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Hemolymphangioma of Greater Omentum

A Rare Case Report

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Abstract: Hemolymphangioma is a rare vascular developmental error. It comprises malformed venous and lymphatic component in various proportion. To the best of our knowledge, only a few cases have been reported in the literature so far. Here, we report a case of huge intraperitoneal cystic mass in a 3-year-old boy that was presented to hospital with intractable abdominal pain. On examination, he had fever along with associated symptoms like cough and sputum. Abdomen was distended with no tenderness or rebound tenderness. On computed tomography scan, huge cystic mass was seen and was diagnosed as intraperitoneal benign cystic lesion. Excisional surgery of the lesion was planned. On surgery, lesion was found to be originated from greater omentum and no adhesion was seen in surrounding tissue. Complete excision of the lesion was done. Histopathological specimen after surgery suggested it to be hemolymphangioma. Follow-up for 6 months showed no recurrence.

Hemolymphangioma of the greater omentum is benign tumor and accurate diagnosis before surgery is still a challenge. Presentation of disease may vary from simple well-defined cystic lesion to aggressive ill-defined lesion mimicking malignancy. Therefore, further research is needed to help doctor with preoperative radiological diagnosis and avoid unnecessary radical surgery.

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Abbreviation: CT = computed tomography.

INTRODUCTION

Hemolymphangioma, also known as venolymphatic vascular malformation is a rare vascular developmental error.¹ The formation of this tumor may be explained by obstruction of the venolymphatic communication among dysembryoplastic vascular tissue and the systemic circulation.²⁻⁵ A few cases of hemolymphangioma had been reported in the orbit, pancreas, mediastinum, pericardium, spleen, adrenal gland, cervix hepatica, retroperitoneum, and also in extremities and tongue. To the best of our knowledge, no cases of hemolymphangioma have

been reported in greater omentum (Pubmed). This article is a case report of a greater omentum hemolymphangioma in a 3-year-old boy, the imaging findings and pathological features of the case were also discussed.

CASE REPORT

A 3-year-old boy complained of mild but progressively increasing abdominal pain around umbilical region for 2 days with no other associated symptoms. After 2 days the pain was intractable and he finally presented to hospital. At the time of presentation, he had fever and also other associated symptoms like cough with scanty sputum. He was diagnosed with upper respiratory tract infection. Further evaluation of abdominal pain was done by ultrasonography, where huge cystic mass was found. Then he was transferred to our hospital (the First Affiliated hospital of Sun Yat-Sen University) for tertiary care. His past medical and traumatic history was not significant. On physical examination, abdomen was distended with no tenderness or rebound tenderness.

Laboratory examinations found that white blood cell count is 26.1 billion/L (normal range, 3.5–10.5 billion/L); alkaline phosphatase (ALP), 163 units/L (normal range, 0–110 units/L); and lactate dehydrogenase (LDH), 293 units/L (normal range, 114–240 units/L). On computed tomography (CT), a huge intraperitoneal mass occupying almost all intraperitoneal spaces including pelvic cavity was seen (Figure 1). Small bowel, transverse colon, and sigmoid colon were pushed up from their normal position (Figure 1). On nonenhanced CT, homogenous low attenuated lesion with average density of 10 HU and intersposed thin septa with isodense (density similar to muscle) amorphous lesion were seen (Figure 2). On contrast enhanced CT, mild enhancement was seen in septa and amorphous part but cystic part did not take contrast (Figure 2). Radiologically, it was diagnosed as intraperitoneal benign cystic lesion and differential diagnosis of lymphangioma and mesenteric cyst was made.

The fever was managed by antibiotics and after it was well controlled, the patient underwent abdominal laparotomy followed by surgical excision. During surgery, a giant polycystic lesion of 20 cm × 15 cm × 6 cm was seen with brown cystic fluid. Superiorly, the lesion extends from inferior margin of spleen up to the pelvic cavity caudally. The lesion was not adherent to neighboring tissue but the base was attached to greater omentum. The tumor was excised after complete separation. On postsurgical histopathological examination, polycystic lesions, with dilated lymphatic vessel and blood vessel, were seen (Figure 3). Dilated endothelial-lined cystic lesion was further infiltrated with inflammatory cells. On immunohistochemistry, some endothelial were relatively positive for CD 31 and CD 34 and other were positive for D2-40 (Figure 3). No any atypical cell with hyper proliferation or mitotic division was seen. Finally, the diagnosis was confirmed to be hemolymphangioma originating from greater omentum. The postoperative

