Extensive Hemangiolymphangioma of the Small Bowel Mesentery: A Case Report

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Abstract

Hemangiolymphangioma is a rare malformation of the vascular and lymphatic system affecting different parts of the human body. Hemangiolymphangioma of the small bowel mesentery is extremely rare. We report the case of a 23-year-old woman who presented with generalized colicky abdominal pain. Computed tomography (CT) findings were initially thought to represent possible secondary mesenteric and peritoneal metastatic deposits. However, no primary tumor was identified. Pelvis MRI was then performed which favored the diagnosis of extensive mesenteric lymphangiomatosis. The diagnosis was confirmed histopathologically as hemangiolymphangioma of the small bowel mesentery.

Keywords: Hemangiolymphangioma; Hemolymphangioma; Mesentery; Mesenteric Cyst; Small Intestine; Oman.

Introduction

Hemangiolymphangioma is a rare type of venolymphatic vascular malformation[1] containing a network of blood vessels and lymphatics.[14] This condition presents on the body surface of infants and young children, and extremely rarely in adults.[8] In adults, most cases of hemangiolymphangioma occur in the head and neck while a few have been reported in the pancreas,[2] spleen,[3] small intestine,[4] rectum,[5] chest wall,[6] and extremities.[7] To date, nine cases of small bowel hemangiolymphangioma have been reported worldwide.[14] Here, we report a case of small bowel mesentery hemangiolymphangioma with characteristic imaging features that differentiate it from other more sinister conditions.

Case Report

A 23-year-old woman presented with five days' history of intermittent generalized colicky abdominal pain, constipation, and vomiting. Six years earlier she had undergone laparoscopic appendectomy. Abdominal examination showed tense abdomen with diffuse tenderness. Laboratory investigations revealed hemoglobin (Hb): 10.8g/dL, erythrocyte sedimentation rate (ESR): 58 and C-reactive protein (CRP): 83. Other blood tests, tumor markers (B-HCG, CA 15-3, CA 19-9, CA 125, CEA, AFP), and microbiological tests (TB, HIV, HepB/C) yielded normal results. Initial bedside B-mode ultrasonography showed cysts anterior to the uterus.

Contrast-enhanced abdominopelvic CT image showed extensive mesenteric involvement by soft tissue masses and several widespread fluid-attenuating cystic lesions of variable sizes, the largest of which measured 5.0 × 4.6 cm with adjacent nodular calcifications and was located in the left upper quadrant. The lesions were engulfing the mesenteric vasculature which was 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 [Figure 1]. The CT findings were initially thought to represent possible secondary mesenteric and peritoneal metastatic deposits. However, no primary tumor was identified.

