

Benign Multicystic Peritoneal Mesothelioma in a Male Teen: Case Report and Review of the Literature

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The case of a Peruvian 15-year-old male with a left-flank abdominal mass suspected to be a peritoneal pseudomyxoma is presented. The patient underwent a R0 surgery, and the pathology review showed a benign multicystic peritoneal mesothelioma. Characterized by recurrent mesothelial peritoneal cysts originating in the epithelial and mesenchymal elements of mesothelial tissue, this benign tumor is unusual among young males. [P R Health Sci J 2020;39:222-225]

Key words: Benign multicystic peritoneal mesothelioma, Multilocular peritoneal inclusion cyst, Intra-abdominal tumor

Mennemeyer and Smith first described a benign multicystic peritoneal mesothelioma (BMPM) in 1979; although usually considered benign, this tumor has a high rate of local recurrence (1, 2, 3). Also known as multilocular peritoneal inclusion cyst, postoperative peritoneal cyst, and inflammatory inclusion cyst of the peritoneum, a BMPM is a rare benign tumor that presents with recurrent peritoneal mesothelial cysts which arise from the epithelial and mesenchymal elements of the mesothelial tissue (4, 5, 6). The incidence of the disease is 0.15/ 100 000 per year (4). This lesion occurs most frequently in women during their reproductive years and is associated with a history of previous abdominal surgery, endometriosis, or pelvic inflammatory disease (1, 4). To date, there are fewer than 200 reported cases in the literature. To our knowledge, this is the first report on a male patient in Latin America. For that reason, there is little evidence on its management and prognosis (4, 5). BMPM rarely occurs in young males, making this presentation of a 15-year-old male with a BMPM significant.

Case presentation

An otherwise healthy 15-year-old male from Lima, Peru, presented with a 1-month onset of abdominal pain, nausea, and vomiting. The physical examination showed a patient in good condition. Upon abdominal examination, a refractory mass was palpated in the right flank, lower quadrant; no testicular masses were found. The tumor markers were within normal limits (CA 19-9: 3.85 U/mL; CEA: 0.79 ng/ml). An abdominal computed tomography (CT scan) (Figure 1) showed an extensive, hypodense, homogeneous tumor with defined borders and internal septa. It crossed the midline and displaced internal structures without infiltrating them. The tumor measured 30 x 25 cm at its greatest width and length, respectively, and was suspicious of pseudomyxoma peritonei.

The patient underwent surgery: An exploratory laparotomy was performed, in which an extensive intra-abdominal tumor was found. It was characterized by thin borders with diffuse vascularization, trabeculae, with large and small clusters, the largest of which measured 30 x 25 cm. The tumor depended from the peritoneum of the right iliac fossa; it displaced the intra-abdominal organs without infiltrating them. A R0 resection was achieved; it consisted of a resection of the intra-abdominal tumor, a right iliac fossa partial peritonectomy, and an appendicectomy; in addition, suspicious nodules on the right pelvic wall were resected. A frozen-section biopsy of the mass was reported as lymphangioma; however, as a peritoneal pseudomyxoma was suspected, an appendicectomy was included in the procedure.

The histologic examination of the intra-abdominal tumor showed multiple cystic formations with fibrous septa, loose connective tissue, and inflammatory infiltrate with eosinophils and neutrophils (Figure 2). The cystic formations were covered by flat mesothelial and cuboidal cells filled with eosinophilic material; no pleomorphism or cellular atypia was observed (Figure 2). The peritoneum of the right iliac fossa and the appendix showed severe mixed inflammatory infiltrates, but no malignancy was found. The immunohistochemistry revealed that the intra-abdominal tumor was positive for calretinin and negative for CD34 (Figure 3); with these findings, the diagnosis of BMPM was made.

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Figure 1. The CT shows an extensive (28 x 22 cm) neoplastic hypodense lesion that is homogeneous and has defined borders.

The post-operative outcome was uneventful, and the patient was discharged after 3 days. At the 1-year follow-up, the chest and abdominal CT scans showed that the patient was free of recurrence.

Discussion

The pathogenesis of BMPM is unclear, with chronic inflammatory and indolent neoplastic causes proposed by various authors. The current consensus says it is not a true neoplasm, but a reactive proliferation caused by trauma, surgery, or chronic inflammation (6, 7). Another theory (known as

the hormonal hypothesis) holds that BMPM's sensitivity to sex hormones might be responsible for its development and progression. This theory is supported by the higher incidence of BMPM in women who are in their reproductive age, this kind of tumor's estrogen hormone receptor expression, and the responsiveness of this tumor to tamoxifen and gonadotropin-releasing hormone analogs (2, 4, 8).