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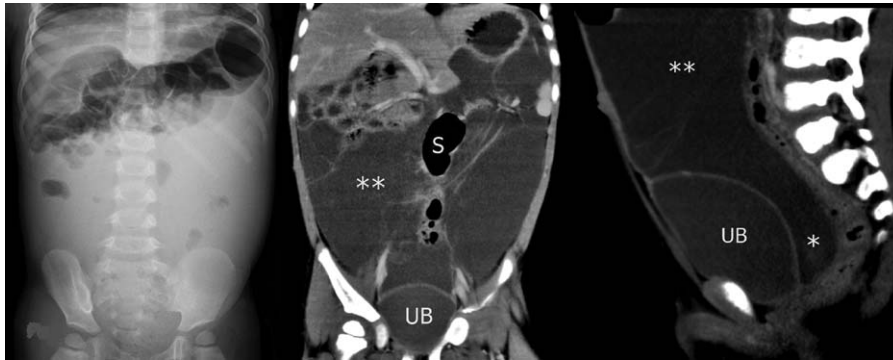


FIGURE 1. Plane x-ray (left), coronal view (middle), and sagittal view (right) of the abdomen; huge intraperitoneal cystic lesion (**) of density similar to UB with interposed septa is seen. All bowels including sigmoid colon (S) are seen displacing superiorly and posteriorly. UB = urinary bladder.

course of the patient was uneventful and the patient was discharged 10 days after the surgery. No complicated or recurrence was noted during 6 months of follow-up. This study was approved by the First Affiliated Hospital of Sun Yat-Sen University Institutional Review Board. Written consent for this case report was obtained from the patient.

DISCUSSION

Hemolymphangioma is a rare benign lesion that appears to arise from congenital malformations of the vascular system. It is thought to originate from the mesenchymal tissue. It comprises both venous and lymphatic components in various proportions. The formation of this tumor may be explained by obstruction of the venolymphatic communication between dysembryoplastic vascular tissue and the systemic circulation.²⁻⁵ The co-existence of lymph and hemangioendothelial cells in hemolymphangioma strengthens the hypothesis of a common stem cell from which both blood and lymph-endothelial cell originate.⁶ The vascular channels are lined with flat mature endothelium exhibiting normal rates of endothelial cell turnover, although some stress conditions like infection, trauma, hormonal influences, or progressive hemodynamic forces cause disproportionate growth, the walls of which appear thin because they lack smooth muscle. The architecture of the constituent vessels on routine H&E histological preparations is usually distinguished, although this can sometimes be difficult. In such cases, immunoreactivity for podoplanin with the D2-40 antibody helps to differentiate it from lymphatic endothelial.⁷

These lesions are presented at birth. Clinically, the onset of hemolymphangioma can vary between a slow-growing lesion

over a period of years and an aggressive enlarging tumor without invasive ability.⁸ These lesions can arise at any sites, can be localized or more extensive, and can be superficial or deep. Deep lesions without superficial involvement may remain unrecognized until the patient presents with clinical symptoms later in life.⁹ The small intra-abdominal cyst may not present any symptoms, unless it enlarges significantly and compresses adjacent organs. The larger cyst could be presented with acute abdomen or dull aching pain. Possible etiologies for acute abdominal pain proposed in the literature include intestinal obstruction, torsion, infarction, traction of the mesentery as the cysts expand superimposed infection, and peritoneal irritation due to leakage of the cyst contents.¹⁰ Infection or trauma may cause these lesions to acutely increase in size.¹¹

As mentioned in the literature, this case was presented with dull abdominal pain for few days; this could be due to the pressure effect of huge lesion into the abdominal wall and internal organs. Later, the pain progressed to become intractable and was superimposed by fever. Raised WBC count in blood and inflammatory cell infiltrates within the lesion on histopathology, probably could be due to acute inflammatory change within the lesion that manifests as fever and newly onset intractable pain. Also ALP and LDH are seen to be raised. The raised ALP and LDH on oncology are used for predilection of metastasis in various malignant cancer, and this represents the worse prognosis. But our case is a benign vascular malformation with no radiological or histological feature of malignancy. ALP is a hydrolase enzyme concentrated in certain organs, such as liver, kidney, bone, and placenta but can also present in many other tissues.¹² ALP in this case is slightly higher than normal range and that could probably be due to

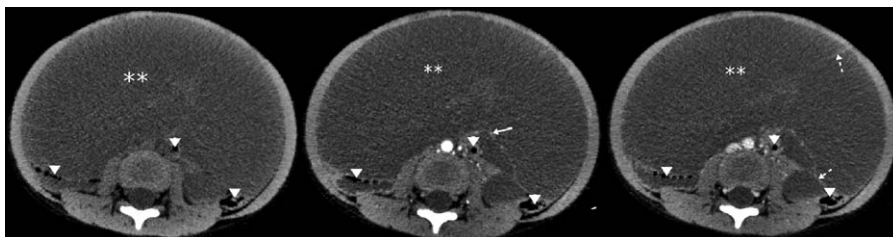


FIGURE 2. Axial view of abdomen (plain, arterial, and venous phase, respectively). Huge hypodense cystic lesion with interposed septa is seen. Cystic lesions do not enhance, but interposed septa show mild enhancement in arterial phase that further enhances in venous phase (dotted arrow). Normal artery (arrow line) is seen within the septal stroma. Bowels (arrow head) are displaced and are compressed posteriorly with abdominal wall.