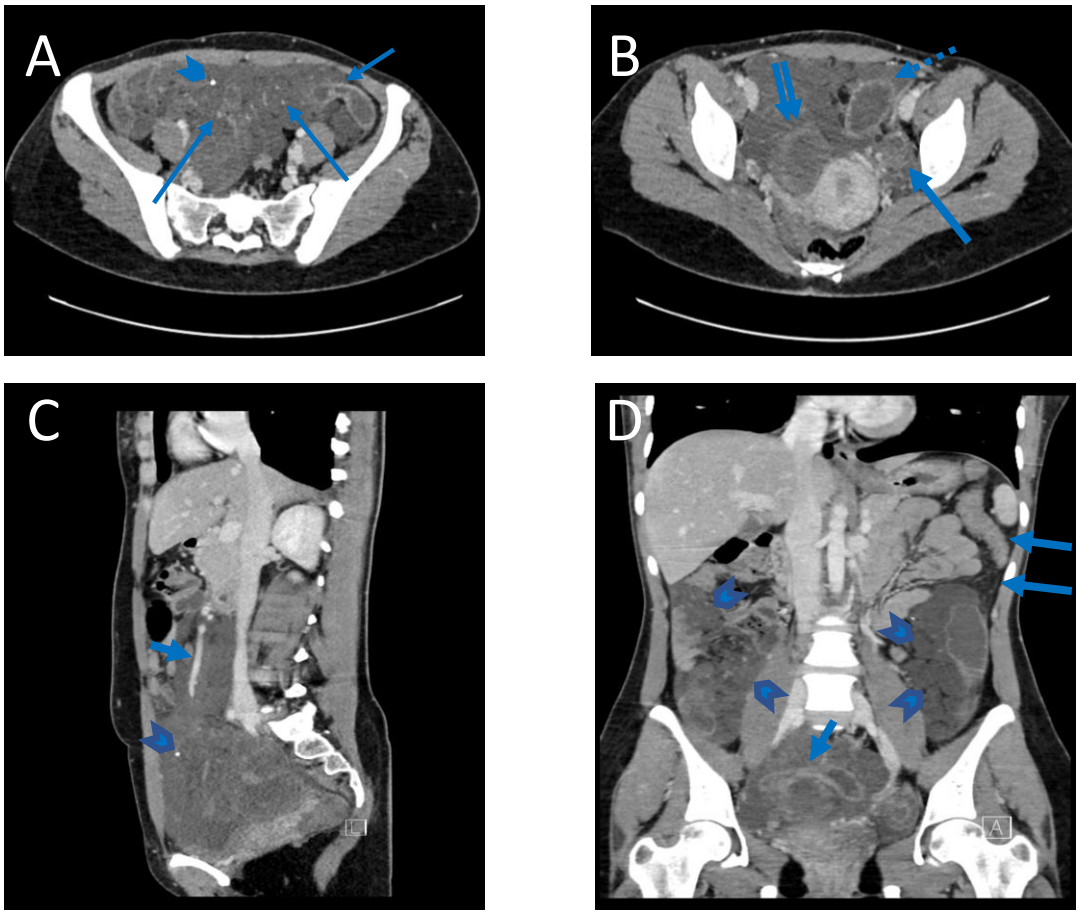

Figure 1: Computed tomography (CT) image with intravenous contrast in the portal venous phase images. (A) Axial image showing soft tissue densities engulfing the mesenteric vascular branches (long solid arrows), with adjacent nodular calcifications (arrow head) and small bowel narrowing (short solid arrow). (B) Axial image showing hemorrhagic cyst (double arrow) with dilated small bowel segment proximal to the hemorrhagic 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 confirmed the presence of extensive innumerable cystic changes within the small bowel mesentery encasing the branches of the superior mesenteric artery (SMA) and superior mesenteric vein (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 × 3.3 × 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, resulting in partial obstruction of the small

intestine. Proximal to this, the small intestinal loops were dilated, 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 [Figure 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.

