MESENTRIAL CYST LYMPHANGIOMA IN 3-YEAR-OLD GIRLS: A CASE REPORT

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Abstract: Mesenterial cysts are rare tumors, incidence 1: 100,000-200,000. We report the case and the treatment. *Case report, 3-year-old woman with complaints of a lump in the abdomen. Complaints in the last 1 year ago, sometimes she felt pain, physical examination showed a lump of cystic impression with a size of ± 5x10 cm in the epigastric region, mobile, firm boundary. It can be diagnosed with a suspected intra-abdominal tumor with a mesenterial cyst, differential diagnosis of teratoma. Surgery was performed and a mass was found in retroperitoneal with a size of ± 20x5 cm up to the pelvic cavity, cystic and mobile, the mass could not be taken at all, we decided to do marsupialization, and left kidney is not visible. Postoperative diagnosis is retroperitoneal (type IV) mesenterial cyst lymphangioma with agenesis ren sinistra. Mesenterial cyst Lymphangioma is a tumor that is very rarely found and is benign. For a good prognosis, it depends on the type and therapy that is performed.*

Keywords: *Mesenterial cyst lymphangioma, a tumor in children, mesenterial tumor*

INTRODUCTION

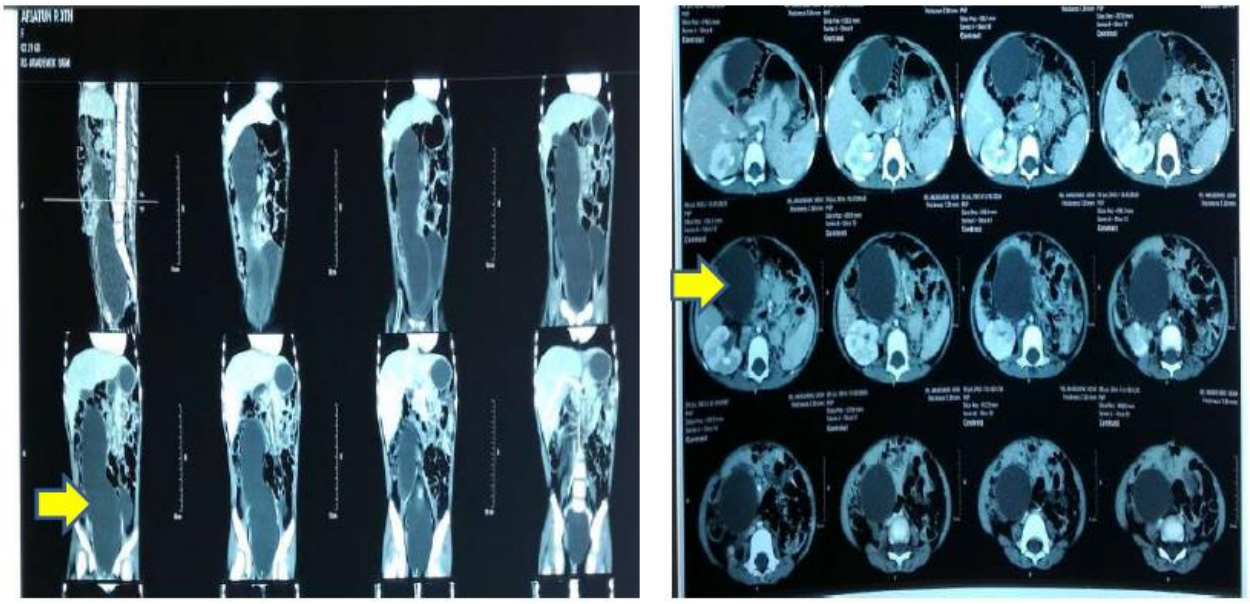
Mesenterial cyst is a rare tumor with an incidence of about 1: 100,000-200,000. Lymphangiomas are benign proliferation of lymph vessels, producing fluid-filled cysts produced from obstruction of the lymphatic system. Mesenterial cyst was first reported by Benevieni, an anatomist from Italy in 1907 while performing an autopsy on an 8-year-old girl, earlier in 1842 Von Rokitansky described the presence of a chylous type of mesenterial cyst.[1] A study revealed that almost a third of mesenterial cysts appears in children <15 years, and a quarter of mesenterial cysts appear in children aged <10 years. These cysts are more common in girls than boys.[1,2]

