Mucinous Adenocarcinoma of the appendix with Retroperitoneal and Pelvic Bone Extension - A Case Report and Literature Review

Alamdari NM1, Gholizadeh B2 and Kimia F3

1Assistant Professor of General and Vascular Surgery, Shahid Modarres Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran
2Assistant Professor of General Surgery, Shahid Modarres Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran
3Assistant Professor of anesthesiology, Shahid Modarres Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Corresponding author: Gholizadeh B, Assistant Professor of General Surgery, Shahid Modarres Medical Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran, E-mail:barmak.gholizadeh@gmail.com


Received Date: September 30, 2017  Accepted Date: October 25, 2017  Published Date: October 27, 2017

Abstract
The Appendiceal mucocele (AM) was considered as a rare dilation of the appendiceal lumen. Four different types of AMs are defined according to the cause of obstruction, for both benign and malignant, including retention cysts, epithelial hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. Mucinous adenocarcinomas of the appendix, which responsible for approximately 40% of appendiceal adenocarcinomas, are proportionality uncommon and rarely associated with the development of pseudomyxoma peritonei (PMP) syndrome so, patients with appendiceal adenoma or cystadenoma should be assessed for the presence of lesions elsewhere in the colon as a result of the strong association with synchronous or metachronous colorectal adenoma or carcinoma.

Keywords: Appendiceal Neoplasms; Mucinous cyst adenocarcinoma; Pseudomyxoma Peritonei; Retroperitoneal; Pelvic Bones

Method: Here presented the case of an incidentally discovered mucinous cystadenocarcinoma in an 85-year-old man, whom subsequently underwent laparotomy and omentectomy in follow.

Result: A mass was in RLQ that extend to the right iliac wing. A computerized tomography (CT) scans of the abdomen and pelvic showed a clear mass with no liver metastases. Laparotomy revealed gelatinous mucus sticking both to the omentum and to the mass of RLQ with no evidence of liver metastases. Although, the mass was adhesive to the retroperitoneal and iliac wings, it was taken en bloc with appropriate all free margins.

Discussion: The presented case reinforces the potential for metastatic spread of mucinous cyst adenocarcinoma to retroperitoneal and pelvic bone, which is not reported before, and solved questions in today's inconsistent practice for treating AMs.

Introduction
The Appendiceal mucocele (AM) was first described in 1842 by Rokitansky [1]. This disease considered as a rare dilation of the appendiceal lumen, which is found in 0.2 to 0.3% of the appendectomies [2], secondary to the accumulation of mucinous secretions [3]. Four different types of AMs are defined according to the cause of obstruction, for both benign and malignant, including retention cysts, epithelial hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. The latter two demonstrate neoplastic processes, with cystadenomas typically referred to as low-grade appendiceal mucinous neoplasms (LAMNs) [4]. Mucinous adenocarcinomas of the appendix which are responsible for approximately 40% of appendiceal adenocarcinomas are proportionality uncommon and rarely associated with the development of pseudomyxoma peritonei (PMP) syndrome [5]. The both terms of adenoma and cystadenoma, which are interchangeable, rigorously used to refer to a neoplastic process, which is benign and does not recur. These neoplasms account for the most majority of cases of pseudomyxoma peritonei (PMP). The mucinous feature in FMP is mucinous ascites without intracellular mucin accumulation in tumor cells, and described as neoplastic lesions analogous to adenomas occurring elsewhere in the gastrointestinal tract but possessing a different appearance, because
of inimitable growth restriction in the appendix [5]. Mucin secreting epithelium has a tendency to accumulate in the regions of the right subdiaphragm and subhepatic space, the left abdominal gutter, greater omentum, and pelvis. In most samples, although the mucin is microscopic, it may occasionally form a localized collection in the right lower quadrant (RLQ). Also, perforation is usually associated with mucin dissection and localized mucus collections attached to the serosa or lying free within the peritoneal cavity. In the neoplasm of low malignant potential, neoplastic cells penetrate the appendiceal wall and spread beyond the appendix in the form of peritoneal implants, which occur on the spleen, liver and ovarian involvement. Spread to the uterus and fallopian tube with partial replacement of the tubal mucosa may also occur. In this case, patients with appendice adenoma or cyst adenoma should be assessed for the presence of lesions elsewhere in the colon as a result of the strong association with synchronous or metachronous colorectal adenoma or carcinoma [6,7].

