

# Radiological Features of Extensive Hemangiolympangioma of the Small Bowel Mesentery with Histopathology Findings: A Case Report and Literature Review

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## Abstract

Hemangiolympangioma is a rare malformation of the vascular and lymphatic system affecting different parts of the human body. Hemangiolympangioma of the small bowel mesentery is an extremely rare entity with only 3 cases reported in the English literature, to our knowledge. We report the case of a 23-year-old female who presented with generalised colicky abdominal pain. The CT findings were initially thought to represent possible secondary mesenteric and peritoneal metastatic deposits. However, no primary tumour was identified. Pelvis MRI was then performed and favoured the diagnosis of extensive mesenteric lymphangiomatosis. The final diagnosis was confirmed histopathologically as hemangiolympangioma of the small bowel mesentery. This case report will cover several imaging features that can help differentiate hemangiolympangioma of the small bowel mesentery from metastatic deposits in the peritoneum and aid radiologists/clinicians reach a preoperative diagnosis with a level of certainty.

**Keywords:** Hemangiolympangioma, Hemolympangioma, Small bowel, Mesenteric tumour, Radiology.

## Introduction

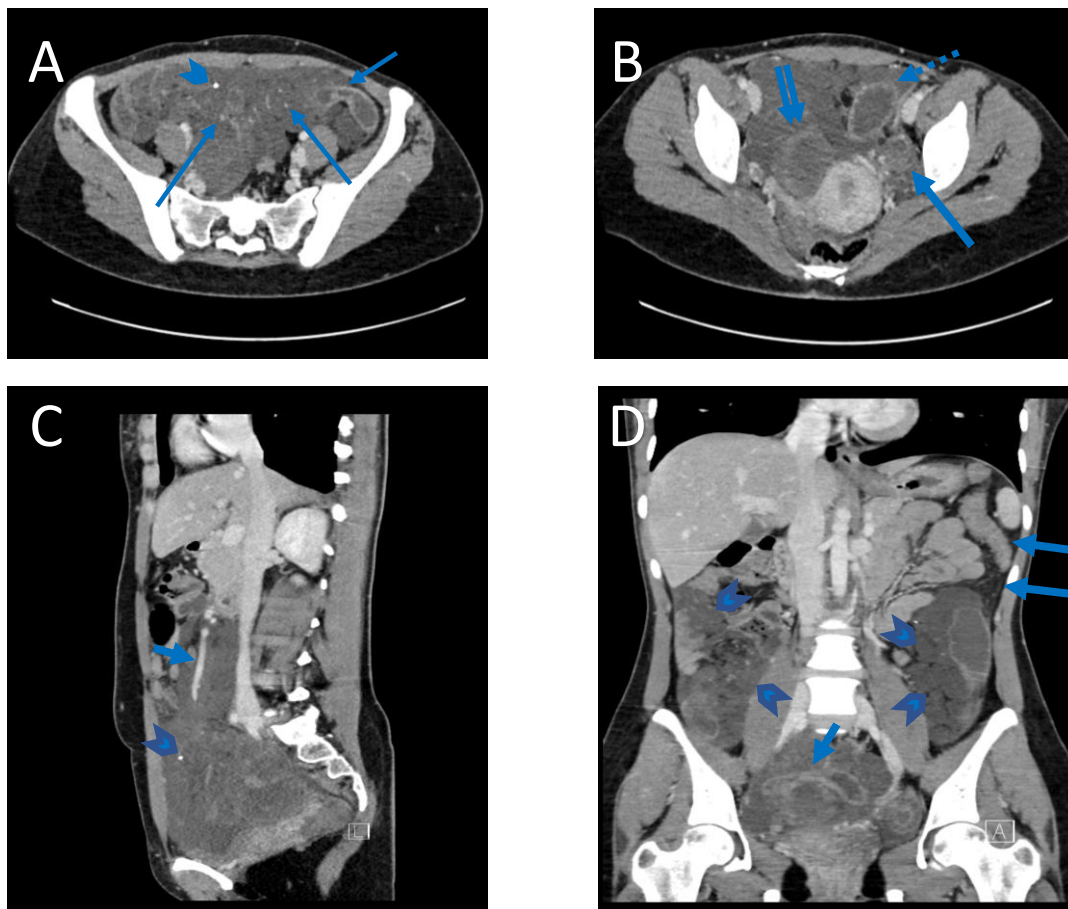
Hemangiolympangioma is a rare type of venolympatic vascular malformation<sup>1</sup> showing a mixture of blood vessels and lymphatics.<sup>14</sup> Hemangiolympangioma most commonly presents in infants and young children on the body surface, with extremely low incidence in adulthood.<sup>8</sup> In adults, most hemangiolympangiomas occur in the head and neck while only a few cases have been reported to occur in other parts of the body including the pancreas,<sup>2</sup> spleen,<sup>3</sup> small bowel,<sup>4</sup> rectum,<sup>5</sup> chest wall<sup>6</sup> and extremities<sup>7</sup> etc. To date, 9 small bowel hemangiolympangioma cases have been reported, and only 3 cases of small bowel mesenteric hemangiolympangioma have been reported in the English literature.<sup>14</sup> Here, we report a case of small bowel mesentery hemangiolympangioma with characteristic imaging features that can help differentiate it from other more sinister conditions namely malignancy and metastasis.

