

Isolated Giant benign Multicystic peritoneal Mesothelioma Developed in the Parenchyma of the Stomach: Report of Two Cases

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Abstract

Benign Multicystic Peritoneal Mesothelioma (BMPM) is an uncommon lesion of the serosal membranes. Many terms used in the literatures to describe this particular entity. There are larger series reported from pathologist consultation files with patients from different institutions with incomplete clinical information and lack of long-time follow-up data. Therefore, this disease is classified as an exceedingly rare medical entity. We reported two cases with such rare entity of BMPM admitted and managed in our pediatric surgery unit with a review of the literature.

Case 1: A 4 years old male was admitted to our pediatric surgery unit with a preliminary diagnosis of appendicitis. For 15 days prior to admission, he had been complaining of a crampy abdominal pain, episodes of non-bilious vomiting, progressive abdominal distention and constipation. The patient was operated with a supraumbilical transverse incision. Operation revealed a cystic mass filled with dark brown-dark green serous fluid, which was attached to the greater curvature of the stomach.

Case 2: A 28-month-old male presented with a few days history of abdominal pain, mild fever with twice episodes of non-bilious vomiting. The CT images confirmed the sonogram studies. The patient was operated with a supraumbilical transverse incision. Operation revealed a cystic mass filled with dark brown-dark green serous fluid, which was attached to the greater curvature of the stomach. In both case the histopathology an Immunohistochemistry revealed and confirmed BMPM. Benign multicystic mesothelioma should be kept in mind as an unusual cause for an abdominal mass in children.

Introduction

Benign Multicystic Peritoneal Mesothelioma (BMPM) is an uncommon lesion of the serosal membranes [1-3]. In the works of literature, many terms used to describe this entity include cystic mesothelioma, multilocular peritoneal inclusion cyst, peritoneal cystosis, postoperative peritoneal cysts and multicystic mesothelial proliferation. It was first described in 1979 by Menemeyer and Smith since then not more than 130 cases have been reported and the information regarding BMPM is derived from a small number of patients from a few institutions or from isolated case reports [1,4]. Despite there are larger series reported from pathologist consultation files with patients from different institutions but with incomplete clinical information and lack long-time follow-up data [5]. Therefore, this disease is classified as an exceedingly rare medical entity, which challenges its origin, pathogenesis, diagnosis and therapy. Although this lesion has the potential to develop in all serosal cavities, reports pointed its occurrence most commonly in the peritoneal cavity, with only rare isolated cases reported in the pleura, pericardium, and tunica vaginalis [6-8]. Most cases occur in young to middle-aged females. In most cases, BMPM remains silent in the abdominal cavity and may invade the underlying organs, and more rarely the retroperitoneum [9]. Presenting symptoms are nonspecific and can include abdominal pain (sometimes-acute abdomen), distention, or a palpable mass. The exact etiology and pathogenesis of BMPM is a controversial entity, and controversy exists regarding its classification as a reactive or neoplastic condition. In many reported patients, multicystic mesothelioma is manifested with a history of previous abdominal surgery, pelvic inflammatory disease, or endometriosis, suggesting the lesion develops as a reactive inflammatory response to chronic serosal irritation [9-11]. The ultrasonography (US), Computed Tomography (CT) or Magnetic Resonance Scan (MRI) as diagnostic modalities can suggest the diagnosis, but the precise confirmation can be had only at surgery then the histopathology [12,13]. Management consists of surgical excision, which unfortunately is not always curative since recurrences have often been described [14]. We here reported two cases with such rare entity of BMPM admitted and managed in our pediatric surgery unit with a review of the literature.

Ethics Statement

Written informed consents were obtained from the patients parents who participated and managed in these two cases for publication of the reported cases and any accompanying images. The scientific and health committee in health directorate office approved this publication.

