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Mesenteric Lymphangioma with Counterclockwise Barberpole Sign-A Case Report

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Abstract Case Report

Lymphangioma is a congenital malformation of lymphatic vessel. Commonly lymphangiomas are seen in the neck and axilla (95%). Rarely (5%) they occur in the mediastinum and abdomen in the mesentery and retroperitoneum. Mesenteric lymphangioma may be associated with counter clockwise barber pole sign on MDCT. It may be associated with volvulus and rarely associated with malrotation of the gut or normal variant. We report a case of mesenteric lymphangioma in a 3 year old female patient with counterclockwise barberpole sign with partial malrotation of small bowel.

Keywords: Mesenteric lymphangioma, counterclockwise, barberpole sign, partial malrotation.

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INTRODUCTION

Lymphangioma is a congenital malformation of lymphatic vessel. Commonly lymphangiomas are seen in the neck and axilla (95%). Rarely (5%) they occur in the mediastinum and abdomen in the mesentery and retroperitoneum.1The incidence of lymphangiomas is 1 per 20,000 paediatric population admitted in the hospitals.2Mesenteric cystic Lymphangioma (MCL) become symptomatic in 90% cases before 2 years of age.

Mesenteric lymphangioma may be associated with counter clockwise barber pole sign on MDCT. It may be associated with volvulus and rarely associated with malrotation of the gut or normal variant [3].

CASE REPORT

3 year old female patient presented with nonbilious vomiting for 7 days, loose stools followed by constipation since 3 days, and pain in peri-umbilical region since 4 days.

Ultrasound (USG) of the abdomen (figures 1,2)showed a well-defined thinned wall anechoic cystic lesion of size 75(cranio-caudal)X45(anteroposterior)X105(transverse) mm in umbilical region beneath the anterior abdominal wall in the midline, extending on either side of the midline and extending

inferiorly in hypogastric region. It showed septations. Branches of superior mesenteric artery (SMA) and superior mesenteric vein (SMV) were seen traversing through the lesion. Bowel loops were compressed and displaced laterally and posteriorly by the cyst.

Computed tomography (CT) abdomen and pelvis (figures 3-7) showed a well-defined thin walled cystic lesion of size 75(cranio-caudal) X45 (anteroposterior) X105 (transverse) mm in umbilical regions, and extending inferiorly in hypogastric region. It was seen just beneath the anterior abdominal wall in midline, extending on either side of the midline and showed septations. Altered relations were noted in SMA and SMV distal to their origin. SMV showed counter clockwise rotation with relation to SMAsuggestive of counterclockwise barber pole sign seen in partial malrotation or as a normal variant (figures 3-5). SMA and SMV branches were seen traversing through lesion suggestive of mesenteric origin (figures 6, 7). The cyst was compressing and displacing bowel loops laterally and posteriorly. Rest of the abdomen was normal.

Diagnosis of mesenteric lymphangioma with counter clockwise barber pole sign seen in partial malrotation was given. Patient was operated elsewhere and the diagnosis was confirmed.

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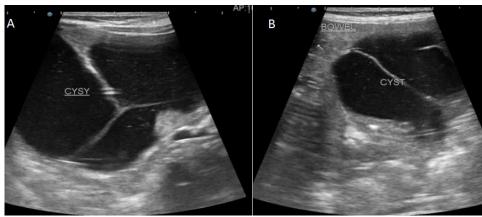
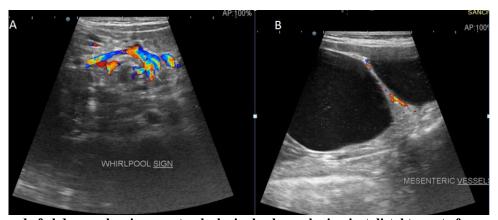


Fig-1: Ultrasound of abdomen showing multilocular cyst in mesentery showing multiple thin septations and fine internal echoes



 $\label{eq:Fig-2: Ultrasound of abdomen showing counterclockwise barber pole sign just distal to root of mesentery (A), and \\vascularity within septations in cyst(B)$

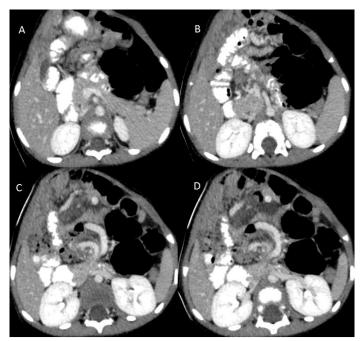


Fig-3 (A-D)-Contrast enhanced CT abdomen showing counterclockwise barber pole sign distal to root of mesentery (SMV showing counterclockwise rotation with relation to SMA)