The primary tumor is predominantly a minimally invasive appendiceal mucinous epithelial neoplasm with a high propensity for spread to peritoneal surfaces, but almost no lymphatic or hematogenous metastases [8,9]. The initial cancer dissemination is through the wall of the appendix into the peritoneal space. Tumor cells from the ruptured appendiceal neoplasm are spread throughout the peritoneal cavity by the intraperitoneal fluid current and gravity. This passive movement might explain through the absence of adhesive characteristics on the cell surface. The older literature states that mucinous adenomas of the appendix are cured by appendectomy, provided the resection margin at the base of the appendix is free of involvement [10]. Also, the complete surgical margin of the appendix should be evaluated for the presence of neoplastic epithelium in associated with prognostic implications of the margin status of appendectomy specimens. In addition, if a tumor has features of a low-grade mucinous neoplasm, the presence of invasion of neoplastic epithelium or carcinoma-like areas influences the diagnosis and has remarkable prognostic ramification. For this reason, recent studies have advocated a more assertive approach to the management of these unforeseeable lesions with right hemicolecctomy to obtain clear margins and thorough surgical debunking with omentectomy. Furthermore, complete removal of all mucinous material and implants would be used, even if it requires a second procedure [8,11,12]. Another surgical treatment of mucinous cystadenocarcinoma that has been revealed a reasonable alternative by clinical studies is Laparoscopic appendectomy [13].

Here, we present the case of an incidentally discovered mucinous cystadenocarcinoma in an 85-year-old man, whom subsequently underwent laparotomy and omentectomy in follow. The patient's diagnosis and course of treatment emphasize a tendency for Mucinous adenocarcinomas to accumulate in new regions which are not reported before.

Case report

85-year-old Iranian men presented to a hospital with an abdominal pain for a month. During the examination, a mass was touched in RLQ that extend to the right iliac wing. The mass was firm consistency and not mobile. Although, the conjunctiva of the eyes appeared pale, no other points were seen in the examination. He did not have any previous history of surgery or malignancy and significant medical comorbidities. On the Laboratory evaluation, the hemoglobin was 9 grams per deciliter. A computerized tomography (CT) scan of the abdomen and pelvis was performed, which showed a clear mass with no liver metastases (Figure 1).

Figure 1: A computerized tomography (CT) scan of the abdomen and pelvis showing a mass in the right lower quadrant extending to pelvic bone.

At this point, the patient was referred to our institution for surgical intervention. Laparotomy revealed gelatinous mucus sticking both to the omentum and to the mass of RLQ (Figure 2) with no evidence of liver metastases.
Subsequently, an omentectomy was performed to remove the mass. Although, the mass was adhesive to the retroperitoneal and iliac wings (figure 2), it was taken en bloc with appropriate all free margins (figure 3). The patient's postoperative course was uneventful, and he was subsequently discharged.

On pathology report the resected mass consisted of skin with subcutaneous tissue and partial fragment of pelvic bone totally (11 × 8 × 4 cm) attached right hemicolectomy (11 cm in length and 3 cm in diameter). After cut section of mass one cystic-mucoidal mass was seen in appendical wall with dilation located in external surface of first part of right colon with extension to attached skin and subcutaneous tissue (extension to muscle layer, 10 × 5 × 3 cm). The nearest distance of mass from radial margin was 0.1 cm. Also, one lymph node (0.5 × 0.3 × 0.4 cm) was seen.

