ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 11, Issue, 11, pp.8552-8556, November, 2019

DOI: https://doi.org/10.24941/ijcr.37349.11.2019

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

RESEARCH ARTICLE

WHAT TREATMENT IN APPENDICEAL MUCOCELE?

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ARTICLE INFO

Article History:
Received 24th August, 2019
Received in revised form
08th September, 2019
Accepted 25th October, 2019
Published online 30th November, 2019

Key Words:

Mucocele,
Reatment.

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ABSTRACT

Introduction: Appendicular mucocele is a rare disease and constitutes 0.2-0.3% of surgically excised appendicitis, more frequently in females (3: 1) and with an average age of 55 (2.4 , 6.12). We distinguish four subtypes characterized with non-specific clinical manifestations. The study proposes, through clinical observation, a focus on the current diagnostic and therapeutic orientation of the pathology. **Materials and Methods:** From January 2017 to December 2019 consulted the database of the AOU "G Rodolico" University of Catania Department of medical surgical and specialist sciences II 4 patients were treated of which 3 female cases and one sex case male with an average age of 38 (range 41-35) years with appendicular mucocele. the clinical symptomatology was characterized by constipation, asthenia, malaise, rapid weight loss and anemia with abdominal pain arising on average, increasing in localization on the right quadrants with ipsilateral lumbar irradiation. The preoperative diagnosis was difficult. **Results:** Blood tests were found to be normal, in particular neoplastic markers, with the exception of CEA> 50 in cistoadenocarcinoma. The surgery showed the presence of a voluminous appendicular mucocele with a maximum diameter of 12 cm and a transverse diameter of 8 cm, without evidence of regional lymphadenomegaly or infiltration of neighboring organs; an appendectomy was performed with complete exeresis of the cystic formation. **Discussion:** The mucocele of the appendix is characterized by the slow accumulation of mucinous material inside its lumen, and includes a heterogeneous group of lesions ranging from the simple accumulation of mucoid material to mucosal hyperplasia, to the cystadenoma up to the cystadenocarcinoma, with an incidence of approximately 0.4% of all cancers of the appendix.). In the cases described by us, the diagnosis was placed, as often happens, in the suspicion of a subacute appendix. The histological examination showed the presence of a proliferation of epithelial cells with abundant mucin-like material (fig3). The laparoscopic approach is preferred to the laparotomic approach to minimize the risks of bowel rupture and dissemination in the peritoneal cavity of mucus-secreting epithelial cells (1, 11, 12). In the case in which the approach in videolaparoscopy is chosen, the appendix can be enclosed in an endo-bag or in a glove in order to prevent the escape of mucus or the rupture of the neoplasm and of the viscus (2). between the mucocele and a concomitant gastroenteric tumor; the mucosa hyperplasia and the appendicular cystadenoma are in fact considered as the hyperplastic polyp of the large intestine, with the possibility of developing areas of dysplasia with a high degree of malignancy. **Conclusion:** Although it is a rare disease, the appendicular mucocele must be taken into consideration when facing an appendicular mass, in order to be able to implement during the intervention the precautions aimed at minimizing the risk of dissemination of mucosal epithelial cells in the peritoneal cavity and therefore onset of pseudomixoma peritoneal.

Citation: Graziano Giorgio Maria, Paolo Antonino Buffone and Antonino Graziano. **2019.** "What treatment in appendiceal mucocele?", *International Journal of Current Research*, 11, (11), 8552-8556.

INTRODUCTION

The appendicular mucocele is a rare disease and constitutes 0.2-0.3% of the surgically removed appendicitis, more frequently in the female sex (3: 1) and with an average age of 55 years (1,2,3 , 4). We can distinguish four subtypes characterized by non-specific clinical manifestations, accompanied by a distension of the bowel from accumulation of mucus in its interior; Possible causes are: retention of an obstruction to emptying due to faecal and congenital anomalies, epithelial hyperplasia, cystadenomas (63-84%) and cystadenocarcinomas (11-20%). (5,6,7,8,) The distension of the lumen is progressively more marked in the four subtypes, up to the possible rupture of the viscus with consequent dissemination in the peritoneal cavity of mucus-secreting epithelial cells, configuring the formidable picture of the pseudomixoma peritoneal (9 , 10,11,12), characterized by

