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Clinicopathological analysis of primary epithelial appendiceal neoplasms

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Abstract

Appendiceal carcinomas are classified into three distinct histopathological disease entities: *disseminated peritoneal adenomucinosis* (DPAM), *peritoneal mucinous carcinomatosis* (PMCA), or *peritoneal mucinous carcinomatosis* with intermediate or discordant features (PMCA I/D). The treatment of appendiceal tumors should be based on accurate histopathological classification, per previously reported case series. The objective of this study was to evaluate the clinicopathologic presentation and outcome of patients with appendiceal tumors treated at our institution over a 15-year period. We identified patients with appendiceal tumors diagnosed or treated at our institution from January 1989 through May 2004. Chart review for age, gender, signs and symptoms at

diagnosis, and treatment was performed. Review of the pathologic specimens was performed and tumors were classified as DPAM, PMCA I/D, or PMCA. Forty patients were identified (median age 52.5 years; males 38%). The number of patients with DPAM, PMCA I/D, and PMCA was 15 (38%), 6 (15%), and 18 (46%), respectively. Peritoneal involvement was seen in 11 (73%) of patients with DPAM, 5 (83%) of PMCA I/D, and 11 (61%) of PMCA. The median survival for patients with DPAM, PMCA I/D, and PMCA was 7.7 years (90% CI: 2.9—upper limit not estimable), 1.2 years (90% CI: 0.9–1.6), and 0.7 years (90% CI: 0.4–1.5), respectively. The difference in survival across the three groups was statistically significant. Three distinct histopathological disease entities exist in appendiceal tumors. The prognosis and management of these tumors should be based on the extent of disease and pathologic diagnosis.

Keywords

Appendiceal carcinoma; Pseudomyxoma peritonei; Disseminated peritoneal adenomucinosis (DPAM); Peritoneal mucinous carcinomatosis (PMCA)

Introduction

Appendiceal carcinomas are rare neoplasms accounting for 0.5–1% of all gastrointestinal malignancies [1, 2] or ~1,500 new cases per year. Incidental primary appendiceal carcinoma is detected in 1% of all appendectomy specimens [3, 4]. Most primary appendiceal adenocarcinomas are of the mucinous type, arise from an adenomatous polyp or serrated adenoma and frequently present with the pseudomyxoma peritonei syndrome [5–7].

Appendiceal neoplasms are distinct from colorectal carcinoma and [8, 9] tend to have a less aggressive natural history. Although lymphatic or hematogenous metastases are only seen in 2% of appendiceal tumors, local spread to the peritoneum is commonly seen in the majority of patients [10, 11].

Tumor histology in appendiceal cancer is prognostic and impacts treatment. Patients with well-differentiated appendiceal tumors have significantly improved survival as was demonstrated by three case series [12–15]. In the case series by Ronnett et al. [14], 109 patients with pseudomyxoma peritonei were classified into two distinct histological categories: *disseminated peritoneal adenomucinosis* (DPAM) or *peritoneal mucinous carcinomatosis* (PMCA). DPAM was characterized by peritoneal lesions composed of abundant extracellular mucin containing scant simple to focally proliferative mucinous epithelium with little cytological atypia or mitotic activity. Characteristic of appendiceal neoplasms classified as DPAM was the rare demonstration of hematogenous or lymphatic involvement. These tumors tended to have a prolonged benign course with peritoneal recurrences over many years. Much of the morbidity and mortality in such cases was because of intra-abdominal mucus accumulation leading to fibrosis and bowel obstruction. DPAM was typically slow growing and the morphology of the peritoneal lesions did not change over time.

In contrast, PMCA was characterized by peritoneal lesions composed of more abundant proliferative epithelium, glands, nests, or individual cells including signet cells, with the cytological and architectural features of carcinoma. Yan et al. [16] in a later report subclassified the PMCA group into well differentiated, moderately differentiated, and poorly differentiated cancer. Tumors classified as PMCA, on the other hand, were invariably more aggressive with frequent parenchymal organ and lymph node involvement and significant morbidity and mortality from intra-abdominal carcinomatosis. A significant difference in survival between cases classified as DPAM compared to those with PMCA was observed. A

third uncommon histological category with features either intermediate or discordant between DPAM and PMCA (PMCA I/D) was also described. These tumors were derived from well-differentiated appendiceal or intestinal mucinous adenocarcinomas and had peritoneal lesions displaying features of DPAM with focal areas of mucinous carcinoma. Tumors with discordant features were derived from atypical appendiceal adenoma with no histological evidence of transformation to carcinoma and had peritoneal lesions composed of mucinous carcinoma. Intermediate survival was observed in patients with PMCA I/D.

