Appendicular Mucocele Simulating Ovarian Tumor Case and Review of the Literature

S.Belaazri¹, FZ.Lamine², T.Berrada², H.Hachi³, A.Baidada², A.Kharbach¹

¹Service Obstetrics Gynecology and Endocrinology M3 Maternity Souissi, Chu Ibn Sina, Rabat, Morocco
²Service Obstetrics Gynecology and endoscopy M1 Maternity Souissi, Chu Ibn Sina, Rabat, Morocco
³Breast gynecological Pole of the National Institute of oncology, Chu Ibn Sina, Rabat, Morocco

Corresponding Author: Sofiane Belaazri
sbelazri23@hotmail.com

ABSTRACT
Mucocele appendicular or tumor mucosécrétante appendicular is a rare disease, defined as a cystic dilation of the light Appendix following intraluminal accumulation of mucinous secretion, translucent, gelatinous. The clinical picture may take several forms. Radiology has an important place for diagnosis. Treatment is primarily surgical; adjuvant treatment is reserved for repeat offenses. We report a case of appendicular mucocele simulating an ovarian tumor collects in the breast gynecology division of the National Institute of Oncology Rabat.

Keywords: Appendicular mucocele, Ovarian tumor, Imaging, Treatment

1. INTRODUCTION
Mucocele appendicular (MA) or mucosécrétante tumor of the appendix is defined as a cystic dilation of appendicular light after an intraluminal accumulation of mucinous secretions, translucent, gelatinous, may affect either the entire body or a segment, usually distal¹. It was described for the first time in 1842 by Rokitansky who then decided to call it "hydrops appendicular" since it has attracted the interest of many authors². We report a case of the appendicular mucinous ovarian tumor and peritoneal extension, treated at the National Institute of Oncology in Rabat. The aim of our work is to clarify the clinical diagnosis, diagnostic and therapeutic modalities of appendicular mucocele.

2. CASE
Patient 58 years old, diabetic on insulin for 14 years, operated for cataract and cholecystectomy a year ago. G7P5 menopause 14 years ago. The onset of symptoms was in 4 months before hospitalization, by the appearance of pelvic pains without bleeding, with urinary frequency. The all moving in a conservation context of the general state. The gynecological examination: healthy-looking collar, no palpable adnexal mass, parameters are free.

The abdominal, pelvic ultrasound revealed the presence of a large cystic multilocular 46 x 41 mm, associated with a siled pelvic ascites, heavy suspicion of a right ovarian tumor with peritoneal carcinomatosis. The magnetic resonance imaging revealed a peritoneal carcinomatosis of ovarian tumor (Figure 1, A, B&C). The biological a CA125 has been requested; it has been slightly increased to 43UI / l.

Fig.1: Abdomen MRI shows peritoneal carcinomatosis

A laparotomy was performed: exploration presence of gelatinous ascites, a large appendicular mucocele. A bilateral oophorectomy with appendectomy was performed. The specimen was sent for pathological study, which was in favor of a low-grade mucinous appendiceal neoplasm associated with an extension to the peritoneum and the surface of the two annexes. The postoperative course was simple. The patient was scheduled for a monitored based on clinical examination, CT, and a dosage of ACE and CA19-9 every six months to 5 years; then once a year for ten years, but the patient lost sight

3. DISCUSSION

The appendicular mucocele is a rare condition estimated at 0.2 to 0.3% appendectomies\(^3\). It represents between 7 and 8% of appendicular tumors. The sex ratio is variable, but it seems that the disease is more common in women with a ratio of four women to one man in the series Aho et al.\(^5\). The age of onset varies from 15 to 71 years\(^6\) with an average age of 55 years. Our patient was a 58-year-old.

It corresponds to an expansion of the appendix by secondary mucus buildup to obstruction of its light is of mechanical origin in case of ligation or stercolith or by locoregional reached in benign tumors: endometriosis, villous adenoma or malignant: carcinoma, carcinoid tumor\(^4\).

