

## Case Report

# Benign cystic peritoneal mesothelioma: an abdominal daze

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### ABSTRACT

Benign cystic peritoneal mesothelioma (BCPM) is a rare clinical entity with varied presentations. Its obscure etiology and association with various intraabdominal conditions makes a precise diagnosis difficult. Diagnostic accuracy and diligent follow-up are essential because, though benign nature, it recurs locally. Herewith reporting a case of BCPM presenting as acute abdomen.

**Keywords:** Benign cystic peritoneal mesothelioma, Mesothelial cells

### INTRODUCTION

The benign cystic mesothelioma of the peritoneum is a rare lesion and is known for local recurrence. Although this lesion is typically benign, its high recurrence rate and potential for malignant transformation warrants diagnosis in a minimally invasive manner, and to formulate a surgical treatment strategy to avoid unnecessary re-operations.

### CASE REPORT

A 13-year-old girl presented to the emergency department with history of sudden onset of lower abdominal pain of 3 days duration. She gave a history of vomiting which was not blood or bile stained. She had no history of fever, dysuria, or bowel disturbances.

Examination revealed tachycardia, pallor, lower abdominal guarding and sluggish bowel sounds. Per vaginal and per rectal examinations were normal. Ultrasonography abdomen revealed 16 cm × 13.4 cm × 6.7 cm sized echogenic irregular ill-defined mass with cystic spaces, septations and calcifications, arising from the pelvis and extending into the supraumbilical region. Uterus was normal and ovaries could not be delineated clearly. An emergency plain computed tomography abdomen confirmed the above findings.

Patient was resuscitated and taken up for emergency laparotomy on view of persisting peritonitis. Laparotomy showed multicystic mass of size 15 cm × 13 cm arising from the pelvis extending up to the umbilical region, appearing as bunch of grapes with clear watery fluid, involving the anterior abdominal wall, pelvis, ovaries, uterus, mesentery of small bowel, transverse mesocolon and appendix. However, there was no luminal narrowing of small or large bowel. Stomach, duodenum, pancreas, spleen and liver were normal. No para-aortic or mesenteric nodes involved.

The tumor was resected except where inseparable from viscera. Post-operative period was uneventful and the patient was discharged home on 10<sup>th</sup> post-operative day. The histopathological report was “benign cystic peritoneal mesothelioma (BCPM).” Patient reported twice for follow-up in the last 6 months and there has been no clinical or radiological evidence of recurrence.

### DISCUSSION

BCPM is an extremely rare entity. Only about 130 cases have been reported in the literature worldwide.

In 1889 Henke<sup>1</sup> reported a case of “multiple cystic lymphangioma like tumor.” Over the next hundred years



various similar growths were described and it was only in 1980 that the term, “benign cystic mesothelioma,” was coined.<sup>2</sup> The mesothelial origin of such growths had been demonstrated by Mennemeyer and Smith in 1979.<sup>3</sup> It is also known as benign multicystic peritoneal mesothelioma or multilocular peritoneal cysts.

BCPM is a localized tumor arising from the epithelial and mesenchymal elements of the mesothelial cells, and does not metastasize.<sup>4</sup> The tumor can attach to the serosal surfaces of the intestine and the omentum or to the retroperitoneal space, spleen, and liver if located in the peritoneal cavity.<sup>5</sup>

The etiology remains obscure. In particular there is no association with asbestos exposure, previous abdominal surgery, or abdominal trauma.<sup>6</sup>

It shows predominant female preponderance, of reproductive age group. Peritoneal irritation caused by pelvic inflammatory disease, endometriosis, ruptured ectopic pregnancy, systemic lupus erythematosus may lead to this condition.<sup>7</sup>

The most common presentation is pelvic and low abdominal pain, but the lesions are sometimes incidental findings at laparotomy. Rarely, weight loss and a huge abdominal mass can be the presenting features.

Ultrasound abdomen can give useful information but computed tomography is the investigation of choice. Differential diagnoses include peritoneal lymphangioma, pseudomyxoma peritonei, and cystadenoma or cystadenocarcinoma of the ovary.

Aspiration cytology can be useful in making a preoperative diagnosis.<sup>1</sup> In general, the aspirate from a BCPM contains mesothelial cells showing focal presence of a brush border epithelium. Immunohistochemistry is also helpful in doubtful cases.<sup>8</sup>

Surgical resection is the treatment of choice. Marsupiation of the remaining cyst is done, where complete resection is impossible.

Grossly, cysts filled with mucinous or gelatinous fluid. Characteristically, the tumor is composed of multiple mesothelial-lined cystic structure, with fragile fibrovascular stroma holding the formation together.

It is not responsive to chemotherapy or radiotherapy. Prognosis is excellent, though there are chances of recurrence. Periodic follow-up for at least once in 3 months for first 2 years, 6 months in next 2 years and yearly

thereafter is needed. The reported recurrence rate is slightly higher in women (40-50%) than in men (33%).<sup>7</sup>

Cystic lymphangioma (cystic hygroma), cystic adenomatoid tumor, cystic forms of endosalpingiosis, endometriosis, Müllerian cysts involving the retroperitoneum, and cystic mesonephric duct remnants are included in the differential diagnosis among benign lesions. Malignant lesions include malignant mesothelioma and serous tumors of the peritoneum.

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