gelatinous ascites and multifocal epithelial implants with copious secretion of mucin (13,14,15). Furthermore, it can also be associated with other neoplasms, such as colon tumors and mucin-secreting ovarian tumors (16,17,18,19,20); there are also described in the literature cases of association with ulcerative colitis (11) and with cystic pancreatic neoplasms (21.22.23.24.25). The clinical presentation is varied. In 25% of cases the appendicular mucocele is asymptomatic and the diagnosis is made during investigations performed for other reasons (26,27,28,29,30). Otherwise it can occur with a framework that simulates an acute appendicitis; in some cases it appears as a palpable mass (31,32,33,34). Finally it can present as a complication, such as occlusion obstruction, enteric hemorrhage, compression hydronephrosis on the right ureter, in the presence of pseudomixoma peritoneal (35,36,37,38). The differential diagnosis is made with acute appendicitis, Meckel's diverticulum, colitis, diverticulosis, hydrosalpinge, ovarian cysts, mesenteric cysts, enteric duplication cysts (39.40.41). clinical observation a focus on the current diagnostic and therapeutic orientation of the pathology.

MATERIALS AND METHODS

From January 2017 to December 2019 consulted the database of the AOU "G Rodolico" University of Catania Department of medical surgical and specialist sciences II 4 patients were treated including 3 female cases and a male case with average age of 38 (range 41-35) years affected by appendicular mucocele. the clinical symptomatology was characterized by constipation, asthenia, malaise, rapid weight loss and anemia with abdominal pain arising on average, increasing, located in the right quadrants with ipsilateral lumbar irradiation. Preoperative diagnosis was difficult (1, 3, 10, 11, 14). Laboratory tests showed no significant alterations. Only in the case of cystadenocarcinoma was there an increase in CEA. The direct radiograph of the abdomen showed the presence of a mass located in the lower right quadrant, with dislocation of the viscera. Abdominal echotomography (Fig 1) showed a capsulated cystic lesion, communicating with the cecum, with or without calcifications. MRI showed the presence of a low intensity cystic mass in T1-weighted and high intensity images in T2-weighted ones. The colonoscopy highlighted the "sign of the volcano", characterized by the finding of a soft and erythematous mass with at the center a crater from which mucus comes out. The needle-aspirate was contraindicated due to the risk of cell dissemination in the peritoneal cavity. The abdominal CT resolutive analysis to place the diagnosis showed a bulky mass (8-13 cm longitudinal diameter and 6-8 cm of transverse diameter), encapsulated , hypo-anechoic, with an onion shell appearance and the presence of a solid prolongation starting from the capsule itself and jutting out inside the lumen. This mass did not seem to have continuity relations with the neighboring organs and seemed to preserve cleavage plans with respect to the surrounding structures. with the contrast medium, the presence of a longitudinal diameter of 8-12 cm was confirmed, with homogeneous liquid content, with regular walls with parietal calcifications (Fig. 2), located in the context of the right mesocolon.

RESULTS

Blood tests were normal, in particular neoplastic markers were negative, with the exception of CEA> 50 in cistoadenocarcinoma.