Due to their rare occurrence and the heterogeneous histological and biological characteristics, the management of appendiceal carcinomas is based on relatively small case series rather than randomized prospective trials. The objective of this study was to evaluate the clinicopathologic presentation and outcome of patients with appendiceal tumors treated at our institution over a 15-year period.

Patients and methods

Patient characteristics

The study was approved by the institutional review board of Wayne State University. The medical records of all patients diagnosed with appendiceal cancer seen at the Karmanos Cancer Center from the period of January 1989 through May 2004 were analyzed. A total of 40 patients were identified from surgical pathology records containing a diagnosis of pseudomyxoma peritonei of intestinal origin or primary appendiceal carcinoma. Patients with neuroendocrine tumors of the appendix, i.e., carcinoids or goblet cell carcinoid and disseminated ovarian adenocarcinomas were carefully excluded.

Patient demographics including age, gender, signs, and symptoms at diagnosis and treatment received were obtained from the medical records. Pathology parameters analyzed were histological grade (well, moderately, or poorly differentiated), T and N stage, and presence of pseudomyxoma peritonei. Information about histological classification of tumors as defined by Ronnet et al. [17] into the three groups of: (1) DPAM, (2) PMCA, and (3) PMCA I/D were also collected. All patients included in this study had their pathology reviewed for this analysis.

Statistical methods

Descriptive statistics were used to summarize the patient characteristics. Patient survival data were obtained from either the patient's medical records, direct communication, or the United States Social Security death index. Overall survival (OS) was calculated from the date of first visit to the Karmanos Cancer Center to the date of death from any cause. Surviving patients were censored as of the last date on which they were known to be still alive. Patients censored for OS with censoring dates >6 months prior to the data collection were recorded as lost to follow-up (LFU), but still treated as censored patients in the statistical analyses.

Median follow-up time for OS was computed using only the censored patients, to more accurately assess data maturity. The Kaplan–Meier (K–M) estimate of the censored OS distribution was computed. From the K–M analysis, the median OS and its confidence interval (CI) were derived. Due to the small sample sizes, survival statistics (e.g., median, 2-year rate) were estimated more conservatively using linear interpolation among successive event times on the K–M curves [18].

OS was compared by histopathological classification (three groups) via the log-rank test. To adjust for multiple comparisons, pairs of histopathological classes were compared using a

procedure based on a Scheffe-type method adapted to log-rank testing of censored survival distributions.

Results

Patient characteristics

A total of 40 patients, 25 female (62%), 15 male (38%), median age 52.5 (range 28–75) with diagnosis of primary appendiceal carcinoma were identified. The most common presenting symptoms were abdominal pain (62%) and abdominal distension (25%). Two patients (5%) had a preoperative diagnosis suggestive of intra-abdominal malignancy from imaging studies (Table 1).

Pathology

Twenty-seven (of 39) patients (69%) had pseudomyxoma peritonei at time of presentation. Fifteen patients' (38%) tumors were classified as DPAM, 18 (46%) as PMCA, and 6 (15%) as mixed PMCA I/D. One patient's pathology material was not available for review. Thirty-one (of 38) patients (82%) had T4 tumors on presentation, 5 (13%) patients were T3, and only 2 (5%) patients were stage T1 or T2. Lymph node involvement status was available for 11 of 14 patients with DPAM, 15 of 18 patients with PMCA, and in 5 of 6 patients with PMCA I/D. Table 2 summarizes the T, N, M stage and tumor grade by histological type.