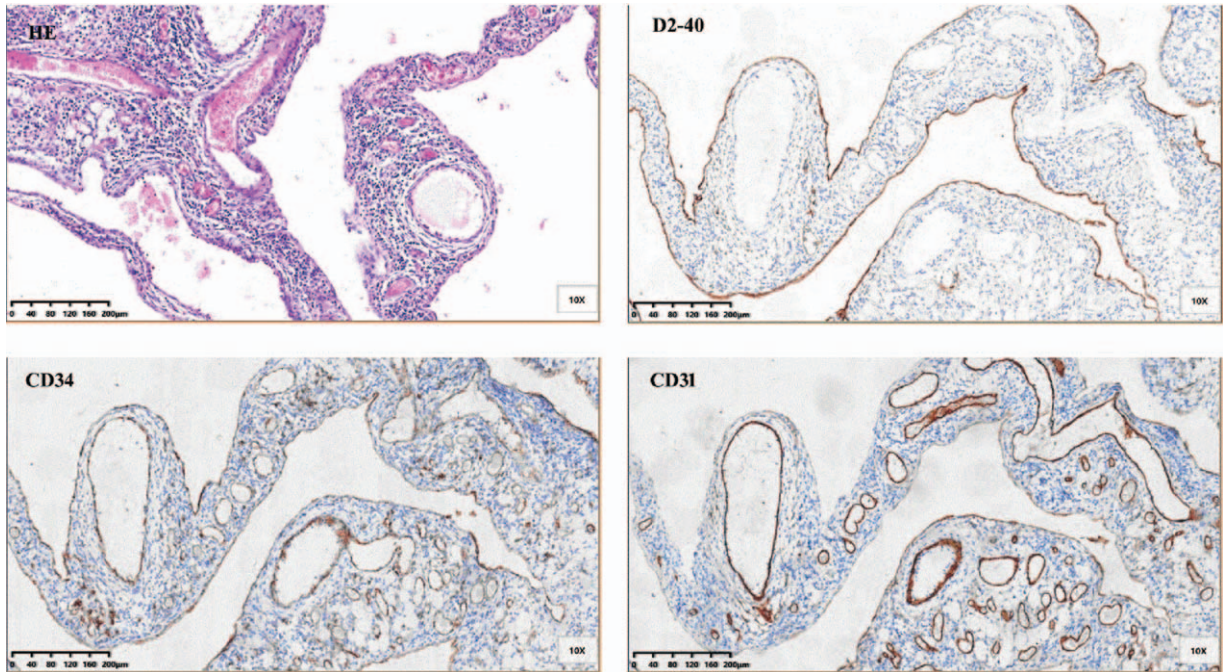


FIGURE 3. Histopathological examination of the greater omentum hemolymphangioma (H&E staining). On H&E preparation, dilated vascular channels with blood cells (in blood vessel) and with clear fluids (in lymphatic vessels) are seen. Inflammatory cells are seen infiltrating the stroma between the dilated vascular channels. Large lymphatic channels are seen positive for D2-40. Vascular endothelial showed positive for CD31 and CD34. H&E = hematoxylin and eosin.

normal bone remodeling for that age. LDH is an important enzyme in energy production in many cell types, which catalyzes the conversion of pyruvate to lactate in hypoxic conditions. Increased LDH in this case could probably be due to hemolysis of stagnant blood within malformed vasculature or due to traumatic hemorrhage. But no significant hemorrhage is seen on radiological examination. Traumatic rupture, hemorrhage, lymphorrhea followed by secondary infection are common complications of hemolymphangioma.⁸

CT scan can be very helpful in radiological diagnosis. Hemolymphangioma shows a combined architecture of dilated vein, dilated lymphatics, and normal stromal tissue and normal vasculature in between. Venous components are isodense (as aorta) lesion in nonenhanced CT that enhances significantly in venous and delayed phase (Figure 3). Malformed and dilated venous vessels are usually presented with thrombosis that can form dystrophic necrosis and calcium deposition. Lymphatic component shows attenuation similar to water corresponding serous and chyle, but occasionally may exhibit higher attenuation reflecting infection, hemorrhage, or inspissated mucoid material. The stroma and septa show various level of enhancement, depending on the thickness and composition of tissue of origin.

A thorough exploration of the abdominal cavity should be performed because of the potential invasion to the surrounding organs. Complete excision provides the best results with a lower recurrence rate. However, a careful follow-up is necessary. The recurrence rate varies depending on the complexity, the anatomical location, and the adequacy of the excision.¹³ It has been established in the literature that lesions that have been completely excised show 10% to 27% recurrence, while 50% to 100% of partly resected tumors may recur.¹³ Other nonsurgical treatments, including cryo therapy, laser therapy, radiotherapy,

and local injection of sclerotic agents, compared with surgical treatment, do not show superiority.⁸

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