## Case Report

A 23-year-old female presented to the hospital with 5 days history of intermittent generalised colicky abdominal pain, constipation and vomiting. She had a prior surgical history of laparoscopic appendectomy done 6 years earlier, Abdominal examination showed tense abdomen with diffuse tenderness. Laboratory investigations showed Haemoglobin (Hb) 10.8g/dL, ESR 58 and CRP 83. Other blood tests, tumour markers (B-HCG, CA 15-

3, CA 19-9, CA 125, CEA, AFP) and microbiological tests (TB, HIV, HepB/C) were normal. Initial bedside B-mode ultrasonography (US) showed cysts anterior to the uterus.

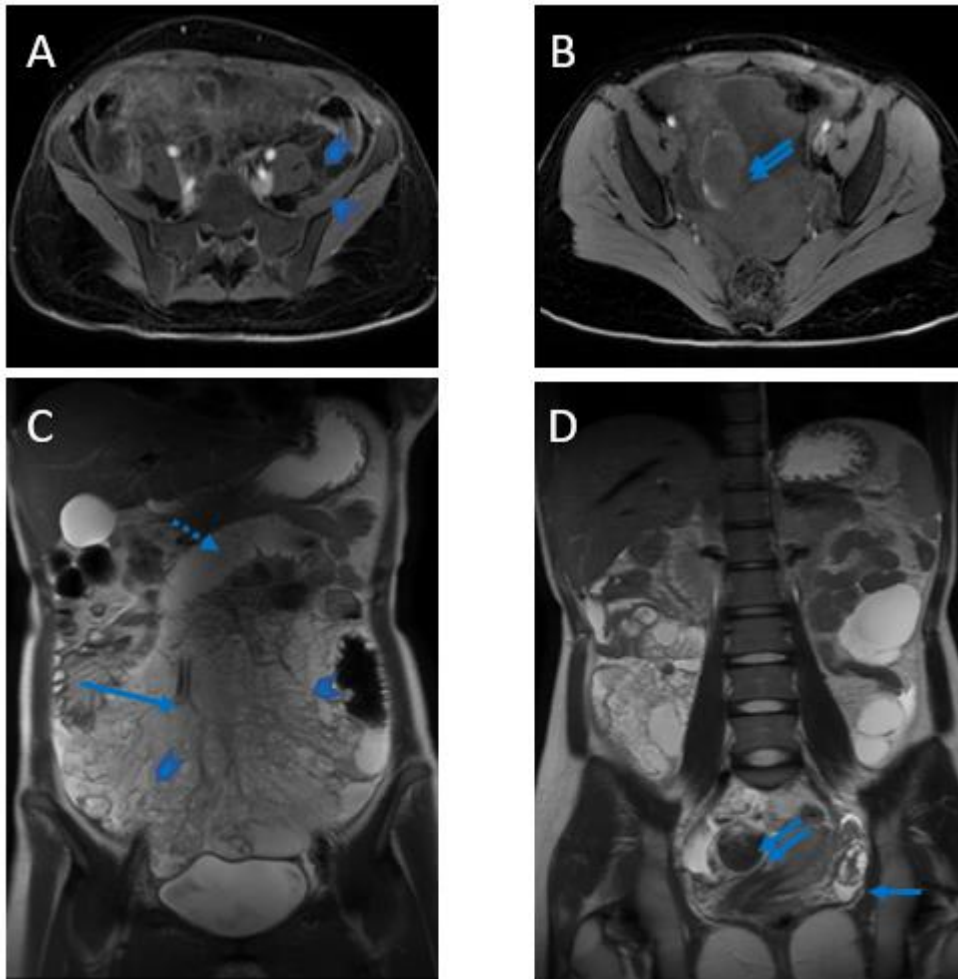
Contrast-enhanced abdominopelvic computed tomography (CT) demonstrated extensive mesenteric involvement by soft tissue densities and multiple wide spread variable sized fluid attenuating cystic lesions, largest in the left upper quadrant measuring 5.0 x 4.6 cm in size with adjacent nodular calcifications. The lesions were engulfing the mesenteric vasculature which were attenuated but patent. There were alternating areas of small bowel narrowing and dilatation with mural hyperenhancement within the engulfed segments. Mild segmental dilatation of the proximal jejunal loops was noted reaching up to 3.3 cm in diameter with associated mild diffuse wall thickening and enhancement of the proximal non-dilated jejunal loops reaching up to 0.6 cm in thickness. A few scattered calcific foci were identified within the cystic lesions. There were multiple prominent mesenteric lymph nodes, largest measuring 0.8 cm in short axis (Fig. 1). The CT findings were initially thought to represent possible secondary mesenteric and peritoneal metastatic deposits. However, no primary tumour was identified.