Treatment

All patients were treated with surgery, chemotherapy, or both. Fifteen patients (38%) were treated with surgery alone which was either radical tumor resection with a right hemicolectomy or debulking surgery for palliation. Three patients (8%) were treated with systemic chemotherapy alone in a palliative setting. Twenty-two patients (55%) received combined surgical and chemotherapy modalities. Four (of 37) patients (11%) received peri-operative intraperitoneal chemotherapy along with systemic chemotherapy. Of the 25 patients (62%) receiving systemic chemotherapy, a majority (18 patients; 72%) received 1–2 lines of therapy. Chemotherapy administered in most cases was 5-fluorouracil-based regimens used in the treatment of colon cancer.

Treatment modalities according to tumor histology were also analyzed. Three patients with early stage DPAM tumors (T3 or less and without pseudomyxoma peritonei) were treated with surgery alone. Advanced DPAM tumors, T4 and pseudomyxoma peritonei on presentation ($n = 11$), were treated with either surgery alone (33%), a combination of surgery and systemic chemotherapy (42%) or with a combination of surgery, systemic chemotherapy and intraperitoneal chemotherapy (25%).

Of the 11 patients with PMCA and pseudomyxoma peritonei on presentation, 4 (36%) underwent debulking alone, 3 (27%) received palliative chemotherapy alone, 3 (27%) received a combination of surgery and systemic chemotherapy, and 1 (9%) patient received a combination of surgery, systemic chemotherapy, and intraperitoneal chemotherapy.

In patients with PMCA I/D, one patient with early stage tumor (T3 and no pseudomyxoma peritonei) was treated with a combination of surgery and chemotherapy. Of the five patients with advanced disease and pseudomyxoma peritonei on presentation, 3 (60%) were treated with a combination of surgery and systemic chemotherapy, while 2 (40%) underwent debulking.

Survival and histopathological classification

Among the 40 patients being followed up, 32 (80%) have expired with six patients (15%) alive and two patients (5%) lost to follow up at the time of analysis. Median follow-up for OS among the eight censored patients was 57 months. The median OS of the entire patient cohort was 25.2 months (95% CI: 14.2–46.4 months). The 2-year OS rate for all the patients was 52%, which dropped to 23% by 5 years. Further statistical details are provided in the legend for Fig. 1.

OS was highly statistically significantly different ($P < 0.0001$) according to the histopathological classification of appendiceal tumor (Fig. 2). OS duration of patients with DPAM was highly significantly different from OS of both patients with PMCA and patients with PMCA I/D ($P < 0.0001$ in each case). OS duration of patients with PMCA was not significantly different from OS of patients with PMCA I/D ($P = 0.0564$). The median OS for patients with DPAM was 7.7 years (90% CI: 2.9—upper limit not estimable). All 15 patients with DPAM were alive at the end of 2 years. The 5-year OS rate of the DPAM group was 62% (90% CI: 30–95%). Of the three patients with early stage tumors without pseudomyxoma peritonei on presentation, two are still alive with a median follow-up of 7 years and one patient died from other causes 9 years after the initial diagnosis.

The median OS of the patients with PMCA was 0.7 years (90% CI: 0.4–1.5 years). The 1- and 2-year survival rates for the PMCA group were 36% (90% CI: 17–55%) and 21% (90% CI: 5–37%), respectively. The median OS of the patients with PMCA I/D was 1.2 years (90% CI: 0.9–1.6 years). The 1- and 2-year survival rates for the PMCA I/D group were 70% (90% CI: 39–100%) and 15% (90% CI: 0–40%), respectively. There was only one patient with PMCA between both groups alive at 5 years, with that patient dying at 5.1 years.

Discussion

The limited information in the literature on the management of appendiceal malignancies makes it challenging to both appropriately diagnose and manage these cases [9, 12–15, 19]. Appropriate treatment decisions require a recognition of the different histopathological disease entities. In our single institutional study, we analyzed the outcomes of all patients with appendiceal carcinoma based upon the well characterized pathological parameters.

A preoperative diagnosis of appendiceal carcinoma is rarely made as seen in our series and demonstrated by others [9, 20]. The most common presenting symptoms are nonspecific abdominal pain and abdominal distension indicating an advanced stage of disease because of invasion into the peritoneal cavity. Often these patients are preoperatively diagnosed with appendicitis or right lower quadrant tumor abscess before appendiceal carcinoma is found on surgery, with the possibility of ensuing suboptimal surgical management [9, 21].

