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# Peritoneal Polycystic Mesothelioma (PPM): Need and Place of Second Look

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## Abstract

Polycystic péritonéal mesothelioma (MPP) is a rare and benign condition. Its treatment is surgical. After surgical resection, MPP tends to recur in the peritoneal cavity. Our observation concerns a 30-year-old patient with a history of non-specific abdominal pain evolving for a year. She was operated on urgently and benefited from a resection of cystic lesions disseminated in the peritoneal cavity in addition to an appendectomy. The peritoneal cancer index (PCI) was estimated at 12 points. The postoperative course was simple. The histological study was in favor of a polycystic péritonéal mesothelioma. Despite the fact that the patient was asymptomatic and a normal CT scan 3 months postoperatively, a second look was retained and the patient was operated on again. Residual cystic lesions of 04 to 15mm were highlighted and resected. This surgery was associated with intraperitoneal chemotherapy based on 150mg of Cisplatin over 5 days. The postoperative course was simple and the patient was discharged from the service on the 10th postoperative day. A 3rd intervention by laparoscopic approach was carried out five years later for the exploration of primary sterility. The latter found a cyst of the left ovary and a hydrosalpinx which were resected She is alive and without recurrence at 108 months. Conclusion: Faced with polycystic péritonéal mesothelioma for which there is doubt about radical resection, additional surgery using a second look

should be considered. This attitude makes it possible to avoid residual recurrence, which is quite frequent in this disease.

**Keywords**
Polycystic mesothelioma; Complete radical resection; Tumor recurrences; Second-Look.

## Introduction

Polycystic peritoneal mesothelioma (MPP) is a rare condition, representing 30% of mesothelioma. MPP is a form of mesothelioma first described in 1979 by Mennemeyer and Smith [1]. Its management is surgical and requires radical resection of all the lesions [2].

## Case Report

This is a 30-year-old patient who has no medical or surgical history other than nonspecific abdominal pain that has been evolving for a year. She was admitted urgently for an acute appendicular syndrome to another department of a regional hospital. Intraoperative exploration found cystic lesions disseminated throughout the peritoneal cavity and more concentrated at the level of the greater omentum. These predominantly translucent lesions range from a few millimeters to several centimeters (Fig. 1, 2 and 3). The peritoneal cancer index (PCI) was estimated at 12 points. An appendectomy was performed with partial omentectomy and resection of other lesions on request. The postoperative course was simple. The histological study of the surgical specimens was in favor of an MPP. The clinical examination, the biological examinations and the postoperative abdomino pelvic computed tomography (Fig. n°4 and n°5) were without abnormalities. Faced with the risks of resection of an incomplete resection of the lesions and after discussion with her surgeon, we decided to operate the patient again 3 months after the intervention.

The intraoperative exploration of this 2nd intervention after the realization of a peritoneal cytology, highlighted the presence of some loose adhesions between the large remaining omentum and the abdominal wall as well as the presence of 5 to 6 cystic lesions next to the right colic angle, another located on the pavilion of the left fallopian tube and finally another smaller one on the pavilion of the right fallopian tube. These lesions were between 4mm to 14mm in diameter. Other lesions in the form of firm nodules are located on the greater omentum. An additional omentectomy was performed combined with resection of all suspicious lesions. The intervention was completed by the placement of drains for the performance of immediate postoperative intraperitoneal chemotherapy combining ascites with 2.5 liters of physiological saline and 75mg of Cisplatin after removal of 10cc of peritoneal fluid for a cytological study at the end of the procedure intervention. Chemotherapy was followed over 4 postoperative days with emptying and filling of the peritoneal cavity with 2 liters of saline and 25mg of Cisplatin. The postoperative course was simple and the patient was discharged from the service on the

10th postoperative day. The histology study confirms the diagnosis of benign polycystic mesothelioma on 6 resected lesions while on others, it was postoperative inflammatory reactions. The peritoneal cytology at the start and end of the procedure was benign with the presence of only active and regular mesothelial cells. The postoperative course of this 2nd intervention was also simple. Five years after the 2nd operation, this patient was explored for primary sterility with the discovery of an ovarian cyst of the left ovary and an ipsilateral hydrosalpinx. A laparoscopic approach was performed for a right ovarian cyst associated with hydrosalpinx. A cystectomy and dissection of the hydrosalpinx was performed. This intervention had simple consequences. This patient is alive without recurrence at 108 months.

