives more complex appearances like multiple thin septations, coarse/fine echoes,

echogenic debris and thick appreciable wall (Hyodoh 2005).

Contrast enhanced computed tomography is investigation of choice which shows exact extent, phasic enhancement and wash-out of lesion, any associated secondary complication and visceral involvement. Sometimes lesion can show negative attenuation values due to presence of fatty chyle or air (secondary to interventions) (Lal 2016).

Most symptomatic cases require treatment, due to mass effect of lesion on adjacent structure or due to the complications (haemorrhage/infection). Treatment of choice is direct surgical removal. However, recurrences are common due to insinuation of lesion in adjacent structures and thus leading to difficulty in complete removal. Thus, imaging is important demonstrating accurate anatomic localization, character and definition of the lesion for pre-operative planning and to access recurrent lesions.

Differentiating lymphangiomas from enteric duplications, mesothelial cysts, and pseudocysts may be difficult because the imaging features of these lesions overlap (Levy 2004).Conclusion: Giant macrocystic mesenteric lympho-vascular malformation presenting in an adult patient is a rare entity and can present with varied imaging features and surgical emergencies requiring early and accurate diagnosis. Multiplanar CT plays an important role in anatomic localization, characterization and extension of the lesion for preoperative planning and facilitating complete surgical excision.

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