Appendiceal carcinomas frequently present with pseudomyxoma peritonei, which was seen in 69% of our patients. A characteristic feature of appendiceal carcinoma verified by our series was that none of the patients with advanced DPAM or mixed PMCA I/D tumors had evidence of lymph node involvement, a feature clearly distinct from colon carcinoma.

Appendiceal carcinomas should be viewed as a spectrum of different diseases with differing outcomes rather than a single entity. As clearly demonstrated in our series and corroborated by others, appendiceal carcinomas may range from the low-grade DPAM to the intermediate, mixed PMCA I/D tumors and at the other end of the spectrum the aggressive PMCA tumors. Tumors that have the low-grade, noninvasive histology, classified as DPAM have a prolonged benign course with local peritoneal recurrences over many years. In our

series, the median OS and 5-year survival for patients with DPAM was 7.7 years and 62%, respectively. A similar finding of improved survival in the well-differentiated tumor groups was seen in other case series as well [12–14, 17]. In their original report, Ronnett et al. [17] noted the age-adjusted 5-year survival rates were 84% for patients with the histologically bland, low-grade DPAM. In a subsequent report with longer follow-up and updated survival data [14], patients with DPAM had 5- and 10-year survival rates of 75 and 68%, respectively, with a median survival not reached after 104 months of follow-up.

On the other hand, poorly differentiated, invasive tumors with complex architecture classified as PMCA are more aggressive with frequent parenchymal organ and lymph node involvement and significant morbidity and mortality from intra-abdominal carcinomatosis. In our series, the median OS and 2-year survival was 0.7 years and 21%, respectively. In the series by Ronnett et al., the 5-year survival rate for PMCA was 6.7%. Tumors with mixed histologies such as PMCA I/D do not have frequent lymph node or parenchymal organ involvement yet appear to follow a more aggressive clinical course than the low-grade, noninvasive histologies. In our series, the median and 2-year survival of patients with PMCA I/D were 1.2 years and 15%, respectively, in comparison with the previously reported 5-year survival of 37.6%. Therefore, tumor histology was strongly related to survival in all patients in our series confirming the results of previously reported series [12–15, 17].

Over the past 15 years at our institution, the management of appendiceal carcinomas has included surgery, systemic chemotherapy, and intraperitoneal chemotherapy. This variability in the management of this disease in our series is due to the lack of randomized, controlled trials that define the optimal management of appendiceal carcinomas, the different histologies and presence or absence of peritoneal lesions. Patients with well-differentiated, noninvasive DPAM appendiceal tumors confined to the appendix are usually treated by resection alone and may not require any further intravenous or intraperitoneal chemotherapy. Curative resection in such cases would include right hemicolectomy with regional lymph node dissection, although the chance of nodal spread is quite small. In our series, the three patients with DPAM and early stage disease were treated with surgery alone with excellent outcome. Greater variability was seen in the management of patients with DPAM and peritoneal disease. In this group, surgery alone, surgery and systemic chemotherapy, or intraperitoneal chemotherapy was used in 33, 42, and 25% of patients, respectively. Patients with peritoneal dissemination of these well-differentiated tumors (DPAM) are most likely to benefit from an aggressive peritoneal directed treatment strategy consisting of resection of the primary tumor combined with complete peritonectomy and planned peri-operative intraperitoneal chemotherapy [22]. The completeness of cytoreduction correlates with survival, with patients achieving a complete cytoreduction doing significantly better than patients with incomplete cytoreduction [9, 22, 23]. Sugarbaker et al. [8] reported a survival of 70% at 20 years for patients with the indolent appendiceal pseudomyxoma peritonei syndrome with cytoreductive surgery and intraperitoneal chemotherapy which is vastly different from results with serial debulking. Failure to make the diagnosis of DPAM preoperatively occurs frequently in this disease and was reflected by our series. That limits the ability to plan for appropriate cytoreductive surgery with intraperitoneal chemotherapy. The role of systemic chemotherapy in this setting is less established especially since parenchymal distant metastasis is rarely seen.