**Figure 1:** Computed Tomography (CT) with IV contrast in the portal venous phase images. (A) Axial image showing soft tissue densities engulfing the mesenteric vascular branches which are attenuated but patent (long solid arrows), with adjacent nodular calcifications (arrow head) and small bowel narrowing (short solid arrow). (B) Axial image showing haemorrhagic cyst (double arrow) with dilated small bowel segment proximal to the haemorrhagic cyst (dotted arrow) and normal left ovary (long solid arrow). (C) Sagittal images showing the distribution of the abnormality within the lower half of the abdomen with sparing of the upper half with encasement of the patent SMV without luminal narrowing (short solid arrow) and calcific focus (arrow head). (D) Coronal image showing distribution of the cystic changes along the mesentery (arrow heads) with relative sparing of the peritoneal regions (long solid arrows) and compression of ileal loop resulting in partial low grade small bowel obstruction (short solid arrow).

Pelvis MRI was then performed which confirmed the presence of extensive innumerable cystic changes within the small bowel mesentery encasing the branches of the SMA and SMV without significant luminal stenosis. The cystic changes demonstrated hyperintense signal on T2 weighted images and hypointense signal on T1 weighted

images with numerous thin septations. However, one of the cystic lesions within the right side of the pelvis measuring about 4.6 x 3.3 x 3.2 cm showed peripheral T1 hyperintensity and diffuse T2 hypointensity suggestive of hemorrhagic content. That hemorrhagic cystic lesion was having a tubular extension superiorly and was compressing one of the distal ileal loops and resulting in partial small bowel obstruction. Proximal to this, the small bowel loops were dilated measuring up to 3.4 cm in maximum diameter. This was suggested to represent an associated band causing the obstruction. There was a small amount of pelvic free fluid. No definite peritoneal nodules were seen. Apart from thin enhancing septations, no soft tissue masses were identified. In addition, the distribution of the abnormality involved the lower abdomen and pelvis with relative sparing of the upper abdomen (Fig. 2). Considering the extensive small bowel mesenteric involvement, presence of hemorrhage and calcifications, lack of solid component and absence of mass effect on the mesenteric vessels despite the large size, it was favored to represent extensive mesenteric lymphangiomatosis.



**Figure 2:** Magnetic Resonance Imaging (MRI) images. (A) Axial T1WI post contrast venous phase showing multiple enhancing septations without solid mass (arrow heads). (B) Axial T1WI without IV contrast showing hyperintense periphery of hemorrhagic component (double arrow). (C) Coronal T2WI showing mesenteric hyperintense cysts with variable signal intensity of different locules with numerous thin septations (arrow heads), normal flow void mesenteric vessels passing through the cystic structures without luminal narrowing (long solid arrow) and dilated small bowel (dotted arrow) (D) Coronal T2WI showing hypointense haemorrhagic cyst (double arrow) and normal left ovary (short solid arrow).