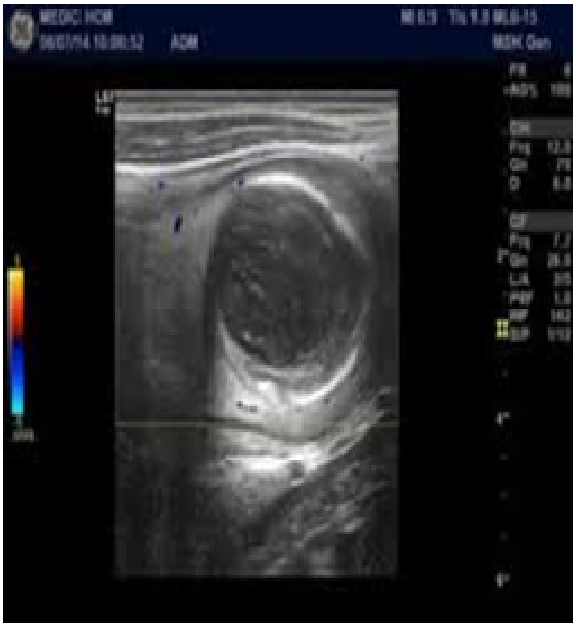

Fig. 1. Echo appendicular mucocele

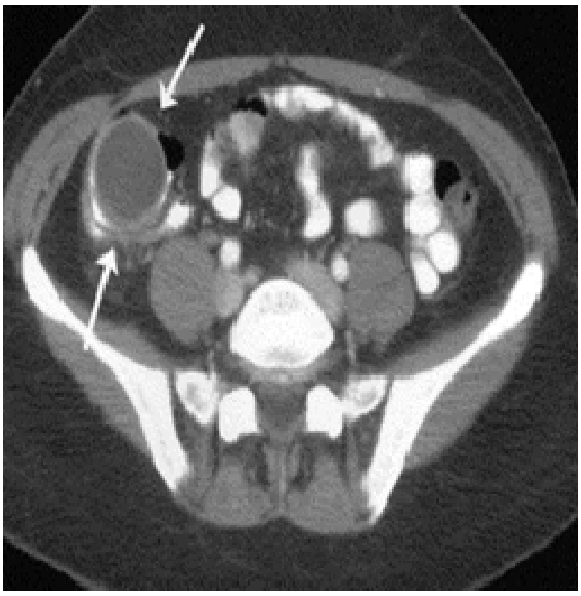

Fig. 2. Ct appendix mucocele calcifications

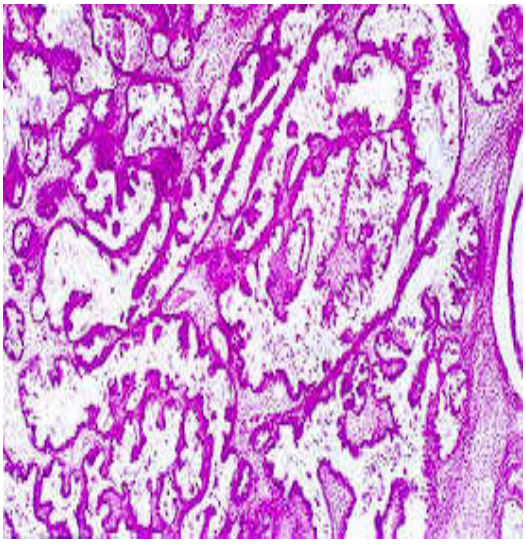

Fig 3. cistoadenocarcinoma

The CT exam proved to be decisive in the preoperative period, in order to determine the diagnosis, and the contiguity with the other organs. The surgery showed the presence of bulky appendicular mucocele with a maximum diameter of 12 cm and a transverse diameter of 8 cm, without evidence of regional lymphadenomegaly or infiltration of neighboring organs; an appendectomy was performed with complete exeresis of the cystic formation. The definitive histological examination showed in 3 cases it was a mucinous tumor of the "coherent" cecal appendix with mucinous cystadenoma, with exeresis margin on typical mucosa and containing abundant dense mucoid material, and in one case cistoadenocarcinoma. (Fig 3) it proceeded after extemporaneous examination of the lesion which confirmed its malignancy to a right hemicolectomy. The postoperative course was regular with discharge in 4 postoperative day. A colonoscopy performed two weeks after surgery excluded synchronous colonic neoplasms.

DISCUSSION

The mucocele of the appendix is characterized by the slow accumulation of mucinous material inside its lumen, and includes a heterogeneous group of lesions ranging from the simple accumulation of mucoid material to mucosal hyperplasia, to the cystadenoma up to the cystadenocarcinoma., with a incidence of about 0.4% of all the tumors of the appendix. The clinical onset of this pathology is generally aspecific and heterogeneous, including from vague abdominal disorders with localized pain mainly to the right quadrants of the abdomen, up to the presence of a palpable mass or peritonitis. Some cases report a correlation between the type of symptomatology and the characteristics of the neoplasm, correlating the weight loss or the presence of an abdominal mass to the malignancy of the tumor (42,43,44). The instrumental examinations allow today to place the diagnosis with relative certainty. The appearance of this lesion is in fact characterized by a frankly hypodense and well-delimited cystic image, with some calcifications on the CT scans (fig 2), while the typical echographic pattern is represented by the "onionskin sign" of the English-speaking authors, which represents a pathognomonic sign.

