Unilocular Cystic Lymphangioma of the Small Omentum in a Girl of 4 Years

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Abstract

Background: Cystic lymphangiomas (CL) are congenital benign malformations of lymphatic vessels. Their usual locations (90%) are neck and axilla in subcutaneous tissue. Abdominal locations are rare and represent 10–12%. The incidence of mesenteric and omental cysts is 1 in 20,000 among children, and even lower among infants. Only 2.2% are omental cysts of these two types of cystic mass. Of omental CL, unilocular forms are rarer but more common in boys than in girls. We aim to present clinical, radiological and therapeutic management of a small omental unilocular CL, slowly evolved in a girl.

Observation: A girl of 4-years suffered of chronic intermittent abdominal pain without vomiting and fever, managed like parasitic infection during a year. One year after, appears an abdominal swelling leading to pediatric surgery department. There was no prenatal diagnosis. The preoperative radiological diagnosis by ultrasonography and scan was simple mesenteric cyst, especially because of the unilocular presentation aspect. One week before scheduled surgery it was complicated by intracystic hemorrhage. The lesion of 17 cm × 8 cm was implanted in the small omentum under the liver between hepatic pedicle and the gastric artery. It had sero-hematic content and was totally removed by laparotomy. Hystological examination concluded to CL. The outcome was favorable after 13 months follow up.

Conclusion: CL has many forms, locations and cystic contents. Unilocular cystic lymphangiomas located in the small omentum are very rare and can be complicated at any time. It is more often confused with mesenteric cysts but they are distinct from one another by pathological examination which gives the final proof of the diagnosis. Surgical resection should be as complete as possible, putting the patient free from recurrences.

Keywords: Cystic lymphangioma; Abdomen; Omentum; Children; Surgery

Introduction

Cystic lymphangiomas (CL) are congenital benign malformations of lymphatic vessels. Lack of lymphatic vascular connections secondary to an abnormal embryological development of the lymphatic system lead to a sequestration of the lymphatic tissue. It results in obstruction of the local lymphatic flow and the development of lymphagectasias. Their origin remains uncertain [1-3]. Described for the first time by Koch one century ago, it is known that this anomaly occurs primarily in children and 60% of cases are diagnosed before the age of 5 years [4]. Usual locations (90%) of cystic lymphangioma (CL) are neck, extremities and axillary in subcutaneous tissue. Abdominal locations are rare and represent 10%-12% [5]. In abdomen, locations are also variable and involve mesentery, mesocolon, retroperitoneum, omentum, splenic loge, liver and pancreas [2-4,6]. Abdominal locations cause diagnostic and therapeutic problems. We report a CL located in the gastro-hepatic ligament and focus by literature reviewer on its abdominal locations.

Case Presentation

A 4-year-old girl was admitted on a scheduled basis in pediatric surgery department for an abdominal mass about 1 month before, associated to chronic abdominal pain. Second child of a fourth’s siblings without pass history of abdominal mass in the family, she was born at term. There was no previous antenatal diagnosis of abdominal mass. Her parents are alive and well.
In fact, she had abdominal intermittent pain since one year without vomiting or fever. One abdominal ultrasound realized at the beginning of the symptoms was normal. She had analgesics and ant parasitic drug, four times in year duration. One month before scheduled admission, her parents realized abdominal epigastric swelling. That time it was not associated with pain. She had a preserved transit and general condition. She had no fever. The examination noticed an epigastric abdominal swelling (Figure 1) where a moving painless mass was felt. There was no node clinically felt. She had a good cutaneo-mucous coloration. The rest of the examination was normal. Abdominal ultrasound showed a unicystic mass with a regular wall and a homogeneous content. An abdominal scan (Figure 2) revealed a unicystic anterior abdominal tumor with homogeneous content resulting to cystic mesenteric cyst.

The patient was scheduled for exploratory laparotomy and tumor excision. One week before surgery, she had moderate acute abdominal pain. She had maintained a normal hemodynamic state. At laparotomy we discover a unicystic tumor implanted in the small omentum (Figure 3). It was very limited measuring 17 cm × 8 cm. It had cracked and bleeding with hematomas in the great omentum. The content was sero-hematic, mixing yellowish fluid with blood due to the intracystic hemorrhage. Figure 4 shows the post-resection operative site. The patient left hospital five days after. The final histological diagnosis was cystic lymphangioma. The patient was doing well 13 months postoperatively.