Case 1

A 4 years old male was admitted to our pediatric surgery unit with a preliminary diagnosis of appendicitis. For 15 days prior to admission, he had been complaining of a crampy abdominal pain, episodes of non-bilious vomiting, progressive abdominal distention, and constipation. Laboratory findings were not specific except for moderate anemia with hemoglobin of 9.1 g/dl. An erect plain abdominal x-ray image revealed small bowel loops with air fluid levels. Abdominal ultrasonography showed a large, multicystic reservoir filled with fluid full both right and left upper quadrants extending to the umbilicus. Abdominal contrast-enhanced Computed Tomography (CT) showed a 27x19x15 cm multicystic mass extending from the greater curvature of the stomach to the dome of the bladder with no solid components and no infiltration to adjacent organs. The patient was operated with a supraumbilical transverse incision. The operation revealed a cystic mass filled with dark brown-dark green serous fluid, which was attached to the greater curvature of the stomach with an 8 cm thick peduncle. Total excision of the mass was performed by clamping and cutting the narrow attachment (Figure 1, A-E). The histopathology study revealed that the cells lining the cysts stained positive for cytokeratin and calretinin markers on immunohistochemical stains and negative with CD31 and CD34. A diagnosis of benign multicystic mesothelioma was made (Figure 1, F). The patient had an uneventful recovery and was closely followed-up by US and CT. He remained free of symptoms and had no recurrence 16 months after surgery.

Case 2

A 28-month-old male presented with a few days history of abdominal pain, mild fever with twice episodes of non-bilious vomiting. Initial evaluations on admission revealed abdominal guarding, distended abdomen and a palpable mass felt in the right and left upper quadrants. Blood profiles revealed elevation in the ESR to 32 mm/hr, white blood cells WBC $16.0 \times 10^9/L$ and microcytic hypochromic anemia 8 g/dl. Blood biochemistry, renal function, and liver function tests were not specific apart from elevated C - Reactive Protein (CRP). The CT images confirmed the sonogram studies, revealed a $17 \times 13 \times 5.5$ cm abdominal mass or fluid collection filling the entire peritoneal cavity compressing the stomach, bowels and bladder (Figure 2, A-C). The patient underwent the surgical decision. Laparotomy through a supraumbilical transverse incision revealed a multicystic mass. It was comprised of numerous cysts ranging from 2 cm to 11 cm in diameter, filled with white serous fluid, which was attached to the greater curvature of the stomach with a 3 cm thick peduncle. It also had an attachment with the serosal surface of the transverse colon with an inflamed, partially necrotic, $4 \times 6 \times 2$ -cm segment of the greater omentum (Figure 2, D,E). The lesion was totally resected. Gross pathological studies revealed a multiloculated cystic mass measuring 16.5 cm in greatest dimension, involving the greater omentum. No hemorrhage with area of omental necrosis. Microscopic examination showed multiple cystic spaces lined by a single layer of cuboidal cells (mesothelial cells) with no significant pleomorphism or mitotic activity identified. The immunohistochemistry study revealed, the cells lining the cysts stained positive for cytokeratin and calretinin markers and were negative for CD31 and CD34 (Figure 2, F). Diagnosis of benign multicystic mesothelioma was submitted. The abdominal pain relieved, the patient clinically improved, and the ESR normalized on a postoperative day 12 to 11 mm/hr. The patient had an uneventful recovery and was closely followed-up by US and CT studies. He remained free of symptoms and had no recurrence 10 months after surgery.