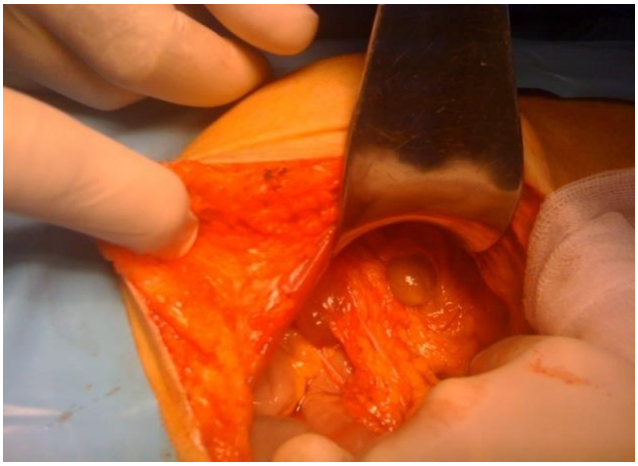

**Figure n°1:** Operative view (First surgery).

## Comments

MPP is a benign condition whose etiology is currently unknown and only hypotheses are put forward. It occurs mainly in women of childbearing age and is under the influence of female hormones [3]. But cases in young men have been reported [4,5]. Malignant transformation has been reported [6].

Macroscopically, it can present from the form of a single multilocular cyst to several cysts, the most frequent form as in our observation and giving rise to the rather evocative "cluster" appearance. The cysts have a thin translucent wall and contain a serious or gelatinous fluid, more or less hemorrhagic. These cysts vary in size from a few millimeters to more than 30 cm. The cysts are separated by conjunctivovascular partitions which can harbor a population of more or less abundant acute or chronic inflammatory cells, as well as haemorrhagic changes. In these partitions, we sometimes find isolated mesothelial cells, arranged in nests or organized in small glandular structures. These sometimes-vacuolated mesothelial cells can simulate a bezel-ring appearance.

Microscopically, cysts are most often lined by a single layer of mesothelial cells. Microscopically, cysts are most often lined by a single layer of flattened or cubic mesothelial cells. The cells are sometimes hyperplastic with some atypia identical to those observed during reactive mesothelial proliferations. These cells may also form small papillary projections in the lumen of cysts. We can also find areas of adenomatoid or squamous metaplasia. More rarely, the mesothelial cells desquamate and float in the lumen of the cysts. The immunohistochemical study highlights the mesothelial profile of these cells which express the cytokeratins AE1E3, Cam 5.2, Calretinin, EMA and HMBE1 and do not express ACE, CD

15 and0020BerEP4 [7]. The etiology of polycystic mesothelioma remains obscure. It is not linked to exposure to asbestos, but certain factors seem to favor its outbreak such as previous abdominal or pelvic surgery, locoregional inflammatory pathology, endometriosis, suggesting for some authors the "reactive" nature of this lesion rather than neoplastic [8].

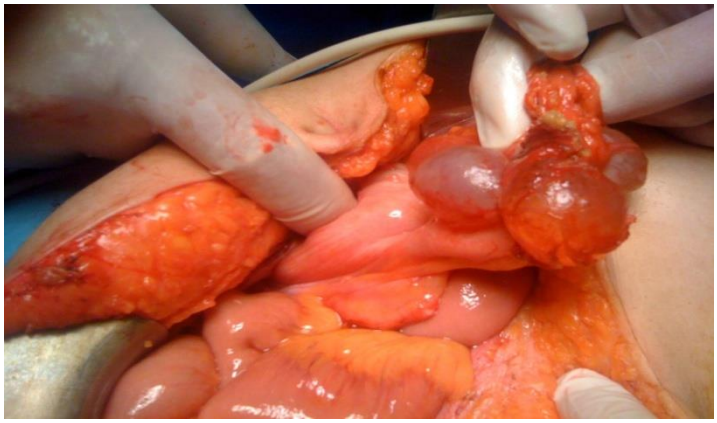

**Figure n° 2**: Operative view (First surgery**)**

Clinically, PPM can manifest as abdominal pain or a picture of palpable masses [9]. The clinician often confuses it preoperatively with an ovarian tumor or a pseudomyxoma in advanced forms, especially in front of advanced forms. The differential diagnosis through the macroscopic aspect is essentially represented by the cystic lymphangioma of the peritoneum and the standard histological study and immunohistochemistry decide. In favor of the latter, they are endothelial cells of uniform size with a wall containing a few muscle bundles and quite a few lymphocytes. In immunohistochemistry, the lymphangioma variably expresses CD31, CD34, and factor VIII. It should be kept in mind that from the macroscopic form, the histology and especially the immunohistochemistry decide in favor of the different differential diagnoses. It should be noted that the diagnosis is established on the bundles of clinical, macroscopic, morphological, intraoperative, histological and immunohistochemical arguments.