**Discussion**

Cystic lymphangioma (CL) is conjunctival malformative vascular tumours corresponding to a detention of lymphatic tissue due to an abnormal embryonic development of the lymphatic system [7]. It is a congenital benign malformation of lymphatic vessels. In fact, the matter would be a lack of lymphatic vascular connections secondary to an abnormal embryological development of the lymphatic system which lead to a sequestration of the lymphatic tissue. These arrested developments of lymphatico-venous connections during the embryogenesis would cause absence of drainage of the primitive lymphatic bags leading to the formation of a cystic lesion containing some lymph. It results in obstruction of the local lymphatic flow and the development of lymphagectasias. Their origin remains uncertain [1-3,8]. Described for the first time by Koch one century ago, it is known that this anomaly occurres primarily in children according to its embryological development and 60% of cases are diagnosed before the age of 5 years [4]. This congenital theory is strengthened by observations of CL detected in the prenatal period. Or mostly in children population however, the diagnosis can be made at any age [8]. In our case there was no any prenatal discover of abdominal mass but the cyst occurs at 4 years old. CL is usually benign but can be locally invasive [9].

Usual locations (90%) of cystic lymphangioma (CL) are neck, extremities and axillary in subcutaneous tissue [10,11]. But variety of other sites have been described including the mediastinum, pleura, pericardium, groin, bones and abdomen [12,13]. Abdominal locations are rare and represent 10%-12% [5]. In abdomen, locations are also variable and involve mesentery, mesocolon, retroperitoneum,
Laparoscopic management has the advantages of lower risk and faster recovery compared to open surgery. Malignant transformation of cystic lesions is rare. Resection of the bowel and recurrence are rare. The preferred treatment of omental cysts is complete excision, whether laparoscopic or not. Resection of the bowel and recurrence are rare. Malignant transformation of cystic lesions is rare.

Clinical presentation can be variable and nonspecific. Acute symptoms include acute abdomen, distension, vomiting, and fever. Chronic symptoms include progressive abdominal distension and pain. Patients admitted to the hospital may be classified into two main groups: those with acute clinical symptoms and those with non-acute clinical symptoms. Diagnosis of a cyst should be considered even if the findings are non-specific and the patient exhibits symptoms over a long period of time. The presence of complicating factors, including hemorrhage, torsion, and infection, or pressure to other structures, is relevant with acute presentations that require urgent surgery. Pathologically, they can be unilocular or multilocular and complicated at any time. It is more difficult in unilocular case, as in our case. "Clinical presentation can be variable and nonspecific. Acute symptoms include acute abdomen, distension, vomiting, and fever. Chronic symptoms include progressive abdominal distension and pain. Patients admitted to the hospital may be classified into two main groups: those with acute clinical symptoms and those with non-acute clinical symptoms. Diagnosis of a cyst should be considered even if the findings are non-specific and the patient exhibits symptoms over a long period of time. The presence of complicating factors, including hemorrhage, torsion, and infection, or pressure to other structures, is relevant with acute presentations that require urgent surgery. Pathologically, they can be unilocular or multilocular," knowing that unilocular form are more rare. Preoperative diagnoses are more difficult in unilocular case, as in our case.

"CL is more often confused with mesenteric cysts that arise from mesothelial, not lymphatic tissue. This differentiation is important because lymphangiomas often behave in an invasive and aggressive manner, while as mesothelial cysts do not. Despite being difficult to differentiate between imaging studies, they are histologically distinct from one another. Lymphangiomas have an endothelial lining, foam cells, and a wall that contains lymphatic spaces, lymphoid tissues, and smooth muscles."

Clearly the case we described caused more preoperative diagnosis difficulties probably because of its location (smalio mentum), form (unilocular) and complication (intracystic hemorrhage).

The content of the cyst is sero-hematic in our case. The content of CL can be, serous or sero-sanguineous. These different aspects can be explained by different degrees of lymphaticstasis, a variable number of connections with the lymphatic system and the protein content of the cyst contained. The sero-hematiccyst appearance is secondary to intra cystic hemorrhage. Rarely CL can be purulent by infection.

The preferred treatment of omental cysts is complete excision, whether laparoscopic or not. Resection of the bowel and recurrence are rare. Malignant transformation of cystic lesions is also rare. Laparoscopic management has the advantages of lower cost and decreased morbidity compared to open surgery.

**Conclusion**

Cystic lymphangioma has many forms, locations and cystic contents. Unilocular cystic lymphangiomas located in the small omentum are very rare and can be complicated at any time. It is more often confused with mesenteric cysts but they are distinct from one another by pathological examination which gives the final proof of the diagnosis.

**References**