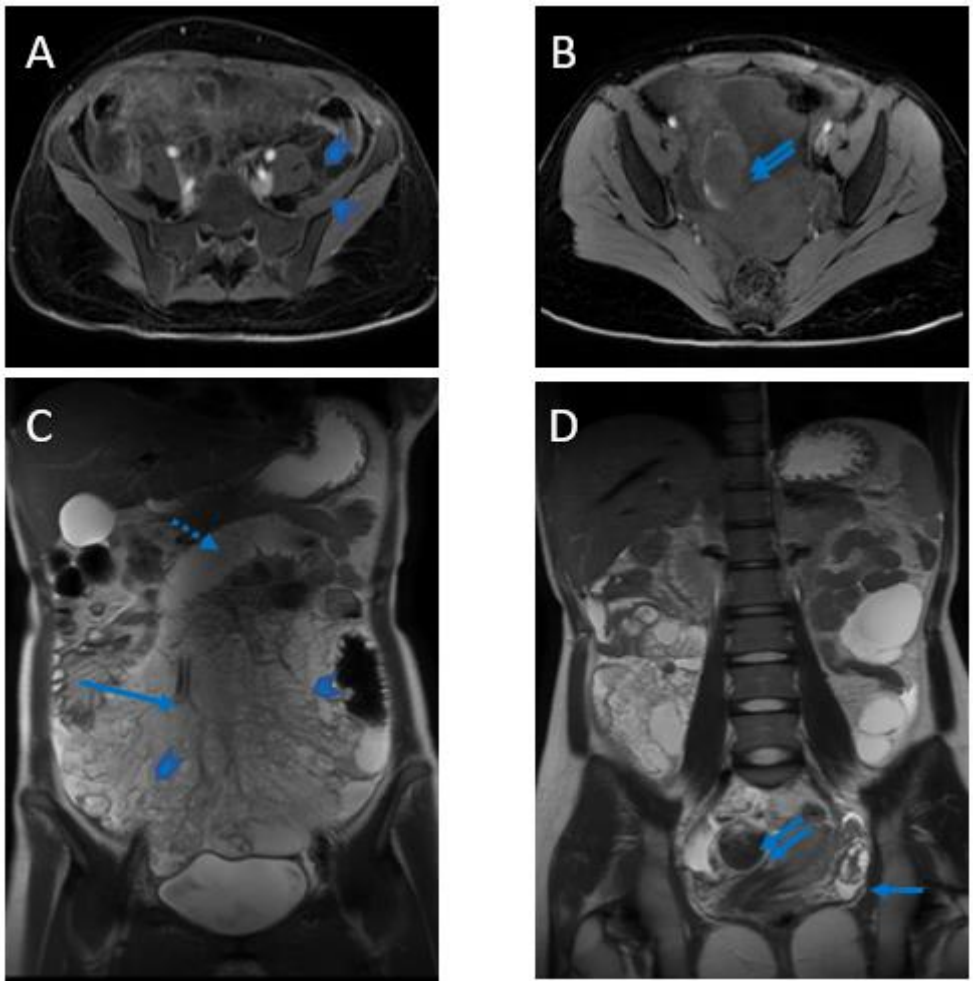

Figure 2: Magnetic resonance imaging (MRI) images. (A) Axial T1-weighted image (T1WI) post contrast venous phase showing multiple enhancing septations without solid mass (arrow heads). (B) Axial T1WI without intravenous (IV) contrast showing hyperintense periphery of hemorrhagic component (double arrow). (C) Coronal T2-weighted image (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 hemorrhagic cyst (double arrow) and normal left ovary (short solid arrow).

Diagnostic laparoscopy with omental and cystic wall biopsy was done. There was hemoserous fluid in the pelvis, left and right paracolic gutters, and between the 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, sensitivity, and TB. Omental and cyst wall biopsies were taken from the deposits. Adhesiolysis was performed, hemostasis was secured, and suction irrigation was performed.

Histological examination of the biopsies 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 [Figure

3]. Smears and cell block showed reactive mesothelial cells, macrophages, and inflammatory cells. No mucin or malignant cells were seen.