The clinical presentation of a BMPM is generally aspecific. The most common presenting symptoms are chronic or intermittent abdominal pain and tenderness and distension due to the mechanical compression of adjacent intra-abdominal organs, as was the case in our patient; even acute abdomen has

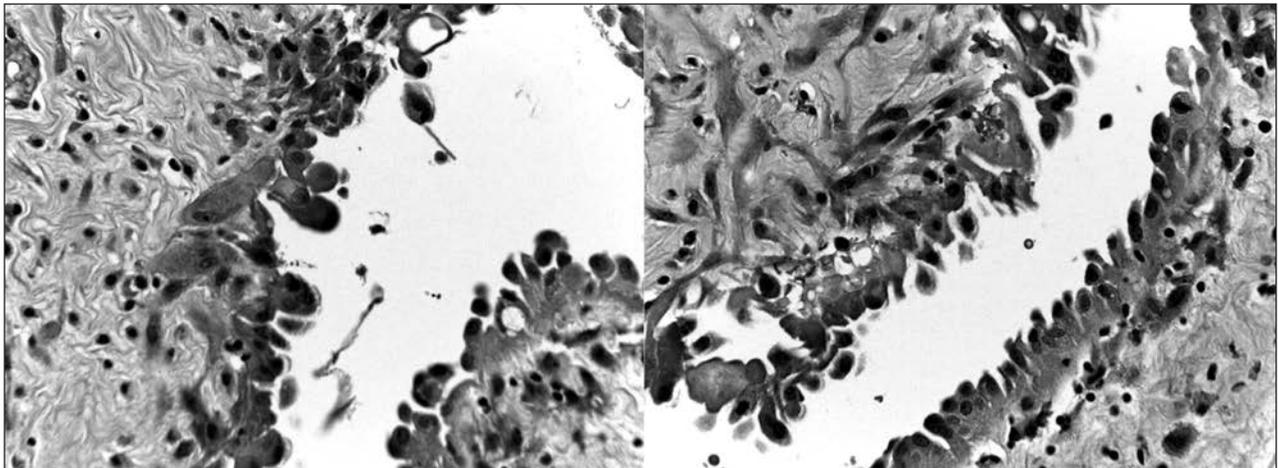


Figure 2. Benign multicystic peritoneal mesothelioma microscopy with H&E: flat mesothelial cells alternating with cuboidal cells.

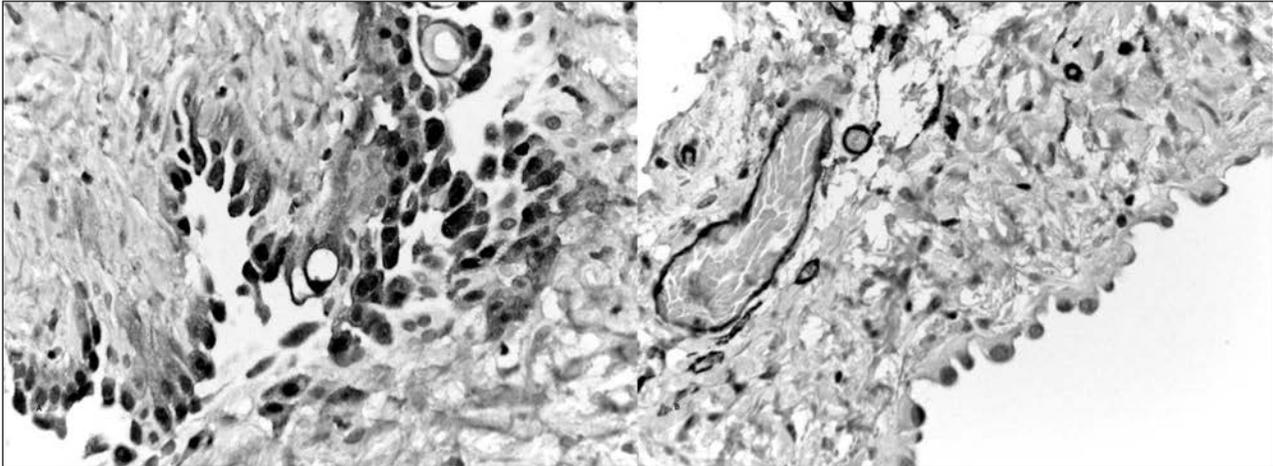


Figure 3. Benign multicystic peritoneal mesothelioma immunohistochemistry. A. Calretinin: positive; B. CD 34: negative.