Mesenterial cysts are often asymptomatic and are often detected unintentionally on abdominal examinations for other diseases and on medical checkups or found accidentally during a laparotomy.[3,4]

In children, the symptom is usually abdominal distension and can be accompanied by a palpable mass during palpation, but in some cases the mass is not found during physical examination. The mass can be large so that it resembles the ascites.[1,3,6]

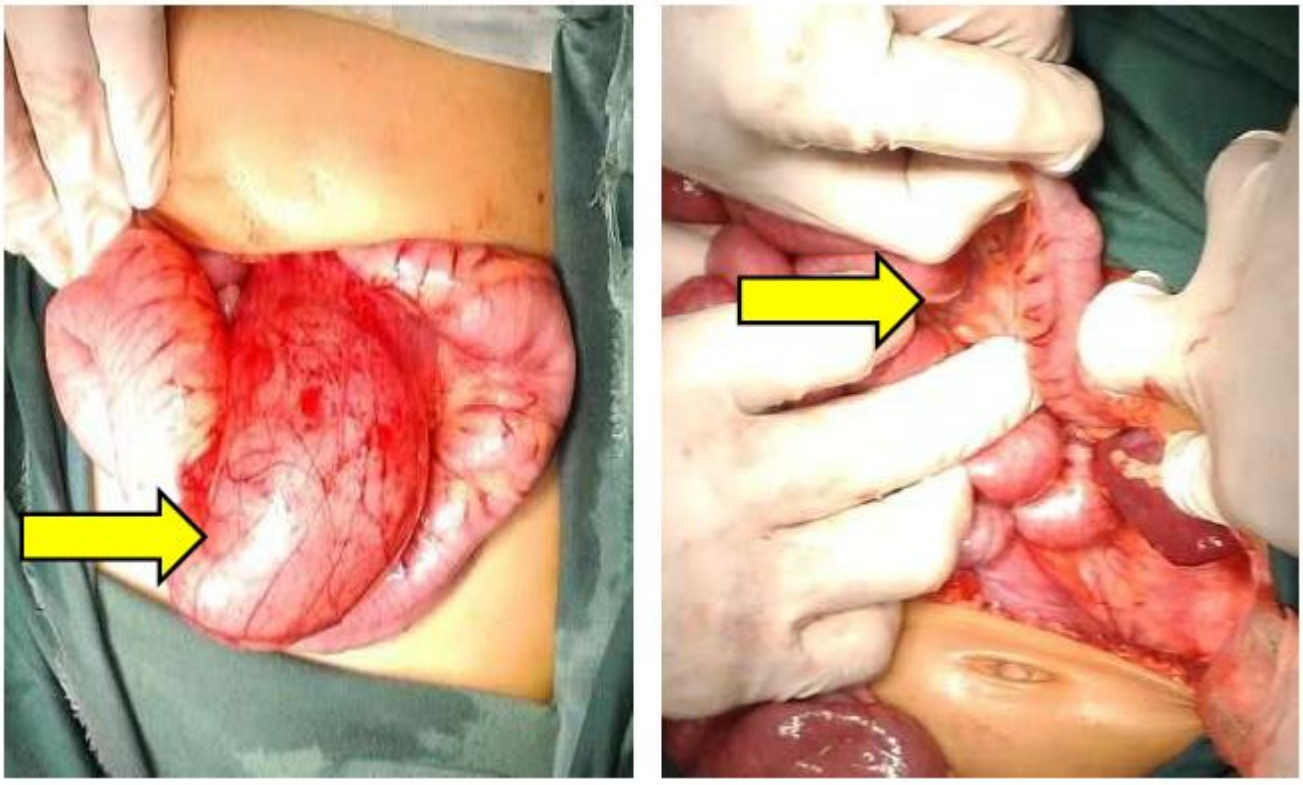
CASE REPORT

Girls aged 3 years with complains of a lump in the abdomen since 1 year ago, sometimes felt of pain, physical examination showed a cystic lump with a size of ± 5x10 cm, located in the epigastric region, mobile, firm boundary. On the history and physical examination, it can be suspected with a diagnosis of a suspected intra-abdominal tumor with a mesenterial cyst with a differential diagnosis of teratoma. Blood tests showed that Hb 10.9 g/dl, Alpha Fetoprotein (AFP) 0.87 ng/ml (normal 0.2–9.0 ng/ml). On contrast CT examination of the abdomen found a tubular oval cystic mass in the middle abdomen tending to the dextra that extends to the pelvic cavity, which causes hydronephrosis dextra of grade I and forces the intestine anteriorly (figure 1). The child underwent exploratory laparotomy through a transverse supraumbilical incision is carried out to the peritoneum. After the peritoneum is opened, a retroperitoneal mass of ± 20x5 cm is found until the pelvic cavity, cystic and mobile (figure 2), are punctured by a mass and out of serous fluid ± 220 cc, when the retroperitoneal cavity is opened, the mass cannot be taken all, it is decided to be done marsupalization. And further exploration, left kidney is not visible. Postoperative diagnosis is retroperitoneal (type IV) mesenterial cyst lymphangioma with agenesis ren sinistra. The results of anatomic pathology examination are mesenterial tissue: supporting the diagnosis of mesenterial cyst lymphangioma, cystic fluid: no malignant cells are found, supporting the diagnosis of cysts (figure 3).