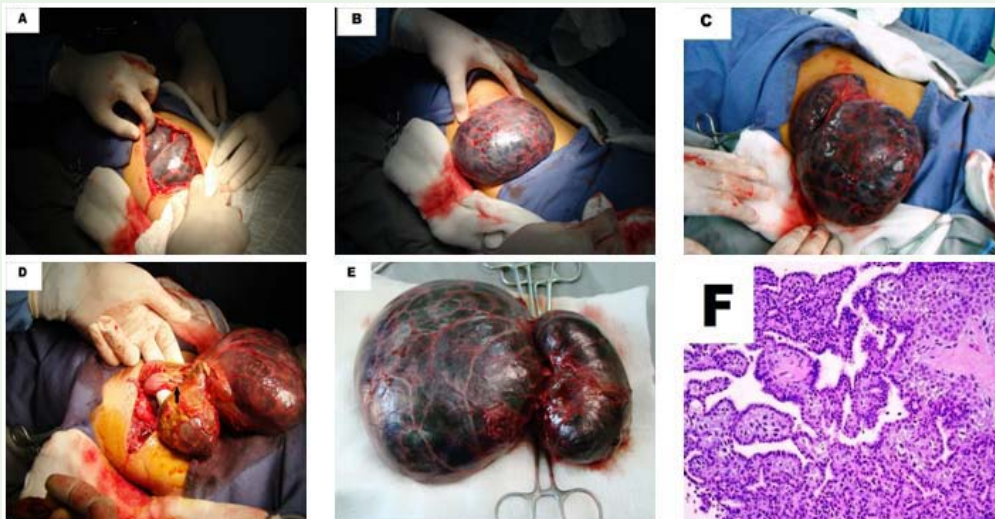


Figure 1: A-F: A- The patient was operated with a supraumbilical transverse incision.
 B-C: Operation revealed a cystic mass filled with dark brown-dark green serous fluid.
 D: Multiloculated cysts with a short segment stomach stalk (black arrow) plastered to its upper surface.
 E: Total excision of the mass was performed.
 F: Histopathology with the immunohistochemistry study revealed that, the cells lining the cysts stained positive for cytokeratin and calretinin markers.

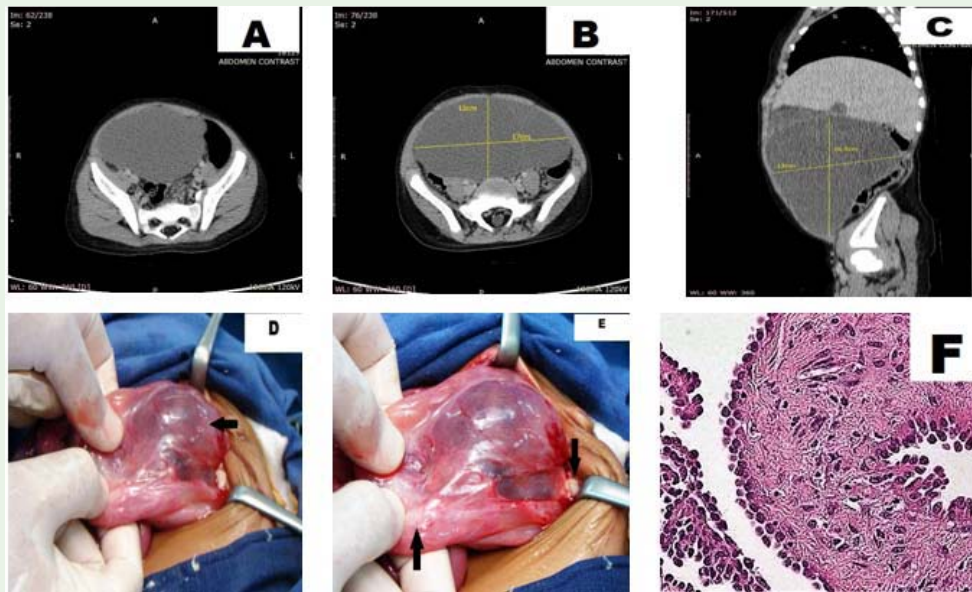


Figure 2 A-F: A-C: CT scan images revealed large cystic mass compressing the stomach, bowels and extended to the dome of the bladder. D: Perioperative image revealed large cystic mass filled with white serous fluid (black arrow). E: The mass attached to the serosa of the greater curvature of the stomach with small patch of omental necrosis (black arrows). F: Histopathological image revealed the cyst lined by numerous mesothelial cell, with no significant pleomorphism.