Aggressive, complex tumors such as PMCA are best treated with curative resection only if confined to the appendix. PMCA tumors invading through the appendiceal wall or with lymph node involvement may benefit from adjuvant chemotherapy after optimal surgical resection, given their otherwise poor outcomes. Chemotherapy used should be in line with the recommendations for similar stage colon cancer. Palliative chemotherapy along with

palliative tumor debulking may be needed for more aggressive tumor types with widely distributed invasive cancer to avoid excessive morbidity and mortality [24]. In our series, 8 of the 11 patients presenting with PMCA and peritoneal spread received systemic chemotherapy as part of their initial management. The chemotherapy used was 5-FU based. Despite the early use of chemotherapy the outcome of this group of patients was relatively poor (median OS of 0.7 years) in comparison with the patients with DPAM or stage IV colorectal cancer. This indicates that the biology of PMCA is different than for colorectal cancer and consequently chemotherapeutic approaches specific for this disease need to be developed. Tumors with mixed histologies such as PMCA I/D follow an intermediate clinical course between DPAM and PMCA and are usually treated in the same way as are the more poorly differentiated tumors. For the early stage disease, surgical resection with adjuvant therapy for T3 or N1 disease is commonly used as was in our series. Palliative debulking and chemotherapy was used in 60% of our patients with advanced stage PMCA I/D.

The differing tumor behavior and outcomes between the indolent DPAM tumor types and the more aggressive mucinous adenocarcinoma type (PMCA) indicates an inherent biological difference between the two tumor types and highlights the need for an experienced pathologist for the accurate identification and classification of appendiceal neoplasms. Appropriate preoperative diagnosis of these tumors will allow for optimal patient management. Clinical trials evaluating systemic therapy in patients with PMCA and PMCA I/D histologies are needed.

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Fig. 1.

Kaplan–Meier graph of OS for 40 appendiceal carcinoma patients. *Tick marks* represent censored patients still alive. The median OS was 25.2 months, with 95% CI 14.2– 16.4 months. The 1-year OS rate was 69%, with 95% CI 0.54– 0.83. The 2-year OS rate was 52%, with 95% CI 0.37–0.68. The 5-year OS rate was 23%, with 95% CI 0.09–0.37. Three patients were still alive beyond 5 years, at 6.0, 7.7, and 19.0 years after their first visit to the Karmanos Cancer Center

Fig. 2.

Kaplan-Meier graph of OS of appendiceal carcinoma patients by histopathological classification. *Tick marks* represent censored patients still alive. The median OS was 7.7 years (90% CI: 2.9—upper limit not estimable) for patients with DPAM; 0.7 years (90% CI: 0.4–1.5) for patients with PMCA; and 1.2 years (90% CI: 0.9–1.6) for patients with PMCA I/D. The 1-year OS rates for DPAM, PMCA, and PMCA I/D were 100, 36, and 70%, respectively. The 2-year OS rates for DPAM, PMCA, and PMCA I/D were 100, 21, and 15%, respectively

Table 1

Frequency of signs and symptoms in patients at presentation

Chief complaint	Frequency
Nausea, vomiting	1 (3%)
Abdominal pain	25 (62%)
Abdominal distension	10 (25%)
Hematochezia	1 (3%)
Radiological finding	2 (5%)

Percentages do not sum to 100 due to rounding

Table 2

Pathological characteristics of 39 tumors according to the histological classification

Pathological parameter	DPAM (N = 15)	PMCA (N = 18)	PMCA I/D (N = 6)
Tumor stage ^a			
T1	1 (7%)	0	0
T2	1 (7%)	0	0
T3	1 (7%)	3 (17%)	1 (17%)
T4	11 (79%)	15 (83%)	5 (83%)
Lymph node involvement ^b			
0	11 (100%)	5 (33%)	5 (100%)
1–3	0	7 (47%)	0
4	0	3 (20%)	0
Grade of differentiation			
Well differentiated	14 (93%)	5 (28%)	3 (50%)
Moderately differentiated	1 (7%)	5 (28%)	3 (50%)
Poorly differentiated	0	8 (44%)	0
Pseudomyxoma peritonei on presentation	11 (73%)	11 (61%)	5 (83%)

DPAM, disseminated peritoneal adenomucinosis; PMCA, peritoneal mucinous carcinomatosis; PMCA I/D, peritoneal mucinous carcinomatosis with intermediate or discordant features

^aTumor stage was unavailable for one of the 15 patients with DPAM

^bReported for the 31 patients who had lymph node status assessed