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CASE STUDY

Benign cystic mesothelioma: unknown cause of abdominal mass

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Abstract
Benign cystic mesothelioma (BCM) is a rare and benign disease that arises from the peritoneal mesothelium. It occurs predominantly in young to middle-aged women. The majority of cases were associated with a history of abdominal or pelvic operation, an endometriosis, and pelvic inflammatory disease. The etiopathogenesis is still unclear. Malignant transformation is extremely rare with only two cases reported in the literature. Like the etiology, the name of this entity is also controversial. Some authors prefer the term “peritoneal inclusion cyst” instead of “benign multicystic mesothelioma” and argue that the term mesothelioma should only be used when there is evidence of atypia. Most cases of BCM are discovered incidentally. Others reflect sequela of tumor mass effect. It appears intra-operatively as large, multi-focal, cystic lesions in the peritoneal and pelvic cavity. Diagnosis is achieved through surgical sampling with histopathological examination. Immunobiologically, BCM exhibits multiple small cystic spaces with flattened lining containing calretinin positive cells without atypical features, mitotic figures, or tissue invasion. Treatment includes cytoreductive surgery. We report on a 38-yearold woman who presented an abdominal mass discovered incidentally. Laparoscopic exploration revealed two cystic masses. The whole tumor was successfully excised and histopathology revealed benign cystic peritoneal mesothelioma.
Keywords: cystic peritoneal mesothelioma, abdominal mass, laparoscopy

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1 | INTRODUCTION

Benign cystic peritoneal mesothelioma (BCM), a rare tumor, occurs mainly in women in their reproductive age. The pathogenesis of BCM is unclear and a controversy regarding its neoplastic and reactive nature exists. BCM is generally considered a benign process; however, given the high rate of recurrence and possible malignant transformation, close followup is important. We would like to report a new case of BCM associated with celiac disease.

2 | CASE REPORT

We report the case of a 38-years-old women followed in our unit for celiac disease. She was asymptomatic and the systematic clinical examination showed an isolated abdominal painless mass in the left flank, measured approximately 20*30mm, fixed at the profond plane.

The blood analysis showed normal findings, leukocytes 5800/mm3, hemoglobin's 13,2 g/dl, and platelets 354.000/mm3. The biochemical test results were as follows: C-reactive protein negative, creatinine 0.83 mg/dl, with normal liver function. The tumor marker tests; CEA and CA 19-9 were negative. Abdominal ultrasonography showed two well-limited rounded hypoechoic masses, measuring 2.54 * 3.20cm and 0.79 * 1.07cm in the left flank.

Abdominal computed tomography (CT) scan revealed a mesenteric lesion on the left flank, oval, well limited, heterogeneous tissue and fluid, measuring 40 * 26mm (Figure 1), evoking a carcinoid tumor or a gastrointestinal stromal tumor.

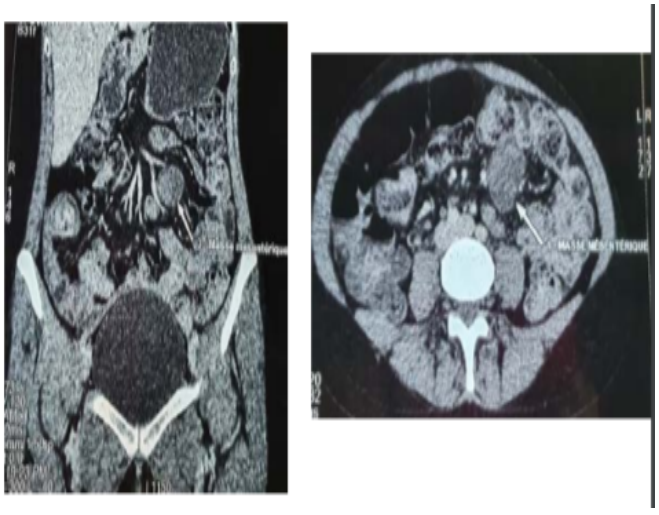

FIGURE 1: CT images: sagittal and axial views of the mesenteric lesion.