Discussion

Cystic mesothelioma is an uncommon tumor with a benign characteristic. Since it was first described a few case reports have been published in the literature including children [15]. There is no consensus about the etiology of BMPM, but neoplastic or inflammatory events have been hypothesized as a reason, the pathogenesis and pathological differential diagnosis remain unclear and controversial [16]. The rarity of such pathology made this challenge with its origin, pathogenesis, diagnosis, and therapy. The most common causes of admission to the hospital are abdominal pain abdominal distention, gastrointestinal complaints, and, less commonly, pelvic pain or a palpable mass. A mass can be determined in some cases, but the lesions can occasionally be incidentally discovered an encountered during laparotomy. An infrequent reason for ascites in childhood should be highlighting such pathology and must be kept in mind [17]. The preoperative diagnosis of benign cystic mesothelioma is difficult because of the nonspecific imaging. The tumor is clinically and radiologically indistinguishable from various benign and malignant mesenteric lesions. Ultrasound and CT scan do not differentiate benign cystic mesotheliomas from other cystic lesions. Fine-needle aspiration in most cases is not reliable and not so informative. Laparoscopy is considered in many centers the most accurate diagnostic method since it allows local biopsy of the suspected lesions, however, it is an invasive procedure. Usually, the confirmation of the diagnosis is submitted only at surgery and accomplished by histopathologic studies. For the time being, surgery is the only effective treatment for such lesions. Complete removal of the cysts if possible remains the mainstay of treatment and the only hope to avoid local recurrence. Most of these masses are usually large at the time of diagnosis (mean diameter, 13 cm) [18]. In our cases, the cysts were very large. Very few such cases of huge cysts have been reported [18]. Exploratory laparotomy reveals characteristic large

thin-walled transparent cysts, which are unevenly distributed in the serosa of the stomach parenchyma in both cases, often forming multicystic masses. Histological differential diagnosis among benign lesions includes cystic lymphangioma, cystic adenomatoid tumor, cystic forms of endosalpingiosis, endometriosis, müllerian cysts involving the retroperitoneum and cystic mesonephric duct remnants while malignant lesions include malignant mesothelioma and serous tumors of the peritoneum [19]. Multicystic mesothelioma should not be confused with malignant mesothelioma. This differential diagnosis is rarely a problem as the two pathologies exhibit marked differences in the macroscopic finding. Benign cystic neoplasm with which BMPM is most likely confused is cystic lymphangioma, which occurs more commonly in males and may develop in children as well [10]. Histology shows round or irregularly shaped multiple cystic spaces of various sizes lined by flat and cuboidal mesothelial cells and immunohistochemistry shows calretinin and keratin staining. Microscopic examination of the lesion reveals an inflammatory component in many cases. Immunohistochemically evaluations are useful in the differential diagnosis, the cells lining the BMPM cysts express markers of mesothelial differentiation including calretinin, Wilms' tumor antigen (WT-1) and keratin 5/6 [20]. By electron microscopy, the cells also display ultrastructural features of mesothelial cells including prominent surface microvilli, monofilaments and desmosomal junctions [21]. In some instance, when the lymphangioma enrolled in the differential diagnosis, it can be distinguished histologically from BMPM by the frequent presence of lymphoid aggregates and poorly developed fascicles of smooth muscle in the cyst walls. In diagnostically difficult cases (we encountered that in our second case), immunohistochemistry can assist in separating multicystic mesothelioma from lymphangioma as the later positively expresses endothelial markers such as CD31, CD34 and factor VIII-related antigen and is negative for keratin, while the opposite is true of multicystic mesothelioma [22].

Conclusion

Multicystic mesothelioma is an uncommon benign mesothelial proliferation. Histological examination with the aid of immunohistochemical techniques is essential for diagnosis. The prognosis is generally good. Benign multicystic mesothelioma, although in many situations it may lead to confused preoperative diagnosis, it should be kept in mind as an unusual cause for an abdominal mass in children.

