



Mucinous Tumor of Appendix: An Uncommon Disease with Rare Presentations



Vijay Sharma^{1*}, Ankur Agarwal² and Richa Sharma³

¹Department of Gastroenterology, Manipal Hospital, India

²Department of Pathology, Manipal Hospital, India

³Department of Public Health, Regional Institute of Health Medicine Research, India

Submission: April 22, 2017; **Published:** April 28, 2017

***Corresponding author:** Vijay Sharma, Department of Gastroenterology, Manipal Hospital, Jaipur, pin code 302022, India, Tel: +919928015254; Email: vijaysadashiv@gmail.com

Abbreviations: EPIC: Early Postoperative Intraperitoneal Chemotherapy; WHO: World Health Organization; PMP: Pseudomyxomateritonei; DPAM: Disseminated Peritoneal Adenomucinosis; PMCA: Peritoneal Mucinous Carcinomatosis; HIPEC: Hyperthermic Intraperitoneal Chemotherapy

Mini Review

Recently in our hospital 54-year-old Indian female patient presented with pain right hypochondrium, underwent cholecystectomy 3 years back for the same pain, now presented in emergency with severe persistent pain right side of abdomen and fullness abdomen for 2 days. On examination there was tenderness and guarding of the right lumbar, right hypochondrium region, post surgery scar was normal, no hepatosplenomegaly. A laparotomy was performed, during which a large amount of mucinous material seen in subhepatic region along with bulky gelatinous appendiceal retrocecal tumor not infiltrating the omentum were found. All mucinous substance aspirated and appendicectomy done. Colon and cecum was inspected and palpated with no recognition of infiltration and separate appendix. Pathologist confirmed mucinous tumor of appendix progressing to pseudomyxomateritonei, opinion of oncologist is being taken while writing the short review

Background

Mucocele of the appendix was first described by Rokitsky [1], It is a morphologic entity explained as dilatation of the appendiceal lumen by abnormal excessive accumulation of mucus. The disease has been reported in 0.3-0.7% of appendiceal specimen and 8% of all appendiceal tumors [2] and it is more frequent in middle aged females [3]. Diagnosis is often mostly difficult preoperatively despite extensive work up and most patients remain undiagnosed until the lesions are discovered incidentally intraoperatively or postoperatively for appendicitis [3].

Etiological factors are mostly inflammatory and also neoplastic. Treatment is directed as per the underlying pathology. Proper management of neoplastic mucocoeles is essential to prevent rupture of the mucocoele and progression to mucinous intraperitoneal dissemination, resulting in a syndrome called pseudomyxomateritonei which is called “jelly belly” [4] and constitutes around 1% of all colorectal cancers in the Western countries [5].

Discussion

There are different reports of patients with mucinous neoplasms of the appendix presenting with clinically varied spectrum of pathologic processes and presentations. Mostly incidental diagnosis as described in our experience [6]. While, pseudomyxomateritonei, though clinically more severe, commonly has a mild course with non-specific abdominal symptoms; therefore, a high level of suspicion is essential, especially if the medical history is suggestive of Appendiceal pathology [7]. The mucinous neoplasms of the appendix are classified into 4 pathological entities as per the characteristics of the epithelium [2,3,6]:

- I. Simple or retention mucocoeles result from non-tumoral obstruction of the appendiceal outflow, usually by a fecalith or inflammatory stricture, and they are characterized by normal epithelium and mild luminal dilatation up to 1 cm.
- II. Mucocoeles with local or diffuse villous hyperplastic epithelium. The luminal dilatation is also mild and they constitute 5-25% of mucocoeles.

III. Mucinous adenoma/cystadenoma are most common type, approx. 63-84% of cases. mostly epithelial villous adenomatous changes with some epithelial atypia, usually with marked (up to 6 cm) distention of the lumen.

IV. Malignant mucinous cystadenocarcinomas represent 11-20% of cases. ON histopathology it presents with glandular stromal invasion, desmoplastic reaction, and/or presence of epithelial cells in the peritoneal implants. The luminal distention is usually severe.

Mucinous cystadenoma is benign end of the disease, with no risk of recurrence, while Mucinous adenocarcinoma is associated with poor survival and high rate of metastases to lymph nodes and liver. Middle in the spectrum lies mucinous neoplasms, which are occasionally are associated with the development of pseudomyxomaperitonei. World Health Organization (WHO) classifies both as "low-grade mucinous neoplasm. Intermediate grade tumors have mucin dissecting into or through the wall of the appendix with or without epithelial cells. Extra appendiceal mucin with epithelial cells characterizes the mucinous neoplasm with a high rate of recurrence, which is most commonly associated with the development of pseudomyxomaperitonei.