CT scan guided biopsy wasn't done to avoide the dissemination of malignant cells. An exploratory laparoscopy which confirmed the presence of two masses measuring 4 cm and 2 cm next to the superior mesenteric axis. The intervention was converted to open laparotomy in order to resect those masses (Figure 2).

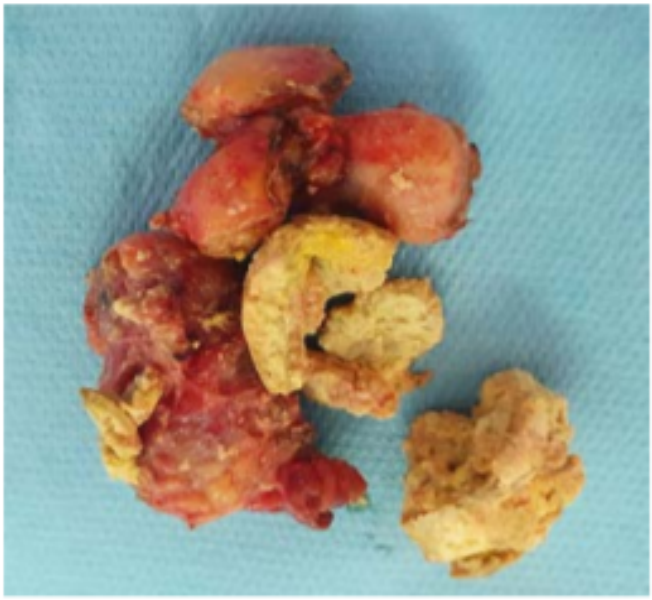

FIGURE 2: Operatory piece of mesenteric mass.

The postoperative outcome was uneventful and the patient was discharged after 2 days without compli-cations. Macroscopic examination revealed 8 formations, which one of them was a cystic mass of 4,5 cm in major axis with a fleshy yellow wall, and the others correspond to a fibrosis tissues. Microscopic examination showed cystic mass bordered by a sim-ple cubic coating, deprived of atypia and resting on a fibrous wall with few cells and crossed by a fine capillary network (Figure 3).

Supplementary information The online version of this article (https://doi.org/xx.xxx/xxx.xx) contains supplementary material, which is available to authorized users.

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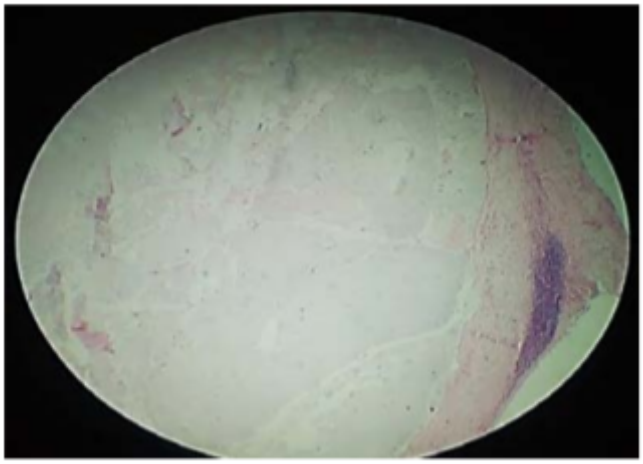

FIGURE 3: Histopathology results: cysts lined by cuboidal mesothelial cells.

At Immunohistochemistry the tumor cells were reactive for calretinin (Figure 4), and non-reactive for Ber-EP4 (Figure 5), consistent with benign multicystic peritoneal mesothelioma.