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References

- Mennemeyer R, Smith M. Multicystic peritoneal mesothelioma: a report with electron microscopy of a case mimicking intraabdominal cystic hygroma (lymphangioma). *Cancer*. 1979; 44: 692-698.
- Moore JH Jr, Crum CP, Chandler JG, Feldman PS. Benign cystic mesothelioma. *Cancer*. 1980; 45: 2395-2399.
- Katsube Y, Mukai K, Silverberg SG. Cystic mesothelioma of the peritoneum: a report of five cases and review of the literature. *Cancer*. 1982; 50: 1615-1622.
- Gonzalez-Moreno S, Yan H, Alcorn KW, Sugarbaker PH. Malignant transformation of "benign" cystic mesothelioma of the peritoneum. *J Surg Oncol*. 2002; 79: 243-251.
- Clement PB, Young RH. Florid cystic endosalpingiosis with tumor-like manifestations: a report of four cases including the first reported cases of transmural endosalpingiosis of the uterus. *Am J Surg Pathol*. 1999; 23: 166-175.
- Ball NJ, Urbanski SJ, Green FHY, Kieser T. Pleural multicystic mesothelial proliferation: the so-called multicystic mesothelioma. *Am J Surg Pathol*. 1990; 14: 375-378.
- Drut R, Quijano G. Multilocular mesothelial inclusion cysts (so-called benign multicystic mesothelioma) of pericardium. *Histopathology*. 1999; 34:472-474.
- Lane TM, Wilde M, Schofield J, Trotter GA. Benign cystic mesothelioma of the tunica vaginalis. *BJU Int*. 1999; 84: 533-534.
- Villaschi S, Autelitano F, Santeusano G, Balistreri P. Cystic mesothelioma of the peritoneum. A report of three cases. *Am J Clin Pathol*. 1990; 94: 758-761.
- Ross MJ, Welch WR, Scully RE. Multilocular peritoneal inclusion cysts (so-called cystic mesotheliomas). *Cancer*. 1989; 64: 1336-1346.
- DeRosa G, Donofrio V, Boscaino A, Zeppa P, Staibano S. Multicystic mesothelial proliferation: immunohistochemical, ultrastructural and DNA analysis of five cases. *Virchows Arch A Pathol Anat Histopathol*. 1992; 421: 379-385.
- Pollack CV, Jordan RC: Benign cystic mesothelioma as acute abdominal pain in a young woman. *J Emerg Med*. 1991; 9: 21-25.
- O'neil JD, Ros PR, Storm BL, Buck JL, Wilkinson EJ. Cystic mesothelioma of the peritoneum. *Radiology*. 1989; 170: 333-337.
- Hafner M, Novacek G, Herbst F, Ullrich R. Giant benign cystic mesothelioma: A case report and review of literature. *Eur J Gastroenterol Hepatol*. 2002; 14: 77-80.
- Shakya VC, Agrawal CS, Karki S, Sah PL, Poudel P, Adhikary S. Benign cystic mesothelioma of the peritoneum in a child-case report and review of the literature. *J Pediatr Surg*. 2011; 46: 23-26.
- Safioleas MC, Constantinos K, Michael S, Konstantinos G, Constantinos S, Alkividas K. Benign multicystic peritoneal mesothelioma: a case report and review of the literature. *World J Gastroenterol*. 2006; 12: 5739-5742.
- McCullagh M, Keen C, Dykes E. Cystic mesothelioma of the peritoneum: a rare cause of 'ascites' in children. *J Pediatr Surg*. 1994; 29: 1205-1207.
- Weiss SW, Tavassoli FA. Multicystic mesothelioma. An analysis of pathologic findings and biologic behavior in 37 cases. *Am J SurgPathol*. 1988; 12: 737-746.
- Koyye R Tagore, Ramineni SA, Bhavani N, Ramachander S. A case of multicystic peritoneal mesothelioma. *Annals of Nigerian Medicine*. 2012; 6: 41-43.
- Sawh RN, Malpica A, Deavers MT, Liu J, Silva EG. Benign cystic mesothelioma of the peritoneum: A clinicopathologic study of 17 cases and immunohistochemical analysis of estrogen and progesterone receptor status. *Hum Pathol*. 2003; 34: 369-374.
- Bhandarkar DS, Smith VJ, Evans DA, Taylor TV. Benign cystic peritoneal mesothelioma. *J ClinPathol* 1993; 46: 867-868.
- Aboud MJ, Kadhim MM. Langerhanscellhisticcytosis (LCH) a presentation of two siblings with two different entities. *Springerplus*. 2015; 4: 351.