The positive diagnosis of MPP is based on morphological examinations such as echotomography (ECT), computed tomography (CT) and magnetic resonance imaging (MRI). ECT can show a multilocular hypoechoic image whereas CT usually shows hypodense images with thin septa, typically with a multilocular cyst appearance. CT can provide information on the precise location and extent of these lesions. On MRI, which is the examination of choice, mesothelioma appears as hypointense on T1 and hyperintense or with an intermediate signal on T2, with moderate enhancement on injection of gadolinium into its walls [10]. In fact, the positive diagnosis requires in all cases the histological examination with immunostaining.

The therapy of MPP calls for complete eradication surgery to prevent recurrence, which is relatively frequent. For some, it is only a radical surgical resection [11]; for others, it must be associated with chemohyperthermia (CHIP) or immediate postoperative chemotherapy (CIPPI) [12, 13]. The MPP is for some authors incorrectly described as a "benign" tumor because it has a strong tendency to recur more

or less quickly (50% of cases), requiring multiple surgical interventions. The degeneration is exceptional.

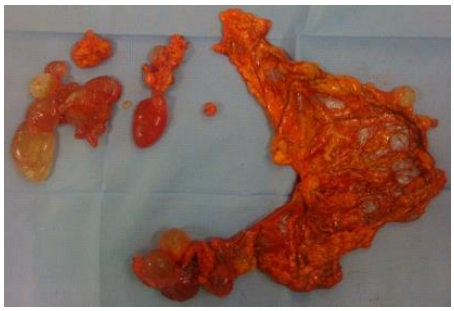

**Figure n°3:** Surgical specimen (First surgery).

Our observation clearly illustrates the possibility of recurrence insofar as 6 tumor foci were left in place after the first intervention. Incomplete resection is the main reason for recurrence in our eyes. A. Singh et al [14] reported a case of mesothelioma resected twice as in our observation. This is the main reason for the high rate of recurrence after surgery for us. We chose to perform additional resection surgery associated with CIPPI despite the absence of clinical signs in the patient and normal CT after the first operation. The histological results after the 2nd intervention proved us right. Indeed, a second-look should be required in any patient if there is no certainty of a complete resection of the lesions during the first surgery even if the clinic and the morphological examinations do not show anything.

## Conclusion

MPP is a benign and rare condition. These clinical manifestations are essentially represented by non-specific abdominal pain. The modern morphological exploration helps in the diagnosis of strong presumption but the diagnosis of certainty remains histological. Therapeutic management is surgical and the latter must be complete to give the patient every chance of not reccur. In the absence of complete resection after resection, additional surgery must be performed associated with intraperitoneal chemotherapy.

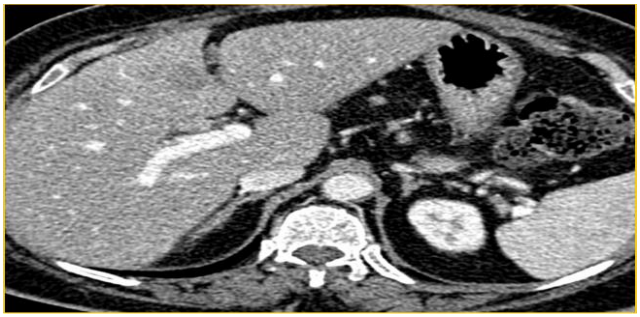

**Figure n°4:** Normal CT scan before the second surgery.

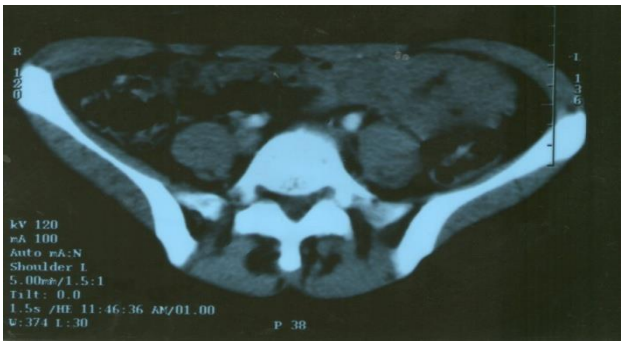


**Figure n°5:** Normal CT scan before the second surgery.

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