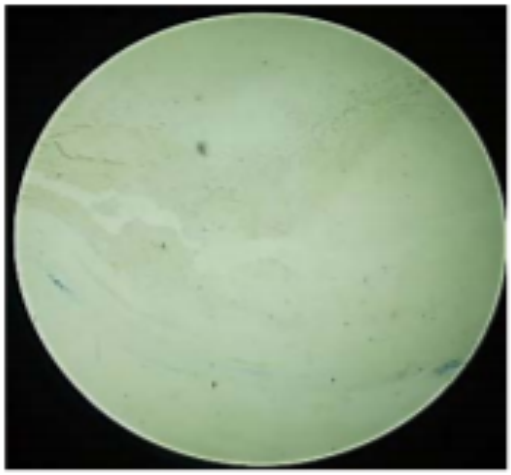

FIGURE 4: Immunohistochemically reaction for Calretinin

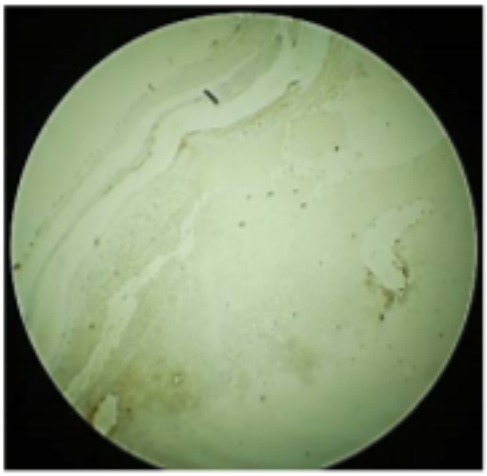

FIGURE 5: Immunohistochemically reaction for BerEP4.

3 | DISCUSSION

BCM, also known as multilocular peritoneal inclu-sion cysts, is an uncommon lesion arising from the peritoneal mesothelium that covers the serous cavity [1].

It is a rare disease that occurs most often in young women with an average age of 37 years and is associated with a history of prior abdominal surgery, endometriosis or inflammatory pelvic disease [1,2].

Until 2019, there are less than 200 documented cases worldwide, and it accounts for approximately 3-5% of the peritoneal mesotheliomas and the estimated incidence is 2 for 1,000,000 per year [2].

This tumor was originally described by Plaut in 1928 as a cyst of the pelvis after being discovered by accident during a surgery of uterine leiomyomas. In 1979, Mennemeyer and Smith first defined the lesion as a « multicystic peritoneal mesothelioma » in a 27- year-old female with multicystic diffuse lesion involving omentum, peritoneum, and pelvic viscera [3].

The pathogenesis of BCM is controversial due to its association with pelvic inflammatory disease (PID), endometriosis, leiomyoma, and a history of previous surgery, some authors believe that it has a possible reactive etiology. One of the hypotheses states that chronic peritoneal inflammation triggers the proliferation and migration of peripheral mesothelial cells and associated connective tissue, giving rise to these cysts. Some authors have proposed a neoplastic origin based on a slow but progressive growth of the lesions, a tendency to recur after surgical resection, and high disease related mortality in advanced stages [4,5]. A genetic and familial association of BCM has been reported by a few studies [4].

In addition, some studies suggest that female sex hormones play a role in its pathogenesis. This hypothesis is supported by the fact that BCM occurs mainly in females of reproductive age [2]. It has a strong predilection mostly for the surface of the pelvic viscera and serosal surfaces of the intestine and omentum or in the retroperitoneal space, spleen, and liver [3].

Malignant transformation is extremely rare, with only two cases reported in the literature, despite the potential recurrence [3,4].

Mostly, BCM is asymptomatic and is often discovered incidentally on imaging or during laparotomy for other indication [4,6]. Typical symptoms include abdominal distention, abdominal tenderness, ascites, nausea, and constipation, urinary symptoms, and dyspareunia [5,7]. When it reaches large sizes, it may present as a painful abdominal mass. In addition, its presentation in the form of acute abdomen had been reported [6]. Abdominal tenderness, abdominal distention, and palpable abdominal or pelvic mass may be present on physical examination [4].

On ultrasound BCM typically appears as a multicystic, vascular mass without calcifications (Figure 6). this aspect is named « spider in the web » [5,7].

CT scan typically shows a multicystic lesion with density similar to blood (Figure 7). It is used to evaluate the location and the extent of the cystic mass, where lesion appears as a low-density, multiloculated, and thin-walled multicystic mass. However, the results obtained by US and CT scans do not differentiate BCM from other cystic mass [2].

