

Benign cystic mesothelioma of the peritoneum: benign neoplasm, not a benign course

Mehdi Benna¹, Asma Sassi², Nadia Boujelbane², Mohammed Ali Ayadi¹,
Sonia Seghaier³, Karima Mrad², Monia Hechiche¹, Khaled Rahal¹

¹Surgical Oncology Department, Salah Azaiez Institute, Tunis, Tunisia

²Pathology Department, Salah Azaiez Institute, Tunis, Tunisia

³Radiology Department, Salah Azaiez Institute, Tunis, Tunisia

Abstract : *Benign cystic peritoneal cystic mesothelioma is a benign neoplasm with high recurrence rate and aggressive behavior. We report a 38 years old female patient who had surgical resections three times in the two last years and we discuss the pathological features and therapeutics options of this disease.*

Keywords: *mesothelioma, cyst, surgery, intraperitoneal chemotherapy, recurrence,*

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I. Introduction

Benign cystic mesothelioma of the peritoneum (CMP) was first described in 1979 by Smith and Mennenmeyer [1]. Since then, less than 200 cases were reported in the literature. It is a rare peritoneal tumor involving the abdomen, pelvis and retroperitoneum with a strong predilection for the peritoneum of pelvic organs [2]. This tumor is misleading as there are several differential diagnoses of which, pseudomyxoma peritonei, cystic lymphangioma and cystic teratoma are the commonest. Although it's benign behavior, CMP is known for local recurrence and rare malignant transformation. Surgery remains the standard of care.

II. Case Report

A 38-year-old previously healthy female patient was referred to the surgery department in January 2016 for a right iliac fossa pain. Physical examination displayed tenderness in the right iliac fossa. Ultrasound revealed peritoneal effusion in the pouch of Douglas and a cystic mass depending on the appendix. The diagnosis of acute appendicitis was suspected. Patient had a laparoscopy revealing multiple cystic masses around the right iliac fossa, which were removed. The patient had an uneventful post-operative course and was discharged **5 days later**.

Grossly, cysts measured between 1 to 2.5 cm in greatest dimension. They had a grape-like appearance, a smooth surface and were thin-walled filled with clear and watery fluid. Histopathological examination revealed an adipose tissue containing cysts lined by cuboidal or flattened mesothelial cells (Fig. 1). Neither hyperplasia nor nuclear atypia was noted. The connective fibrous tissue contained some lymphocytes. On immunohistochemistry, the lining epithelium of cysts displayed an intense and nuclear immunoreactivity for WT1 and calretinin (Fig. 2, Fig. 3). These histological and immunohistochemical findings enabled us to come to a final diagnosis of a benign cystic mesothelioma. The patient had two pelvic recurrences in September 2016 and July 2017 in each case had a debulking surgery.

III. Discussion

Benign cystic mesothelioma of the peritoneum, also referred as mesothelial inclusion cysts, mainly occurs in reproductive age women with a median age on time of presentation of 36 years old in 80% of cases [3]. Its pathogenesis is still unclear but CMP is considered to be a reactive benign mesothelial change secondary to chronic inflammatory conditions such as a history of endometriosis, foreign body fibers and dusts and mechanical injuries [4]. Our patient was young and aged 38, but did not have a past medical history.

Although it is benign, CMP may be associated with appendicitis, peritoneal pseudomyxoma or primary ovarian carcinoma [5]. In this context, it is more likely to be a result of reactive mesothelial hyperplasia. Clinical examination is nonspecific and patients may present with abdominal distension, pain or mass. Sometimes, it can be asymptomatic and incidentally discovered during a routine check, on imaging or intraoperatively [3].

Imaging investigative tools including ultrasound, CT scan and MRI cannot make an accurate diagnostic as they cannot differentiate CMP from cystic peritoneal lesions [6].

On gross examination, CMP is polycystic with a grape-like appearance and cysts are thin-walled and filled with a watery and clear fluid. A definite diagnosis is made by histology, which reveals cysts in the adipose and fibrous peritoneal tissue lined by mesothelial cells. The underlying connective tissue may display inflammation. In up to one third of the cases, adenomatoid or squamous metaplasia is encountered [7]. Histologically, the main differential diagnosis is cystic lymphangioma. The latter is lined by flat endothelial cells and contains lymphoid tissue and smooth muscle in cyst walls [8]. On immunohistochemistry, CMP is positive for WT1, calretinin and cytokeratins. Nuclear immunoreactivity for WT1 and calretinin is mandatory to confirm the mesothelial origin of the tumour. Cystic lymphangioma is positive for endothelial/lymphatic markers as CD31 and D2-40. Besides, clinical presentation is important as cystic lymphangioma commonly occurs in children with head and neck or retroperitoneal location [9].

Treatment consists of multiple aggressive surgical resections. Nevertheless, patients have a like hood of recurrences up to 50% and usually suffer from chronic abdominal and pelvic pain. Several adjuvant treatments such as systemic chemotherapy, radiation therapy, and sclerosis therapy with tetracycline were reported with poor results. The optimal cure is probably cytoreductive surgery eliminating all gross disease with intra-peritoneal chemotherapy [7].