**Discussion**

Pseudomyxoma peritonei (PMP) is a rare clinical condition that has been characterized by extensive accumulation of thick, gelatinous mucus within the abdominal and/or pelvic peritoneal cavity, and is associated with biologically heterogeneous behavior. Tumoral peritoneal spread occurs in most patients throughout the peritoneal cavity often with ovarian involvement, but distant metastases are infrequent [14,15]. To the extent of our knowledge, this is the first work ever reported and shows case of mucinous adenocarcinoma of the appendix with retroperitoneal and pelvic bone extension. The term PMP is used to describe the peritoneal dissemination of mucus-producing adenocarcinomas of the appendix, large and small bowel, and other sites [9,16,17].
The prognosis of mucinous adenocarcinoma depends on the grade of malignancy of the mucinous tumor and success of surgery to remove all the tumors that have metastasized into the abdomen [18]. It is well known that disseminated mucin-producing adenocarcinomas of the appendix cause an aggressive subtype of peritoneal mucinous tumors [19]. Mucoceles may also be categorized by size, which could be rarely malignant as it has been reported <2 cm, be cystadenoma and cystadenocarcinoma while associated with sizes >6 cm, as well as a higher rate of perforation [20]. Both ruptured benign and malignant neoplasia can produce mucinous peritoneal spread leading to diagnosis of PMP [21]. Contrast-enhanced CT imaging is most commonly used modal quality for preoperative diagnosis. CT findings suggestive of a mucocele include an appendical lumen >1.3 cm, with cystic dilatation, and wall calcification [22,23]. Basically, mucinous cystadenomas may have associated with cystic masses, low contrast attenuation, irregular wall thickening, and absence of inflammation [24]. The appropriate treatment of these tumor types is right hemicolecctiony, especially if the disease appears to be confined to the appendix, because complete excision with removal of potentially involved lymph nodes improves the potential for cure [11]. In addition, laparoscopic technique has been recommended with citing advantageous such as reduced risk of seeding, magnification of the surgical field, and a speedier recovery, emphasizing the facility of conversion to open surgery if necessary [25]. Most studies that deal with appendiceal neoplasia focus on tumors confined to the appendix or those that have spread widely throughout the abdomen [4,26]. González Moreno et al. [8] used a laparoscopic surgery resect an appendiceal mucocele caused by a non-perforated mucinous adenocarcinoma. In which, implants of mucinous tumor were found widely disseminated on peritoneal surfaces at laparotomy 9 months later. They showed that all appendiceal tumors, including the most benign-appearing adenomas, could result in diffuse peritoneal implantation [13]. In an attempt, Higa et al. [10] explored twelve patients to obtain a better understanding of the nature, morphology, and behavior of appendiceal “mucocele,” The present clinicopathologic study was carried out because of symptoms known as the right lower quadrant, usually pain or discomfort, and 6 cases had a palpated mass in this area. Also, they had performed a roent genographic studies which revealed the lesion in a single instance as a soft tissue mass at the ileocecal area having a partially calcified wall [10]. In another study of appendiceal tumors done by Carr and Sobin [27] reported that 1 of 5 (20%) appendiceal mucinous neoplasms with cellular mucin was associated with the development of diffuse pseudomyxoma peritonei to the right lower quadrant. Światkowska-Freund et al. [28] presented a case of a woman with diagnosis of adnexal tumor of the right ovary. Laparotomy was performed and not only tumor of appendix was found, also a small tumor in small bowel and some increased lymphatic nodules were recognized. While, they confessed that it is difficult to diagnose pelvic masses when they did not originate from the adnexa [28]. Our patient with pseudomyxoma peritonei arising from a mucinous cyst adenocarcinoma was very interesting because shows a case of mucinous adenocarcinoma of the appendix with retroperitoneal and pelvic bone extension. In this study, CT scan was performed to find the localization and anatomical definition of a mucinous cyst. Laparotomy revealed gelatinous mucus sticking both to the omentum and to the mass of RLQ.

Pelvic mass was resected unblock with skin with subcutaneous tissue and partial fragment of pelvic bone attached right hemicolecotomy were resected. The obtained data revealed that malignant neoplasm was compatible with mucinous cyst adenocarcinoma. The tumor size was 10 × 5 × 3 cm and it located in appendix with extension to pelvic soft tissue. Non-tumoral areas of pelvic soft tissue were in favor for featuring several mixed inflammatory and abscess formations. Proximal and distal margins of the mass were free. The nearest distance of mass from radial margin was 0.1 cm. In addition, the attachment bony surface tissue near the pelvic soft tissue was not involved, and surgical bone margin was free. The inner surface of colonic mucosa with no pathologic change was seen. It was notable that one lymph node was free. About the retroperitoneal mass, pseudo myxoma peritoneum was compatible for mucinous cyst adenocarcinoma.

In conclusion, the presented case reinforces the potential for metastatic spread of mucinous cyst adenocarcinoma, which is not reported before, and solved questions in today’s inconsistent practice for treating.

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