The definitive diagnosis is histopathological. The electron microscopy or immunohistochemistry can assist diagnosis. Serum tumor markers such as CA125, CEA, CA-15.3, CA-19.9, ferritin, 2- microglobulin are usually normals with one case reported, with elevated CA-19.9 with regression after surgical treatment [10,11].

The management protocol isn't consensual. While fine-needle aspiration could be used as a diagnostic tool, in most cases this method is not informative. Laparoscopy is the most accurate diagnostic method since it allows visualization, biopsies of the suspected tissue with intraoperative frozen section if possible, though an invasive procedure [3,6].

The BCM is characterized by solitary or multiple cysts, with fine septa, multiloculated and filled with serous fluid or blood. The size of lesions is few millimeters to 30 centimeters. The microscopy shows up multiple cystic spaces lined by cuboidal epithelium, there is no atypia or mitosis, cysts are separated by fibrous septa with areas of chronic inflammation and proliferation stromal cells [9]. There have been rare reports of transformation to low-grade malignant mesothelioma, but it is generally classified as a benign process [6,7].

The immunohistochemically study is important to exclude possible mimic neoplasms cystic. Generally positive markers include calretinin, D2-40, CK5/6 and WT-1, mesodermal markers present in epithelia, especially the mesothelium. Markers MOC-31, PAX8, BG8, Ber-EP4, B72.3, CEA, and CDX-2 are frequently negative. Most often used an epithelial marker (cytokeratin), and CD34, an endothelial marker that can also be used to exclude lymphangioma or other suspected vascular neoplasms [9,7].

The differential diagnosis of BCM includes benign and malignant cystic or multicystic abdominal tumors such as cystic lymphangioma, endometriosis, cystic forms of endosalpingiosis, cystic adenomatoïd tumor, pseudomyxoma peritonei, malignant peritoneal mesothelioma. This last is a major differential in the diagnosis of BCM, and distinguishing benign and malignant mesothelioma is very crucial to patient care [4,10].

Due to its rarity, BCM treatment options remain an area of controversy and there is no streamlined treatment plan. Currently aggressive surgical resection is the mainstay [10].

Alternative treatments such as hormonal therapy, sclerotherapy, and potassium-titanylphosphate laser vaporization have been proposed in the recent studies. Hormonal therapy with anti-estrogen drugs like tamoxifen and GnRH agonist can be an alternative to surgery in selected patients with estrogen-dependent neoplasm, as they were showed to be associated with a decrease in cyst volume. Laser vaporization with potassium titanyl-phosphate laser were shown to have a significant efficiency at penetrating the tumor, but its therapeutic efficacy is still unknown. Sclerotherapy consists of injecting povidone–iodine

or ethanol through a catheter directly into the cyst [2,11].

The treatment of choice for multicystic mesothelioma is total surgical excision. Chemotherapy and radiation therapy are generally not indicated, because benign nature of this process. Some authors have proposed heated intraperitoneal chemotherapy as a possible treatment option, but there is inadequate evidencebased research on this topic [11,12].

Although BCM is associated with a favorable prognosis in a short term, because it is a completely benign condition; in long term it has been described about 50% after a period of 3 to 27 months (mean 32 months). There is a 40–55% recurrence rate in female patients and a 33% recurrence rate in male patients. No risk factors predicting the recurrence of BCM have been identified yet [5,7,9].

There is evidence that malignant transformation could occur. There are two cases described of malign processing [7].

A long follow-up period is always necessary, although there is still no consensus on when and what additional tests should be ordered. Some authors suggest that CT should be done every 3 months for the first year after resection and then annually for the next 5 years [4].

4 | CONCLUSIONS

BCM is a rare benign neoplasm that is associated with no well-defined symptoms, typical clinical, or imaging data. The diagnosis remains unclear and the definitive diagnosis requires histology and immunohistochemistry. BCM has a high risk of recurrence and a significant potential of malignant transformation.

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