IV. Conclusion

Benign cystic peritoneal mesothelioma is a rare and benign tumor with a high recurrence rate. It's aggressive behavior and the need of several surgical resections could alter the quality of life. Intra peritoneal chemotherapy shows promising results and may be the cure for this "no so benign disease".

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Figure 1: Histological examination of the cyst: adipose tissue containing multiple cysts lined by a cuboidal epithelium (Hematoxylin and Eosin X 40).

Figure 2: Nuclear immunoreactivity of mesothelial cells for WT1

Figure 3: Nuclear immunoreactivity of mesothelial cells for calretinin

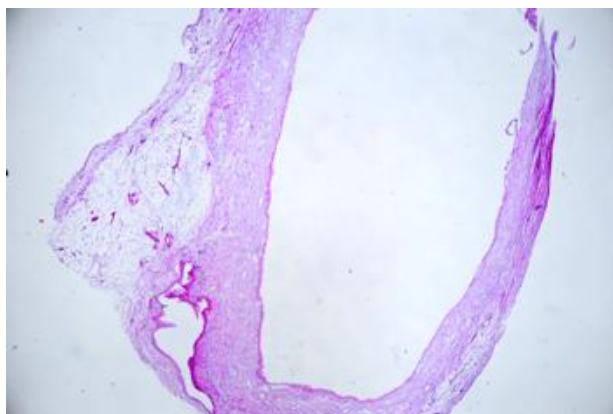


Figure 1: Histological examination of the cyst: adipose tissue containing multiple cysts lined by a cuboidal epithelium (Hematoxylin and Eosin X 40).

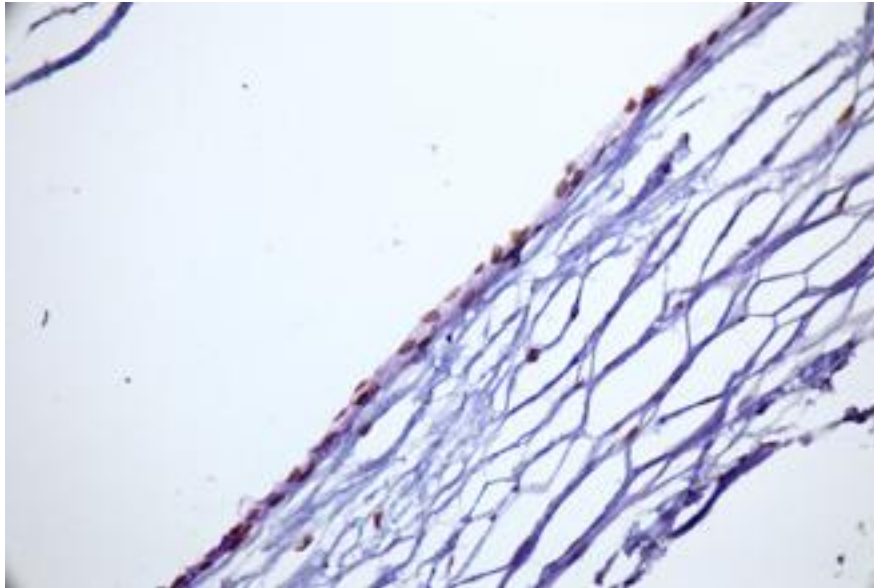


Figure 2: Nuclear immunoreactivity of mesothelial cells for WT1

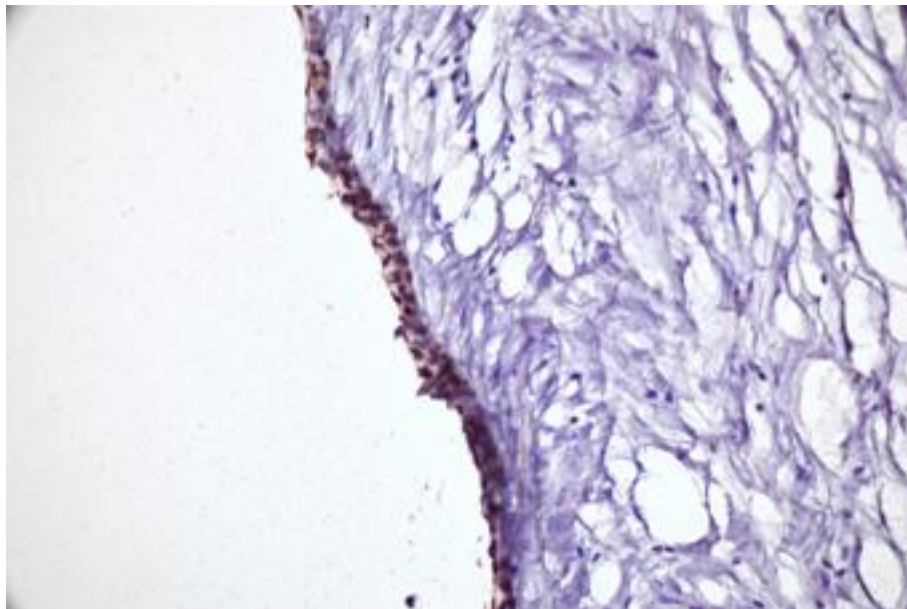


Figure 3: Nuclear immunoreactivity of mesothelial cells for calretinin

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