A **B**

Figure 1. (A,B) CT scan of the abdomen, yellow arrows showing cysts



A **B**

Figure 2. Operation findings (yellow arrows): A. Mesenteric lymphatic cyst mass. B. There is no left kidney

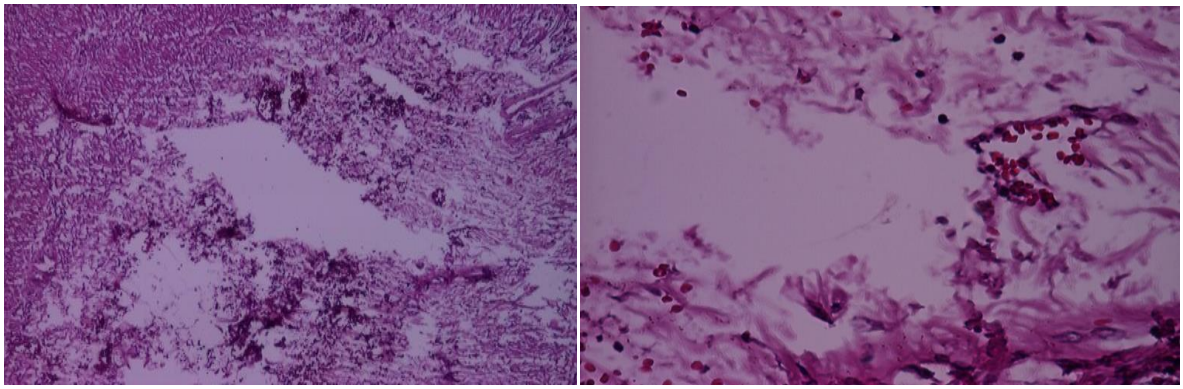


Figure 3. Histology of mesenteric lymphangioma cysts, having endothelial cells, foam cells, and thin walls containing lymphatic spaces, lymphoid tissue, and smooth muscle

DISCUSSION

Mesenterial cyst is a rare tumor with an incidence of approximately 1: 100,000 - 200,000. It can occur at any age, but one-third of cases occur in children under the age of 15 with an average age of 4.9 years. Lymphangioma is a benign proliferation of lymph vessels, producing fluid-filled cyst that result from blockages of the lymphatic system. These tumors can be located anywhere from the duodenum to the rectum even though most locations are found in the ileum.[1,2] In this case, a 3-year-old girl, with complaints of epigastric lumps and sometimes pain, after physical examination and support, the patient was diagnosed with an intraabdominal tumor. Suspicious of mesenterial cysts with differential diagnosis of teratomas. The child underwent exploratory laparotomy through a transverse supraumbilical incision, it is found that the cystic mass is retroperitoneal and, left kidney is not visible. The diagnosis after surgery is retroperitoneal type mesenterial cyst lymphangioma (type IV) with ren sinistra agenesis. Mesenterial cysts are classified based on the cause. Bear et al mention there are four causes of this mesenterial cyst: (1) Embryonic and developmental cyst, (2) Trauma or due to acquisition, (3) Neoplastic, (4) Infection or degenerative. In this classification, mesenterial, omentum, retroperitoneal, dermoid, urogenital, and enteric cyst duplication will be classified as "embryo and developmental cyst." [3]