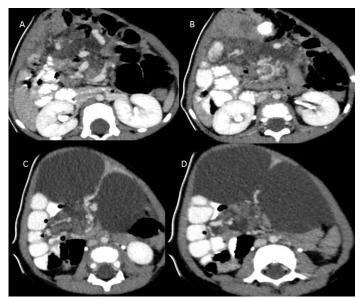


Fig-4-(A-D)- Axial Contrast enhanced CT abdomen showing multilocular cyst in mesentery in umbilical region showing counterclockwise barber pole sign distal to root of mesentery (A-B) and enhancing mesenteric vessels in septations (C-D)

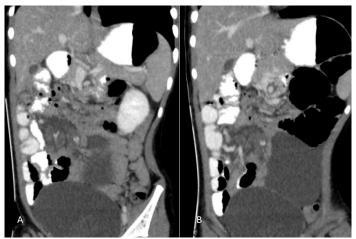


Fig-5: (A-B)- Coronal Contrast enhanced CT abdomen showing multilocular cyst in mesentery in umbilical region showing counterclockwise barber pole sign distal to root of mesentery

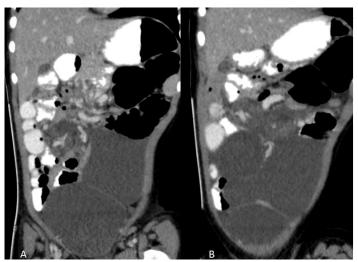


Fig-6 (A-B): Coronal Contrast enhanced CT abdomen showing multilocular cyst in mesentery in umbilical region showing enhancing mesenteric vessels in septations

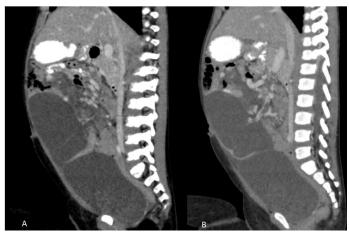


Fig-7(A-B): Sagittal Contrast enhanced CT abdomen showing multilocular cyst in mesentery in umbilical region showing enhancing mesenteric vessels in septations.

DISCUSSION

65% of MCL are congenital [4]. They are usually present as palpable abdominal mass and abdominal distension [5]. It can cause intestinal obstruction due to compression and traction.⁶ Rarely, it can cause closed loop obstruction of the involved bowel loops by causing rotation with resultant volvulus of connected mesentery and small bowel.7 Both USG and multi detector computed tomography(MDCT) can diagnose MCL. USG is superior to CT in assessment of cystic lesion as it can detect wall thickness, presence of septa, presence of echogenic contents and the presence of infection and internal haemorrhage. On MDCT, MCL is seen as a multilocular cystic mass with a homogeneous fluid component of low attenuation values. Rarely negative attenuation values can be seen due to presence of chyle. Calcification is uncommon [8]. Due to multiplanar reformation, MDCT can diagnose the lesion and assess complications like volvulus and intestinal obstructions. As compared to retroperitoneal lymphangiomas, MCL's have a tendency to present with complication as they are mobile. The complications occurring with MCL are volvulus, intestinal obstruction and bowel gangrene, intra-abdominal bleeding, infection and infarction of the cyst. Its pathogenesis is unclear. Interrupted communication of draining the lymphatic system can cause lymphatic congestion . However; this theory cannot explain the occurrence of MCL in adults. Adult manifestation occurs due to delayed proliferation of congenital acquired lymphoid nests after stimuli like local trauma or respiratory infection [9]. MCL may be primarily or secondarily associated with malrotation of gut [10]. Buranasiri et al. first described counter clockwise barber pole sign on angiography. It is an angiographic sign, in rare case of malrotation with counter clockwise volvulus [11]. However, this sign may not always be associated with volvulus and can be association of malrotation of the gut or normal variant [3]. Normally, SMV is anterior and on right side of SMA. Several centimeters inferior to SMA origin the

SMV is wrapped counterclockwise around SMA forming the counterclockwise barber pole sign.

The third part of the duodenum (D3) was seen passing anteriorly while the fourth part (D4) was coursing towards right side -suggestive of partial malrotation. Counter clockwise rotation of SMV around SMA by 180^{0} to 200^{-0} noted 1.8 cm distal to the origin of SMA without any twisting of mesentery and small bowel. There were no signs of proximal small bowel obstruction; hence the diagnosis of MCL with counter clockwise barber pole sign with partial malrotation of the gut was made.

Conclusion

Demonstration of counterclockwise barber pole sign on MDCT in mesenteric lymphangioma can be associated with volvulus or malrotation and rarely as a normal variant. The radiologist should always rule out volvulus and malrotation if this sign is present on MDCT before labelling it as a normal variant.

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