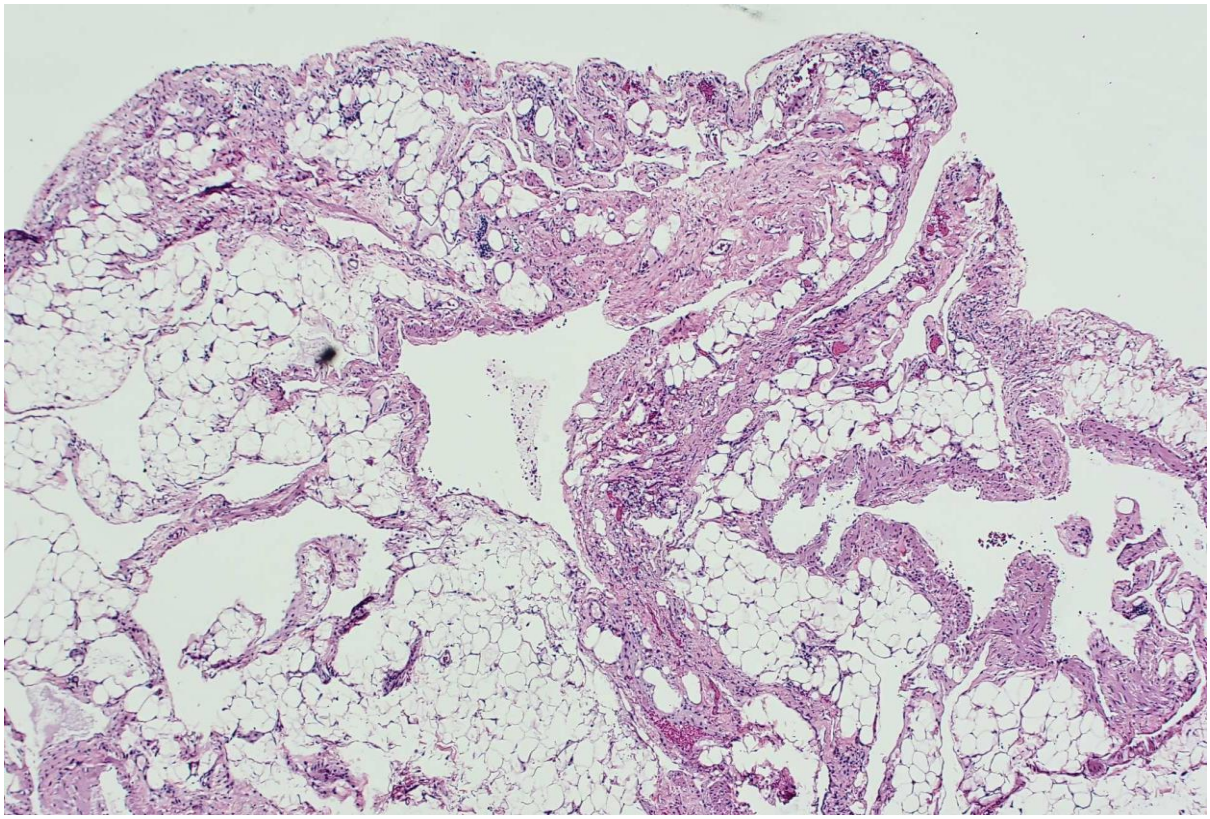

Figure 3: Fibrofatty tissue with dilated irregular vessels, some with prominent smooth muscle layer (hemoxylin eosin stained; x40). Inset: Positive staining for the vascular marker CD31 in the endothelial cells (x100).

The hospital tumor board reached their final diagnosis of hemangiolymphangioma of the small bowel mesentery and noted that due to the extensive involvement of mesentery and peritoneum with a vascular malformation, the tumor was unresectable. In view of the patient's youth and the extreme rarity of her condition, she was referred to a specialized center for a second opinion.

Discussion

Hemangiolymphangioma of the small bowel mesentery is extremely rare.[9] Our literature search yielded nine cases of hemangiolymphangioma of the small bowel, of which only three cases involved hemangiolymphangioma of the small bowel mesentery.[8]

Our patient presented with intermittent abdominal pain and signs of small bowel obstruction. Blood results were normal apart from mild decrease in Hb and slightly elevated CRP/ESR. There are several radiological features that can help differentiate hemangiolymphangioma of the small bowel mesentery from metastatic deposits in the peritoneum. These findings can aid with the preoperative diagnosis but they primarily depend on the composition of the cysts and the amount of blood vessels in them. First, in nonenhanced abdominal CT, hemangiolymphangioma appears as mesenteric cystic or cystic-solid masses and deposits that are isodense but may show increased enhancement in venous and delayed phases. Enhancement may not be obvious if the cystic blood vessels are in small proportion. Mural and septal enhancement may be present depending on the thickness of the tissue. Second, these malformed vessels may thrombose and cause necrosis which might appear like calcium deposits. Calcific foci may overlap with the possibility of mucinous tumors. Third, 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.[2,10,11] 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.

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

Conclusion

Hemangiolymphangioma of the small bowel mesentery is an extremely rare entity that makes it diagnostically challenging. This case report has covered several imaging features that might help clinicians differentiate hemangiolymphangioma of the small bowel mesentery from metastatic deposits in the peritoneum and aid radiologists/clinicians reach correct preoperative diagnosis.

Disclosure

The authors declare no conflicts of interest. Informed consent was obtained from the patient.

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