The confirmation, in fact, of a cystic mass occupying the right abdominal quadrants with an onion-like echographic pattern, in the presence of uterine appendages of regular morphology in the woman, so as to make preoperative diagnosis of appendicular mucocele (45.46.47). The treatment of the appendicular mucinous cystadenoma is essentially surgical and must be evaluated based on the histological picture and the extent of the disease. therefore the type of intervention to be performed is linked as a function of the identified subtype, including the simple resection of the mucocele in the case of the mucinous cystadenoma, which represents the most frequent variant, up to the right hemicolectomy. case of malignant variants. However, it is necessary to remember the possibility of a possible intraoperative rupture of the mucocele both spontaneous and accidental, with the consequent insemmentation in the peritoneal cavity of tumor cells, thus configuring the peritoneal pseudomixoma. A precise preoperative diagnostic definition is therefore important in order to plan the correct surgical procedures. In this regard, as far as the exeresis of the tumor is concerned, next to the case studies that report the classic advantages of the laparoscopic approach also in this type of pathology, some authors report cases of rupture of the appendicular mucocele with relative peritoneal dissemination and need for subsequent re laparotomic intervention (48.49.50). In the cases described by us, the diagnosis was placed, as often happens, in the suspicion of a subacute appendix. The histological examination showed the presence of a proliferation of epithelial cells with abundant mucin-like material (fig3). The laparoscopic approach is preferred to the laparotomic approach to minimize the risks of bowel rupture and dissemination in the peritoneal cavity of mucus-secreting epithelial cells (51,52,53). In the case in which the approach in videolaparoscopy is chosen, the appendix can be enclosed in an endo-bag or in a glove in order to prevent the escape of mucus or the rupture of the neoplasm and of the viscus (54,55,56). In particular, for the mucinous cystadenoma the treatment of choice is appendectomy; only in the case of forms with a large base of implant and located in correspondence with the appendicular or extended base to the caecum can blind indication or right hemicolectomy be found (57.58,59); for the mucinous cystadenocarcinoma we can limit ourselves to appendectomy if the mass is small and localized in the two distal thirds of the appendix, otherwise we resort to the right hemicolectomy (69 61.62).

In any case it is good to check both ovaries and the colon to rule out the presence of other neoplasms. In the case of pseudomixoma peritoneal, a more frequent situation for malignant forms, intra-peritoneal and systemic chemotherapy are associated with radical surgical therapy. Sometimes bilateral ovariectomy is necessary. The prognosis of the mucinous cystadenoma of the appendix is good, even in case of extension outside the appendix, with a survival of 91-100%. The 5-year survival drops to 25% in malignant forms. It is also necessary to remember the relative frequency, reported by some authors, of the association between the mucocele and a concomitant gastroenteric tumor; the mucosa hyperplasia and the appendicular cystadenoma are in fact considered as the hyperplastic polyp of the large intestine, with the possibility of developing areas of dysplasia with a high degree of malignancy. For this reason it is recommended, if the diagnosis of mucocele is not made during the operation, to perform a post-operative endoscopic examination of the large intestine that allows the exclusion of synchronous colonic neoplasm.

Conclusion

Although it is a rare disease, the appendicular mucocele must be taken into consideration when facing an appendicular mass, in order to be able to implement during the intervention the precautions aimed at minimizing the risk of dissemination of mucosal epithelial cells in the peritoneal cavity and therefore of occurrence of pseudomixoma peritoneal. This very serious condition can indeed be established in a subtle way even after some time; ultrasound diagnostics, completed by a CT scan, allows certain preoperative diagnosis to be made. Surgery must guarantee adequate exeresis, therefore a minimally invasive open surgery is preferred.

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