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Management of appendiceal carcinoid tumors in children

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ABSTRACT

Background: Appendiceal carcinoid tumors, also know as well-differentiated neuroendocrine neoplasms, are rare lesions detected incidentally after appendectomy in children. There are limited data about the natural history of these tumors, and guidelines regarding family counseling and need for additional surgery or follow-up imaging are not established in the pediatric age group. The purpose of this study was to review our institutional experience with appendiceal carcinoid tumors to provide data that might improve management.

Methods: After institutional review board approval, the charts of all patients treated at our institution for an appendiceal carcinoid tumor between 2002 and 2014 were reviewed. Data collected included patient demographics, pathologic details, postoperative management, and follow-up information. Descriptive analyses were performed.

Results: Twenty-eight patients were identified, which represents an incidence of 0.2% of children undergoing appendectomy during that time interval. The mean age at surgery was 13.8 ± 2.1 y; 54% were females. Two patients had symptoms suspicious for carcinoid syndrome at presentation, though none had evidence of metastatic disease. The mean tumor size was 0.73 ± 0.4 cm. Five patients (18%) underwent subsequent ileocecectomy or right hemicolectomy because of pathologic findings of invasion of the mesoappendix ($n = 4$) or lymphovascular invasion and subserosal extension ($n = 1$), two of whom had residual disease in the resected specimen (one in a lymph node). No recurrences have been detected at mean follow-up of 1.8 y.

Conclusions: Appendiceal carcinoid tumors are discovered incidentally in about 0.2% of children undergoing appendectomy. Based on findings from a large contemporary series, we can conclude that these tumors are generally small and demonstrate lymphovascular invasion or mesenteric extension in fewer than 20% of cases. Prospective, multicenter studies are necessary to better define the indication for ileocecectomy and follow-up imaging protocols.

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1. Background

Appendiceal carcinoid tumors, also known as well-differentiated neuroendocrine neoplasms, are rare, usually incidentally detected lesions in children at the time of appendectomy for acute appendicitis with a reported incidence of less than 0.1% [1]. Unlike in adults, where algorithms for additional surgery after incidental detection and postoperative surveillance are defined by a consensus guideline published in 2010 by the North American Neuroendocrine Tumor Society, no such recommendations exist in children [2]. A recent review in 2013 provided an algorithm for management in children, but these recommendations appear to be based on data from adult patients [1].

Barriers to the development of treatment algorithms in children include the rarity of the disease with limited single-institution experience and a lack of a defined natural history of these tumors [3]. Additionally, comparison to adult literature has limitations as these tumors behave differently in children [4,5]. At our institution, postoperative management guidelines have been developed for patients with acute appendicitis. During the past year, four patients have been diagnosed with carcinoid tumors on pathologic review of the appendix specimen, and best practice recommendations were not included in our current protocol. The purpose of this study was to review our institutional experience with these tumors to determine the approximate incidence, the pathologic details of the tumors, the percentage of patients who underwent additional procedures, and long-term patient results so as to update our patient management protocols for this small yet significant group of children.

2. Methods

The study is a retrospective case series of patients diagnosed with an appendiceal carcinoid tumor from 2002–2014 at Texas Children's Hospital. Patients were identified through a query of the pathology records. A chart review was performed to extract data regarding patient demographics, pathologic details, hospital course, and follow-up information. After institutional review board approval, patients who had been lost to follow-up were contacted to determine if additional imaging or treatment had been obtained at another institution. Descriptive analyses were performed.

3. Results

Twenty-eight patients were identified during the study period, representing approximately 0.2% (28 of 11,460) of patients treated for acute appendicitis during that period. The patients were 54% female (15/28) and presented at a mean age of 13.8 ± 2.1 y (median 14.0, range 10.3–17.4) y. The most common presenting symptom was abdominal pain (89%), followed by nausea and/or vomiting (43%) and fever (18%). Although most patients presented with the typical acute appendicitis report of pain for 1–3 d, four patients reported abdominal pain for >1 wk.

All patients underwent initial laparoscopic appendectomy. No patients were suspected of having a carcinoid tumor at the time of operation. On review of the pathology specimens, 13 of 28 patients (46%) had evidence of appendicitis (10 simple, 3 perforated). The mean tumor size was 0.73 ± 0.4 cm (median 0.7 cm, range 0.2–1.6 cm). Only 4 patients had a tumor size of >1.0 cm, and none were >2.0 cm. Of those patients with the depth of extension of the tumor reported, 3 extended to the submucosa, 1 to the muscularis propria, 5 to the subserosa, 6 to the serosa, and 7 to the mesoappendix. No patients had positive margins at the base of the appendix specimen. On review of the specimens, 2 (7%) had lymphovascular invasion and 4 (14%) had perineural invasion.

Five patients underwent additional surgical resection; one received an ileocecetomy and four received a right hemicolectomy, performed 2 wk–2 mo after the initial appendectomy. In the pathology specimens of the additional resection, one patient had additional tumor in the mesentery of the cecum without lymph node disease. In another patient, 1/43 lymph nodes were positive, but there was no further tumor in the resected colon. The other three patients were free from further nodal or residual colonic disease. All patients recovered well from their colectomies with a mean hospital stay of 5.4 d (median 5 d, range 4–7 d). One patient developed an infected hematoma postoperatively that was drained percutaneously and treated with intravenous antibiotics.

Other than the patient with the positive lymph node, there was no evidence of metastatic or synchronous disease. Two patients had symptoms that in retrospect would have been suspicious for carcinoid syndrome (one with flushing and diarrhea, the other with diarrhea), but these patients did not have evidence of metastatic disease. The mean follow-up for the study was 1.8 y (median 8 mo, range <1–10 y). On contacting those patients who were lost to follow-up (5/15 contacted), none had received additional procedures at another institution. One patient had a repeat computed tomography scan for abdominal pain that was unremarkable. Nine of the twenty-eight patients had repeat postoperative imaging (computed tomography and/or ultrasound) without detection of additional disease.

4. Discussion

The current largest pediatric series of appendiceal carcinoid tumors has 47 patients (the mean number of patients in 15 series, including our own, is 20 patients), and no prospective data or trials regarding this patient population have been reported [6]. As a result, recommendations regarding management of these tumors in children are derived from adult data. In adults and children, patients with tumors <1.0 cm may be treated with appendectomy alone [1]. The treatment of those with a 1.0–2.0 cm tumor is less clear in both the pediatric and adult populations, and additional resection is based on patient-specific risk factors [2,3,7].

General indications for more extensive resection are based on the risk of more invasive disease and include size >2.0 cm,