ISSN 2689-8268 | Volume 3

American Journal of Surgery and Clinical Case Reports

Case Report Open Access

Incidental Diagnosis of a High Grade Mucinous Neoplasm of the Appendix: A Case Report

Dannaoui K[1*], Hernandez M[4], Young A[4], Clouse K[4], Gomez J[2], Matlin C[3] and Arias A[2]

[1]Abrazo Central Medical Center, Phoenix, Arizona

[2]Medical Student, Universidad Autonoma De Guadalajara, Jalisco, Mexico

[3]Medical Student, A.T. Still University School of Osteopathic Medicine, Mesa, Arizona

[4]Attending, Abrazo Central Medical Center, Phoenix, Arizona

[*]Corresponding author:

Kareem Dannaoui,
Abrazo Central Medical Center, Phoenix, Arizona,
Tel: +1 (602) 781-3973;
E-mail: kdanna123@gmail.com

Received: 14 Sep 2021
Accepted: 22 Sep 2021
Published: 27 Sep 2021

Citation:

Dannaoui K. Incidental Diagnosis of a High Grade Mucinous Neoplasm of the Appendix: A Case Report. Ame J Surg Clin Case Rep. 2021; 3(13): 1-3

1. Abstract

1.1. Presentation of the Case

A 74-year- old female presented to the emergency department for altered mental status secondary to severe microcytic anemia. A colonoscopy found a large ulcerated, polypoid mass nearly 40mm in size that was nearly obstructing the ascending colon. Histopathologic examination of a biopsy taken from the mass proved to be moderately differentiated adenocarcinoma of the ascending colon. The patient subsequently underwent an extended right-hemicolectomy with ileocolic anastomosis. Postoperative histopathologic examination of the surgical specimen was positive for invasive adenocarcinoma with moderate differentiation. In addition, a high-grade appendiceal mucinous neoplasm was located at the distal half and tip of the appendix.

2. Introduction

Appendiceal neoplasms are rare tumors of the gastrointestinal tract that comprise approximately 1% of all cancers, however their incidence has been rising in recent years [1-3]. The most commonly reported appendiceal cancers are neuroendocrine tumors (NET) and epithelial neoplasms. Benign tumors may be asymptomatic, or they may present with a clinical picture similar to acute appendicitis. Diagnosis of appendiceal neoplasm is often made upon histologic

examination of appendectomy specimens. Malignant neoplasms and advanced stage disease may cause symptoms associated with peritoneal spread such as increasing abdominal girth, chronic abdominal pain, and weight loss. The classification system of appendiceal carcinomas has been the topic of much debate with various organizations using different diagnostic terminology.

Recently, a generalized consensus was reached regarding the diagnostic terminology for epithelial appendiceal neoplasms. This new nomenclature is based primarily on histologic characteristics and degree of infiltration. Histologic appearance of appendiceal neoplasms is now classified as mucinous, non-mucinous, ex-goblet cell, or signet ring. Mucinous adenocarcinoma of the appendix can be further subdivided based on grade and differentiation. Low grade neoplasms are well differentiated, whole high-grade neoplasms are poorly differentiated and frequently present with peritoneal involvement [3-5]. Evidence-based guidelines for the management of appendiceal neoplasms are lacking and current treatment strategies mirror those for colorectal carcinoma (CRC). However, recent molecular profiling analyses have demonstrated that appendiceal neoplasms and CRC are distinct entities with separate driving genomic mutations [6,7]. Therefore, treatment strategies for appendiceal neoplasms should be targeted separately from CRC, even when these two disease processes are found concurrently.

Here we present and analyze the case of a 74-year-old female who was found to have a high grade appendiceal mucinous neoplasm after an extended right hemicolectomy for adenocarcinoma of the ascending colon.

3. Description of Case

A 74-year-old female with a significant past medical history of diabetes, hypertension, lymphoma, breast cancer, and CVA, presented to a local medical center's emergency department via EMS for altered mental status. The patient was found by her family who

reported a decreased level of her responsiveness & proceeded to call EMS. Upon EMS arrival, the patient was unresponsive to sternal rub but slowly regained consciousness en route to the ED.

On admission the patient's vitals are as noted: heart rate of 90 beats per minute, blood pressure of 140/80 mmHg, SpO2/pulse oximetry of 100% on, respiratory rate of 10 & temperature of 97.9F. On physical examination the patient was noted to have right-sided hemiparesis and dysarthria which, per family, was her baseline due to previous CVA. Labs in the ED showed a RBC of 2.65 (10^3/mcL), Hgb of 4.8 g/dL, Hct 16.9%. Computerized tomography of head without contrast and Computed tomography angiography head/neck were negative for any acute intracranial abnormalities. The patient was transferred to ICU with consultation for hematology/oncology & gastroenterology in regard to the patient's severe anemia.

