



A PUZZLING PRESENTATION: ABDOMINAL LYMPHANGIOMA MASQUERADING AS SMALL BOWEL MASS

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BACKGROUND

- Acute abdomen is a very common presentation in A&E with varied pathology, and CT is usually the first line investigation used to evaluate it.
- Abdominal lymphangiomas are rare cause of acute abdomen. They are rare benign entities found in mesentery and can have very varied clinical presentation.
- Imaging findings of lymphangiomas are usually typical but sometimes can mimic tumour, as in our case, especially when complicated by superimposed inflammation.
- These cases are difficult to diagnose preoperatively. Surgery is often necessary and final diagnosis is usually made on histopathology.
- Radiologists and surgeons need to be aware of unusual presentation of this pathology and how to manage it.

CLINICAL PRESENTATION

- 53 year-old male presented acutely to A&E with 24-hour history of non-specific abdominal pain. No associated bowel complaints were present. On examination, there was tenderness to palpation in lower abdomen.
- Past medical history was non-contributory .
- Blood investigations revealed raised inflammatory markers (WCC – 16.7, CRP – 211).



IMAGING

 CT Abdomen-Pelvis showed wall thickening of ileum with adjacent lymph nodes, free fluid and mass-like mesenteric stranding. No evidence of small bowel obstruction. Differentials included inflammatory etiology with possibility of underlying small bowel lymphoma or primary.





Figure 1 and 2 : CT at first presentation, axial and coronal images showing thickened bowel loop with adjacent lymph nodes, mass-like mesenteric stranding and fluid (arrows).

- Patient's symptoms and inflammatory markers settled with antibiotics (WCC 7.1, CRP 66) and patient was discharged after 4 days.
- CT after 6 weeks showed interval decrease of inflammation and mesenteric lymph nodes but persistent bowel wall thickening and adjacent mass-like mesenteric stranding.
 Differentials again included inflammatory etiology or bowel tumor.





Figure 3 and 4 : Followup CT after 6 weeks, showing resolution of inflammation but persistence of bowel wall thickening and mass-like mesenteric stranding (arrows).

Figure 4

TREATMENT

• Given persistent abnormality and difficult access with colonoscopy, surgical management was opted after discussion in MDT. Patient underwent elective laparotomy, where bowel mass involving ileum was resected and sent for histopathology.

PATHOLOGY

 Gross pathology specimen showed normal appearing bowel mucosa and honey-comb lesion with hemorrhage in mesentery. Microscopy revealed large number of thin-walled dilated vessels (cystic spaces) with accompanying fat necrosis and hemosiderin deposit – appearances consistent with lymphatic malformation (lymphangioma). The lining of dilated vessels stained positive for CD31 endothelial immunomarker which further confirmed the diagnosis.

FINAL DIAGNOSIS

ABDOMINAL LYMPHANGIOMA of mesentery tethered to ileum.



Figure 5: Macroscopic image of resected small bowel with lesion in mesentery. Figure 6: Sliced specimen showing honey-comb lesion with hemorrhage in mesentery.



Figure 7 and 8: Low power microscopic image showing normal bowel wall with thin-walled dilated vessels (arrows) on mesenteric side. **Figure 9**: High power microscopic image showing lining of dilated vessels staining positive (brown) for CD31 endothelial immunomarker.

DISCUSSION

- Abdominal location is rare for lymphangiomas and accounts for 5% of all cases. Can arise from mesentery, omentum, GI tract and retroperitoneum.
- Mesentery most common site in abdomen. Clinical presentation extremely variable, can also present as acute abdomen.
- Mesenteric lymphangiomas can produce serious complications including hemorrhage, intestinal ischemia, obstruction and volvulus.
- Usually appear as multilocular cystic masses on imaging. Less commonly, may appear as predominantly solid. Can mimic tumours, especially when complicated by superimposed inflammation.
- On CT, fluid component usually homogeneous with low attenuation values. Less commonly, can show negative HU values due to presence of chylous fluid.
- Mesothelial cysts, enteric duplication cysts and pseudocysts mimic closely.
- Treatment is recommended in all (even asymptomatic) cases with laparoscopic resection, as incidence of complications tends to increase with time as lesion size increases. Complete excision is best option to prevent recurrence.



Figure 10: Typical imaging appearance of lymphangioma as a multiloculated cystic mass usually in mesentery or retroperitoneum¹.

LEARNING POINTS

- Abdominal Lymphangiomas are a rare cause of acute abdomen and can have varied clinical presentation and imaging appearances.
- Although typical imaging appearance is of multilocular cystic mass in mesentery, it can masquerade as small bowel tumour especially when associated with superimposed reactive and inflammatory changes, which often mask the true lymphatic/cystic nature of the tumour.
- In such cases, diagnosis is confirmed on histopathology by typical microscopic features and staining for immunomarkers.
- Awareness of this entity is important among radiologists, surgeons and pathologists to guide the management and avoid overtreatment.

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