Some authors distinguish between cyst lymphangioma and mesenterial-omentum cyst on the basis of histology. Cyst lymphangioma is a simple cyst with endothelial cells, foam cells, and thin walls that contain small lymphatic spaces, lymphoid tissue, and smooth muscle. The mesenterial cyst wall has no smooth muscle and lymphatic space, and the lining cells are cuboidal or columnar. Lymphangioma occur in the mesenterial or retroperitoneal and tend to appear with acute symptoms of the abdomen, whereas mesenterial cyst are confined to the mesenterial and appear in adulthood as an asymptomatic mass.[1] Antenatal detection of cyst abdominal lesions may be performed on the fetus during antenatal ultrasound scanning. Sonological features can help differentiate the lesion from many other differential diagnoses. As this usually does not change obstetrical management, a definitive diagnosis is usually made in the postnatal period.[6,7] Existing pathological classification systems: Type 1 (pedicled) and 2 (sessile) are limited to the mesenterial and can be fully excised with or without resection of the intestine involved. Types 3 and 4 (Multicentric) extend to the retroperitoneum and require complex surgery and often with sclerotherapy to be good.[8] In this case, a type 4 lymphangioma cyst that extends to the retroperitoneum, so that in the management of operations performed exploratory laparotomy, cystectomy, marsupialization due to mass

cannot be excised, and would not form a pocket again. The patient was not found left kidney. Unilateral renal agenesis is usually asymptomatic when the other is normal. Renal agenesis caused by a failure in the development of the uereteric bud and metanefric mesenchyme. In unilateral renal agenesis caused by a mutations from several genes such as RET (10q11.2), BMP4 (14q22-q23), etc.[9] The prognosis of this lymphangioma mesenterial cyst is good, depending on the therapy carried out on the patient. In this case exploratory laparotomy, cystectomy, and marsupialization were performed, and in the treatment for seven days the patient's condition improved.

CONCLUSION

Mesenterial cyst lymphangioma is a rare tumor and benign. it can be asymptomatic and detected through investigations. For a good prognosis, it depends on the type and therapy that is performed.

REFERENCES

1. Ricketts RR. Pediatric Surgery: Mesenteric and Omental Cyst. Elsevier 7th edition. 2012
2. Lockhart C, Kennedy A, Ali S, et al. Mesenteric cysts: a rare cause of abdominal pain. *Ulster Med J*. 2005 May. 74(1):60-2
3. Feins NR, Raffensperger JG. Cystic hygroma, lymphangioma, and lymphedema. Raffensperger JG, ed. *Swenson's Pediatric Surgery*. 5th ed. Norwalk, Conn: Appleton & Lange; 1990. 172-3.
4. Lee DL, Madhuvrata P, Reed MW, Balasubramanian SP. Chylous mesenteric cyst: A diagnostic dilemma. *Asian J Surg*. 2013 June 13
5. Losanoff *et al*. Mesenteric cystic lymphangioma: unusual cause of intra-abdominal catastrophe in an adult. International Journal of Clinical Practice Volume 59, Issue 8, pages 986–987, August 2005
6. Wootton-Gorges SL, Thomas KB, Harned RK, et al. Giant cystic abdominal masses in children. *Pediatr Radiol*. 2005 Dec. 35(12):1277-88
7. Ma A, Ayre K, Wijeyekoon S. Giant mesenteric cyst: a rare cause of abdominal distension diagnosed with CT and managed with ultrasound-guided drainage. *BMJ Case Rep*. 2012 Sep 3. 2012
8. Losanoff JE, Richman BW, El-Sherif A, Rider KD, Jones JW: Mesenteric cystic lymphangioma. *J Am Coll Surg* 2003, 196(4):598-603.
9. Schreuder. Renal Agenesis, Unilateral. Www. Orpha.net.July 2014