Clinically, in most cases, it is a chronic pain in the right iliac fossa (48 to 64% of cases). This pain can be intermittent or simulate an array of acute appendicitis. In 18 to 32% of cases, the mucocele is revealed by a palpable mass in the right iliac fossa\(^7\). More rarely, it is manifested by acute intestinal obstruction\(^8\), lower GI hemorrhage\(^9\) or peritonitis\(^10\). Rarely, appendicular mucocele may be manifested by urinary symptoms type: urinary frequency, urinary urgency, hematuria, hydronephrosis, pyelonephritis. This being related to the tumor size and the compression of the ureter and bladder\(^10,11,12\). We found in the literature a case of appendicular mucocele manifested by vaginal mucoides\(^13\). Our patient had pelvic pain with urinary manifestations types of urinary frequency. The simultaneous existence of an ovarian mucinous tumor, whether benign or malignant, and MA is estimated at between 8 and 18% of the cases\(^18\). In fact, these lesions can cause peritoneal Pseudomyxoma (PMP)\(^18\). The appendicular or ovarian origin if PMP has long been discussed, currently immunohistochemical studies have demonstrated the appendicular origin of almost all cases, with a possible extension ovarian secondary\(^14\). The ovarian disease is usually unilateral, and it’s right ovary that is most
affected. In our case, the pathological study revealed the achievement of two annexes. Imaging plays a key role in the diagnosis. At the ultrasound, it looks like an oblong mass of the iliac fossa hypoechoic right with fine echoes, parietal calcifications are possible\(^{(3,4)}\). The scanner is the most efficient examination. It shows a very limited hypodense mass which more or less calcified wall enhances after contrast medium injection. It allows searching peritoneal dissemination of signs with a dense presence of ascites or peritoneal nodules\(^{(3,4)}\). The often massive gelatinous ascites, compartmentalized, lobular, distorting the intestines it can have the appearance of a solid ascites. In our patient, the ultrasound objectifies a large multilocular cyst associated with a siled pelvic ascites, evoking a right ovarian tumor with peritoneal carcinomatosis. The magnetic resonance imaging shows if mucocele appendicular, cystic mass hypointense on T1-weighted images, and hyperintense on T2-weighted images, with a thick wall, enhancing gradually and uniformly after injection of gadolinium\(^{(14)}\). MRI is only useful in evaluating the extension of peritoneal pseudomyxoma in the aftermath of a perforation of the mucocele\(^{(3)}\), it shows the gelatinous masses, hepatic and splenic scalloping, septa intraascitiques\(^{(15)}\) and can differentiate tissue implants in the peritoneal effusion T2 in sequence. In our case, the imaging magnetic resonance revealed the presence of carcinomatosis. The differential diagnosis will be done with a cyst or ovarian neoplasia; a cecal diverticulum, appendiceal abscesses, inflammatory bowel disease, mesenteric cyst or omental well as a hematoma or mesenteric tumor\(^{(2)}\). The case of our patient was diagnosed as an ovarian tumor with ultrasound and MRI. The diagnosis of appendicular mucocele was made intraoperatively. Biologically Serum CEA is increased in AD cases neoplastic, it decreases after the reductive surgery and thus be a means of surveillance to detect early recurrence of cystadenocarcinoma. The increase of CEA and CA19-9 would be in favor of peritoneal carcinomatosis mucinous\(^{(16)}\). The CA-125 may be useful in search of a disease of the peritoneum if it is high\(^{(16)}\). Several authors recommend the determination of CEA, CA19-9, and CA-125, but little information on this in the literature. These markers, especially CA 19-9 may be useful in monitoring the disease and diagnosis of tumor recurrence\(^{(17)}\). The biology, therefore, has no interest in the positive diagnosis of AD. Our patient underwent a CA125 assay has been slightly increased which may indicate an associated peritoneal or ovarian aggression. Treatment has three main goals: evacuate ascites; find and treat tumors or initials and peritoneal lesions; if possible prevent recurrences and complications. Surgery is, for almost all of the authors, the "treatment of choice" of the gelatinous disease of the peritoneum. It meets two objectives:

- totally evacuate ascites and perform a complete cleaning of the peritoneal cavity (some gelatinous masses sometimes being very adherent, it may be necessary to resect certain viscera);
- practicing excision of the primary tumor, possibly extended according to the data of intraoperative histological examination.