Pseudomyxomaperitonei (PMP) is a broad term including a wide spectrum of tumors, ranging from benign to the frankly malignant lesion. Ovary was considered as the commonest primary site in the past, now with immunohistochemical analysis and molecular biology, it is confirmed that ovary is a rare source of pseudomyxoma, and most of the lesions diagnosed as "borderline mucinous tumors of the ovary" are typically metastatic from the appendix [7]. Ronnett classification explained "disseminated peritoneal adenomucinosis" (DPAM) from "peritoneal mucinous carcinomatosis" (PMCA) [8]. DPAM represents the classic PMP with paucicellular mucinous ascites with mild clinical course, while MCA has a higher percentage of overtly malignant cells and a poor prognosis [9].

The clinical symptoms are usually absent or non-specific in most cases of appendiceal mucocoeles or even in the presence of pseudomyxomaperitonei. The diagnosis is established mostly by abdominal CT scan now a day. In the presence of an appendiceal mucocoele, CT scan shows a well-encapsulated, round, thin-walled cystic mass. Calcification is seen in 50% of cases [10], while enhancing nodules in the mucocoele wall are suggestive of cystadenocarcinoma [11]. Mucocoeles less than 2 cm are rarely malignant but larger mucocoeles (6 cm or more) are usually associated with cystadenoma or cyst-adenocarcinoma and a higher perforation rate (20%) [12,13]. Visceral scalloping is a diagnostic sign on CT. As the mucin producing cells in pseudomyxomaperitonei are poorly adhesive, they are easily dislodged by peristaltic movement and adhere at sites of relative stasis. The pouch of Douglas/rectovesical pouch, right and left subphrenic spaces, and surface of the liver and spleen are the commonest sites involved [14,15].

Although right hemicolectomy has been traditionally the standard treatment for mucinous appendiceal malignancies, recent papers shows that an intact mucocoele represents benign process and does not progression. [16]. Gonzalez-Moreno et al. [17] reported no survival advantage with right hemicolectomy versus appendectomy after reviewing 501 patients diagnosed with appendiceal epithelial neoplasms. They suggested need of right hemicolectomy:

- total removal of the primary tumor or complete cytoreduction,
- lymph node involvement and
- A non-mucinous neoplasm identified by histopathology. Few authors suggest that laparoscopic appendectomy is a safe option formucocoele of the appendix [18].

Possible rupture and dispersion of mucus or epithelial cells into the peritoneal cavity is associated with a poorer prognosis. Grasping of the appendix specimen should be minimal, pneumoperitoneum levels should be low, and a retrievable bag should always be used when the laparoscopic approach is favored [18]. Any mucinous fluid within the abdomen should be carefully examined and if epithelial cells are identified, a diagnosis of pseudomyxomaperitonei syndrome or mucinous peritoneal carcinomatosis of appendiceal origin should be established. It is probably safer to convert to an open procedure if such mucocoeles are visualized during a laparoscopic approach [19].

Treatment of pseudomyxomaperitonei is variable, both due to the rarity of the disease and to its frequently slow-growing nature [20]. Current treatment strategies range from watchful waiting to extensive cytoreductive surgery alone or with hyperthermic intraoperative peritoneal chemotherapy (HIPEC) or early postoperative intraperitoneal chemotherapy (EPIC) [21]. Recent study showed that cytoreductive surgery with intraperitoneal hyperthermic perfusion permitted complete tumor removal, confirmed efficacy of combined treatment in terms of improved long-term survival and better regional control of the disease [22]. Other studies reported fluorouracil-based adjuvant systemic chemotherapy better for patients with PMP of appendiceal origin [23]. In situations where surgery is not immediately required, patients can be monitored via CT scans, tumor marker laboratory tests, and physical symptoms, to determine when and why surgery is warranted. Risk of adenocarcinoma of the colon is 6 times greater in patients with a mucocoele than in the general population, colonic surveillance is warranted in these cases [24].

Conclusion

Patients with appendiceal mucocoeles can present with varied sign and symptoms. Preoperative diagnosis, although challenging, helps in planning right management and avoiding both intra-operative and post-operative complications.

Pseudomyxomaperitonei is a complex medical situation that is best dealt with in a specialized center, where doctors should focus on the prevention of loco-regional recurrence.