Diagnostic laparoscopy with omental and cystic wall biopsy was done. It showed hemoserous fluid in the pelvis, left and right paracolic gutters and between bowel loops. Multiple clumped cystic lesions were arising from the mesentery and omentum and were adherent to small bowel loops. Some of those cysts were filled with serous fluid and others with what appeared grossly to be mucin. There were scattered mucin-like deposits within the mesentery. Peritoneal fluid was aspirated and sent for cytology, microbiology culture and sensitivity and TB.

Omental and cyst wall biopsies were taken from the deposits. Adhesiolysis was performed, hemostasis was secured, and suction irrigation was done.

Post-operative histological examination revealed omentum with many irregular dilated vascular channels. Most of those vascular channels had smooth muscle layer in their walls. Immunohistochemically, the lining endothelial cells were positive for CD34 and CD31 while D2-40 was positive in few thin-walled vessels (Fig 3). Smears and cell block showed reactive mesothelial cells, macrophages, and inflammatory cells. No mucin or malignant cells were seen. The final diagnosis was hemangiolympangioma of the small bowel mesentery.

**Figure 3:** Fibrofatty tissue with dilated irregular vessels, some with prominent smooth muscle layer (HE x40). Inset shows positive staining for CD31 in the endothelial cells (CD31 x100).

The case was discussed in the tumor board. A conclusion was reached of extensive involvement of mesentery and peritoneum with a vascular malformation [Hemangiolympangioma] which is unresectable. Given that the patient is young and the condition is extremely rare, patient was referred to be seen in specialized center for second opinion.

## Discussion

Hemangiolympangioma is a rare malformation of the vascular and lymphatic systems. It is classified as either congenital or acquired secondary to prior trauma, surgery or infections. It can involve any site, superficial or deep and can be localised or extensive.<sup>9</sup> Hemangiolympangioma of the small bowel mesentery is an extremely rare entity. A literature review was done using the key words "Hemangiolympangioma" OR "Hemolympangioma" in both PubMed and OVID. There has been 9 total reported cases of Hemangiolympangioma of the small bowel, but only 3 reported cases of Hemangiolympangioma of the small bowel mesentery.<sup>8</sup>

In our case, the patient presented with intermittent abdominal pain and signs of small bowel obstruction. Blood tests were normal apart from mild decrease in Hb and mild elevation in CRP/ESR. There are several radiological features that can help differentiate hemangiolympangioma of the small bowel mesentery from metastatic deposits in the peritoneum. These findings can aid with the preoperative diagnosis but primarily depend on the composition of the cysts and the amount of blood vessels it contains. Firstly, on abdominal CT, hemangiolympangioma appears as mesenteric cystic or cystic-solid masses and deposits that are isodense in non-enhanced CT but show increased enhancement in venous and delayed phases. Enhancement may not be obvious if blood vessels are in small proportion. Mural and septal enhancement may be present depending on the thickness of the tissue. Secondly, these malformed vessels may thrombose and cause necrosis which might be seen as calcium deposits. Calcific foci may overlap with the possibility of mucinous tumours. Thirdly, the lesions may engulf the mesenteric vascular branches without causing mass effect, so these vessels remain patent. Lastly, on MRI, T1-weighted images of the cystic lesions appear hypointense while on T2-weighted images they appear as homogeneously hyperintense with thin enhancement of the septations.<sup>2,10,11</sup> A useful clue to the diagnosis in our case was the distribution of the abnormality along the small bowel mesentery and the fact that the abnormality was confined to the lower half of the abdomen with sparing of the upper part. On the other hand, metastatic deposits usually spread along the peritoneum and would be distributed throughout the abdominal and pelvic cavity. In addition, isolated mesenteric metastatic deposits are rare in the absence of peritoneal involvement.

Hemangiolympangioma diagnosis needs to be confirmed pathologically as it is a rare condition and its imaging appearance may overlap with other diagnostic possibilities. The main stay of treatment is complete surgical resection. Other options include sclerotherapy, laser therapy, cryosurgery, electrocautery and radium implantation.<sup>12</sup> Post operative follow-up is crucial for early and timely detection of recurrence.<sup>13</sup>

## Conclusion

In conclusion, hemangiolympangioma of the small bowel mesentery is an extremely rare entity that makes it diagnostically challenging. This case report aimed to highlight the key radiological findings that might help with preoperative diagnosis.

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