A new surgical approach was described by Sugarbaker\(^{(19,20)}\) advocating immediately maximal surgery improves survival. In all cases, the first track must be broad, above and below the navel, to allow treatment of the primary lesion and a complete peritoneal toilet. Laparoscopy has little place in this pathology. The evacuation of gelatinous ascites should be as complete as possible. It must be careful; it can be facilitated by the use of mucolytic agents and repeated washes in serum. The maximum surgery or "debulking surgery" aims to eliminate macroscopic signs of gelatinous disease\(^{(19)}\). Maximum surgery includes a series of péritonectomies procedures that remove all disease parietal peritoneal surfaces and resect all visceral peritoneal surfaces that are involved.

If Benin cystadenoma of the appendix, a simple appendectomy is sufficient. Some authors propose to achieve it systematically when there is a PGM associated with a mucinous ovarian tumor, although the appendix is macroscopically normal. For some, when the pseudo myxoma peritoneal appendicular origin (or cystadenocarcinoma tumor border) is diagnosed, a systematic bilateral oophorectomy should be considered, in all women or only in postmenopausal women. Life expectancy is increased if the removal of the two sites is performed. Chemotherapy: Chemotherapy should be administered only when there is a clinical tumor recurrence or malignant forms\(^{(21)}\). Platinum derivatives and adriamycin are recommended when the gelatinous disease of the peritoneum is ovarian origin while...
preference is given to mitomycin C and 5-FU when origin appears colic or appendicitis. 

Chemotherapy delivered by intraperitoneal route has the advantage of tumor tissue intraperitoneal (little or vascularized at the beginning of their growth) in contact with high concentrations of cytotoxic agents by limiting systemic concentrations and therefore the risk of toxicity\(^{23}\). 

The chemo-hyperthermia intraperitoneal (CHIP): It has been shown that hyperthermia increased the efficacy of certain molecules (mitomycin C, cisplatin, oxaliplatin), or by increasing their cytotoxicity or increasing their penetration into the tumor tissue\(^{24,25}\). 

Proteolytic enzymes: Peritoneal lavage with proteolytic enzymes (trypsin, hyaluronidase, 5% dextrose, dextran sulfate at 10%) was used to sometimes get good results\(^{15}\). 

Radiotherapy: Some authors consider known unnecessary or even harmful because it would foster the emergence of flanges, fibrosis and increase the risk of developing intestinal obstruction. Radiation therapy is of limited value in preventing the re-accumulation of mucus\(^{15}\). 

Other treatments: The estrogen supplement has been used from time to time with inconsistent effect. 

Intraperitoneal interferon therapy after chemotherapy has been attempted but has not been effective\(^{48}\). Treatment with intraperitoneal mitomycin C absorbed on “activated carbon particles” is not safe in patients who have undergone surgery\(^{15}\). The hyper chronic intermittent power helps maintain digestive rest for long periods while allowing higher nutrient inputs to the daily ration theoretical\(^{22}\).

4. CONCLUSION

The appendicular mucocele is a rare condition, generally postoperative histological discovery. However, with advances in imaging, preoperative diagnosis is possible based mostly on the couple echo-CT This is essential in order to adapt the surgery. CEA is essential for the monitoring and detection of recurrence. 

Simple appendectomy is the rule in most cases, but for some, when the pseudo myxoma peritoneal appendicular origin is diagnosed, a systematic bilateral oophorectomy should be considered, in all women or only in postmenopausal women. Other therapeutic alternatives are being evaluated.

REFERENCES