References

- Rokitansky CF (2011) A manual of pathological anatomy. Blanchard & Lea, Philadelphia (PA), USA, 1855: 89.
- Ruiz-Tovar J, Teruel DG, Castiñeiras VM (2007) Mucocele of the appendix. *World J Surg* 31(3): 542-548.
- Aho AJ, Heinonen R, Lauren P (1973) Benign and malignant mucocele of the appendix. *Acta Chir Scand* 139(4): 392-400.
- Landen S, Bertrand C, Maddern GJ (1992) Appendiceal mucoceles and pseudomyxomaperitonei. *Surg Gynecol Obstet* 175: 401-404.
- Fann JL, Vierra M, Fisher D (1993) Pseudomyxomaperitonei. *Surg Gynecol Obstet* 177: 441-447.
- Pai RK, Longacre TA (2005) Appendiceal mucinous tumors and pseudomyxomaperitonei: histologic features, diagnostic problems, and proposed classification. *Adv Anat Pathol* 12(6): 291-311.
- Carr NJ (2014) Current concepts in pseudomyxomaperitonei. *Ann Pathol* 34: 9-13.
- Ronnett BM, Zahn CM, Kurman RJ (1995) Disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis. A clinicopathologic analysis of 109 cases with emphasis on distinguishing pathologic features, site of origin, prognosis, and relationship to "pseudomyxomaperitonei". *Am J SurgPathol* 19: 1390-1408.
- Ronnett BM, Yan H, Kurman RJ (2001) Patients with pseudomyxomaperitonei associated with disseminated peritoneal adenomucinosis have a significantly more favorable prognosis than patients with peritoneal mucinous carcinomatosis. *Cancer* 92: 85-91.
- Pickhardt PJ, Levy AD, Rohrmann CA, Jr, Kende AI (2003) Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation. *Radiographics* 23: 645-662.
- Chiou YY, Pitman MB, Hahn PF (2003) Rare benign and malignant appendiceal lesions: spectrum of computed tomography findings with pathologic correlation. *J Comput Assist Tomogr* 27: 297-306.
- Deans GT, Spence RA (1995) Neoplastic lesions of the appendix. *Br J Surg* 82: 299-306.
- Lau H, Yuen WK, Loong F, Lee F (2002) Laparoscopic resection of an appendiceal mucocele. *Surg Laparosc Endosc Percutan Tech* 12: 367-370.
- Fairise A, Barbary C, Derelle AL (2008) Mucocele appendiculaire et pseudomyxome peritoneal. *J Radiol* 89: 751-762.
- Diop AD, Fontarensky M, Montoriol PF, Da Ines D (2014) CT imaging of peritoneal carcinomatosis and its mimics. *Diagn Interv Imaging* S2211-5684(14)00037-0.
- Misdraji J, Yantiss RK, Graeme-Cook FM (2003) Appendiceal mucinous neoplasms: a clinicopathologic analysis of 107 cases. *Am J SurgPathol* 27: 1089-1103.
- Gonzalez-Moreno S, Sugarbaker PH (2004) Right hemicolectomy does not confer a survival advantage in patients with mucinous carcinoma of the appendix and peritoneal seeding. *Br J Surg* 91: 304-311.
- Singh MK, Kumar MK, Singh R (2014) Laparoscopic appendectomy for mucocele of the appendix. *J Nat Sci Biol Med* 5: 204-206.
- Gonzalez Moreno S, Shmookler BM, Sugarbaker PH (1998) Appendiceal mucocele. Contraindication to laparoscopic appendectomy. *SurgEndosc* 12: 1177-1179.
- Buell-Gutbrod R, Gwin K (2013) Pathologic diagnosis, origin, and natural history of pseudomyxomaperitonei. *Am Soc Clin Oncol Educ Book* 221-225.
- Dhage-Ivatury S, Sugarbaker PH (2006) Update on the surgical approach to mucocele of the appendix. *J Am Coll Surg* 202: 680-684.
- Deraco M, Baratti D, Inglese MG (2004) Peritonectomy and intraperitoneal hyperthermic perfusion (IPHP): a strategy that has confirmed its efficacy in patients with pseudomyxomaperitonei. *Ann Surg Oncol* 11: 393-398.
- Chen CF, Huang CJ, Kang WY, Hsieh JS (2008) Experience with adjuvant chemotherapy for pseudomyxomaperitonei secondary to mucinous adenocarcinoma of the appendix with oxaliplatin/ fluorouracil/ leucovorin (FOLFOX4). *World J Surg Oncol* 6: 118.
- Federle MP, Anne VS (2004) Mucocele of the appendix. In: Federle MP, editor. *Diagnostic Imaging. Abdomen* Salt Lake City, UT: Amirsys, pp. 26-27.



This work is licensed under Creative Commons Attribution 4.0 License
DOI: [10.19080/OAJS.2017.03.555623](https://doi.org/10.19080/OAJS.2017.03.555623)

Your next submission with Juniper Publishers will reach you the below assets

- Quality Editorial service
- Swift Peer Review
- Reprints availability
- E-prints Service
- Manuscript Podcast for convenient understanding
- Global attainment for your research
- Manuscript accessibility in different formats (Pdf, E-pub, Full Text, Audio)
- Unceasing customer service

Track the below URL for one-step submission
<https://juniperpublishers.com/online-submission.php>