Computerized tomography of abdomen & pelvis without/with contrast as well as EGD with biopsies were unrevealing for any pathologies. On colonoscopy, a large, ulcerated polypoid mass was nearly obstructing the ascending colon leading to non-visualization of the cecum.

Tattoo with India ink was applied 3cm distal to the obstructing mass. Biopsy of the ascending colon proved to be moderately differentiated adenocarcinoma by histopathology. The patient underwent an extended right hemicolectomy from the terminal ileum to the distal transverse colon with ileocolic anastomosis. Findings during the procedure included a large mass in the mid-ascending colon without any obvious metastatic disease. Postoperative Surgical histopathology showed a right colon mass with invasive adenocarcinoma that was moderately differentiated. Distance of the invasive tumor to the closest margin was 2.5cm from the nearest retroperitoneal margin/surface. Pathologic stage classification (pTNM, AJCC 8th edition) of the colonic adenocarcinoma was found to be pT4a, pN0, pM(N/A) as well as an appendix with high grade appendiceal mucinous neoplasm. The appendiceal mass measured 2.0cm in size, located at the distal half & tip of the appendix. The proximal and distal margins of the specimen were uninvolved by high grade appendiceal mucinous neoplasm or acellular mucin. Pathologic stage classification (pTNM, AJCC 8th edition) of the appendiceal mucinous neoplasm was found to be pT4a, pN0, pM(N/A). Zero of the nineteen regional lymph nodes were positive for metastatic carcinoma.

4. Discussion

Mucinous neoplasms of the appendix are extremely rare histopathological entities that are often discovered incidentally upon resection of the appendix for other conditions. Here, we report a case of a 74-year-old female who presented to the hospital with altered mental status and acute blood loss anemia. Initial CT of the abdomen and pelvis was unremarkable except for trace fluid in the right paracolic gutter. A colonoscopy revealed a large, ulcerated, obstructing mass involving the proximal ascending colon and an extended right hemicolectomy was performed. Intraoperative findings included a large mass in the mid ascending colon with no obvious metastasis noted. There were no peritoneal masses or omental seeding. Upon visualization of the appendix, there appeared to be a lesion at the tip of the appendix with matting of nearby mesenteric lymph nodes. Pathological evaluation of the surgical specimen revealed moderately differentiated, invasive colonic adenocarcinoma along with a high-grade mucinous neoplasm (HAMN) of the appendix. HAMN is defined as a mucinous neoplasm with high grade cytological atypia [5,8]. There was also fibrous obliteration of the appendiceal tip and the presence of acellular mucin invading the visceral peritoneum. There were no affected margins or lymphatic invasion.

The incidence of a synchronous appendiceal neoplasm in patients with colorectal cancer is about 4% [9]. The clinical features of an appendiceal neoplasm are widely distinct from those of a colorectal neoplasm. The clinical characteristics of appendiceal cancer can vary from completely asymptomatic to acute appendicitis or signs of peritoneal invasion. Colorectal carcinomas usually present with signs of either obstruction or a lower gastrointestinal bleed.

Despite these unique clinical attributes, appendiceal neoplasms and colorectal carcinomas have been managed similarly due to their relative anatomic proximity and shared embryological origin. However, recent molecular profiling analyses using next-generation sequencing (NSG) have shown that appendiceal neoplasms have distinct molecular profiles and genetic mutations [6]. One study revealed high rates of KRAS and GNAS (28% vs 2%) mutations in appendiceal carcinoma compared to CRC [6,7]. Appendiceal carcinoma was also associated with fewer APC (9% vs 55%) and TP53 (27 vs 67%) mutations compared to CRC 6. It has been reported that the prevalence of microsatellite instability in appendiceal carcinoma is low compared to right sided CRC. Microsatellite instability testing of the right hemicolectomy specimen revealed no loss of nuclear staining for several DNA repair enzymes, including MLH-1, PMS-2, MSH-2, and MSH-6. This indicates that our patient's primary lesion is microsatellite stable. Optimal treatment strategies for the management of appendiceal carcinoma are lacking due to the elusive nature of the disease. Current recommendations for post-operative management of appendiceal mucinous neoplasms depend on the invasiveness of the disease [8].