been reported in the literature (1, 5, 6, 9). Tumor sizes ranging from 3 to 20 cm have been described. To our knowledge, this is the largest BMPM tumor reported (10, 11). An association between BMPM and increased serum CA 19-9 concentrations has been described; however, the tumor markers were within normal limits in our patient (9). Available imaging techniques such as ultrasound, CT scan, and MRI were able to show the lesion. In the ultrasound images, the BMPM could be seen as a multicystic mass, with vascularized septa but without calcification. On the CT scan, a multicystic mass with lobes of different density could be seen, while the MRI showed a multicystic lesion with areas of low and intermediate intensity in T1-weighted images and intermediate high intensity in T2-weighted images (5). However, none of the images previously indicated could be used to make a definitive diagnosis of a BMPM as opposed to some other cystic multilocular lesion (5).

The most important differential diagnoses of BMPM (as it relates to benign lesions) include cystic lymphangioma and adenomatoid tumor. In contrast, malignant lesions include malignant mesothelioma and invasive serous tumors of the peritoneum (5, 8). For the most part, a lymphangioma consists of a cystic mass that both is multiloculated and contains chylous fluid, bundles of smooth muscle, and aggregates of lymphocytes in its walls. The cystic adenomatoid tumor often has a solid component. Malignant mesothelioma usually affects older males with a history of asbestos exposure. It presents as multiple small nodules in addition to cystic structures and shows malignant features, such as infiltrative growth and cellular atypia (9, 12). In this case a peritoneal pseudomyxoma was suspected, due to the CT scan finding of a predominant right iliac fossa abdominal mass whose origin was thought to be the appendix. This is a rare malignant neoplasm characterized by the accumulation of mucus in the peritoneal cavity. It is currently accepted as being caused by the rupture of a low-grade appendiceal mucinous neoplasm with the subsequent dissemination of its mucus-producing cells across the peritoneal surface; generally, peritoneal nodules and omental caking are present (9, 13, 14).

The macroscopic and microscopic findings agreed with those of the literature, in that the presence of cystic lesions lined by mesothelial cells demonstrated immunohistochemically positivity for calretinin and negativity for the vascular marker CD34 and excluded the possibility that the patient had a cystic lymphangioma. Lymphangiomas are benign tumors that are found in children, typically, and only rarely in adults; they are histologically characterized by the presence of large and irregular vascular spaces lined by smooth and flat epithelial cells with fibroblastic stroma or collagen (13, 14). In contrast, malignant tumors such as malignant mesotheliomas are rarely entirely cystic or histologically stratified; moreover, pleomorphism and nuclear atypia are present in such tumors and generally have an aggressive course (5, 15). Finally, the microscopic evaluation was consistent with a BMPM and a non-tumoral appendix.

The current treatment for a BMPM consists of its complete surgical excision. Chemotherapy and radiation therapy do not have a clear role due to the benignity of this tumor. Some authors suggest that cytoreductive surgery, followed by the instillation of hyperthermic intraperitoneal chemotherapy, can provide better disease control than can debulking surgery alone, considering the high rate of recurrence and the possibility of malignant transformation (9). But strong evidenced-based information about this topic is lacking. As mentioned, the recurrence risk remains high: of those women and men who underwent complete cytoreduction, approximately 50% of the former and 33% of the latter experienced recurrence of their tumor (15). Recently, Noiret et al, proposed novel treatment options that include hormonal therapy, sclerotherapy, and potassium-titanyl-phosphate laser vaporization (16). In addition, some authors have proposed extrapolating the treatment for BMPM from malignant mesothelioma, but data are missing (15–17). The follow ups are usually made with periodic abdominal CT scans, but the impact on overall survival has yet to be established (4). The prognosis of a given BMPM is generally good, with a 5-year survival rate of 100%, despite the high recurrence rate and the low rate of malignant transformation (1, 2).

Conclusion

Finding a BMPM in a male teen is extremely rare. This benign reactive lesion can mimic different entities, including serous tumors of the peritoneum and cystic lymphangioma. Therefore, a histopathologic examination and an immunohistochemistry panel are necessary for the final diagnosis. Surgery is the current treatment for a BMPM, with a high risk of local recurrence, even if the tumor is completely excised.

Resumen

Se presenta el caso de un varón peruano de 15 años con una masa abdominal en el flanco izquierdo que sospechosa de pseudomixoma peritoneal. El paciente se sometió a una cirugía R0 y la revisión de la patología mostró un mesotelioma peritoneal multiquístico benigno. Este es un tumor benigno raro entre los hombres, que se presenta como quistes peritoneales mesoteliales recurrentes y que surgen de los elementos epiteliales y mesenquimales del tejido mesotelial.

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