Non-invasive low-grade tumors can be successfully treated with surgical resection alone and there is little risk of tumor recurrence. Low grade tumors with invasive features such as vascular or lymphatic involvement may require hyperthermic intraperitoneal chemotherapy (HIPEC) following primary resection [10]. The cytoreductive surgery (CRS)/HIPEC approach has gained desirability since it has been associated with long-term survival in patients with low grade vappendiceal neoplasms [10]. High grade appen-

diceal neoplasms (HAMNs) pose more of a challenge regarding management since literature on the natural history of the disease is limited. Treatment options for high grade neoplasms include surgical debulking, HIPEC, and possible preoperative systemic chemotherapy [11]. The risk of synchronous peritoneal metastases (PM) in appendiceal cancer is high, with a 5-year cumulative incidence of 9.0% 12. One population-based study found that appendiceal cancer, right sided CRC, advanced tumor and node stage, mucinous histopathology, and vascular invasion were all independent risk factors for PM [12]. In this same study, 23.5% of patients with appendiceal cancer were diagnosed with PM [12]. As stated above, our patient's appendiceal lesion was classified as pT4a, pN0, pM(N/A) according to the AJCC 8th edition criteria. This indicates that the tumor was invading the visceral peritoneum. Based on these high-risk features, the threshold for surveillance of PM in this patient is low. Current recommendations for surveillance include cross sectional imaging with contrast-enhanced CT or diagnostic laparoscopy if there is no detectable disease on imaging [10]. Early detection of PM is crucial since prompt recognition and initiation of HIPEC can improve patient outcomes.

The incidental finding of an appendiceal malignancy during resection for primary colonic masses has been previously documented. It is important to understand the origins of these cancers and treat them as separate entities. New insights into the genetic behavior of appendiceal neoplasms may aid in the development of targeted therapies, leading to more personalized treatment options and enhanced survival. This information can be used to further the development of management protocols for appendiceal neoplasms.

References

1. Lohsiriwat V, Vongjirad A, Lohsiriwat D. Incidence of synchronous appendiceal neoplasm in patients with colorectal cancer and its clinical significance. World J Surg Oncol. 2009; 7: 51.
2. Singh H, Koomson AS, Decker KM, Park J, Demers AA. Continued increasing incidence of malignant appendiceal tumors in Canada and the United States: A population-based study. Cancer. 2020; 126(10): 2206-2216.
3. Guzmán GA, Montealegre I, Obando AM. Incidental diagnosis of a low-grade mucinous appendicular neoplasm: A case report. Int J Surg Case Rep. 2021; 83: 105998.
4. Lu P, Fields AC, Meyerhardt JA, Davids JS, Shabat G, Bleday R, et al. Systemic Chemotherapy and Survival in Patients with Metastatic Low Grade Appendiceal Mucinous Adenocarcinoma. J Surg Oncol. 2019; 120(3): 446-451.
5. Carr NJ, Cecil TD, Mohamed F, Sobin LH, Sugarbaker PH, Gonzalez-Moreno S,et al. A Consensus for Classification and Pathologic Reporting of Pseudomyxoma Peritonei and Associated Appendiceal Neoplasia: The Results of the Peritoneal Surface Oncology Group International (PSOGI) Modified Delphi Process. J Surg Pathol. 2016; 40(1): 14-26.
6. Raghav K, Shen JP, Jácome AA, Guerra JL, Scally CP, Tagart MW, et al. Integrated clinico-molecular profiling of appendiceal adenocarcinoma reveals a unique grade-driven entity distinct from colorectal cancer. Br J Cancer. 2020; 123(8): 1262-1270.
7. Tokunaga R, Xiu J, Johnston C, Goldberg RM, Philip PA, Seeber A, et al. Molecular profiling of appendiceal adenocarcinoma and comparison with right-sided and left-sided colorectal cancer. Clin Cancer Res Off J Am Assoc Cancer Res. 2019; 25(10): 3096-3103.
8. Van de Moortele M, De Hertogh G, Sagaert X, Van Cutsem E. Appendiceal cancer : a review of the literature. Acta Gastro-Enterol Belg. 2020; 83(3): 441-448.
9. Khan MN, Moran BJ. Four percent of patients undergoing colorectal cancer surgery may have synchronous appendiceal neoplasia. Dis Colon Rectum. 2007; 50(11): 1856-1859.
10. Bartlett DJ, Thacker PG, Grotz TE, Graham RP, Fletcher JG, VanBuren WM, et al. Mucinous appendiceal neoplasms: classification, imaging, and HIPEC. Abdom Radiol. 2019; 44(5): 1686-1702.
11. Shaib WL, Assi R, Shamseddine A, Alese OB, Staley C, Memis B et al. Appendiceal Mucinous Neoplasms: Diagnosis and Management. Oncologist. 2017; 22(9): 1107-1116.
12. Enblad M, Graf W, Birgisson H. Risk factors for appendiceal and colorectal peritoneal metastases. Eur J Surg Oncol. 2018; 44(7): 997-1005.