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Carcinoid tumours of the appendix in children having appendicectomies at Princess Margaret Hospital since 1995

Alecia Vandevelde *, Parshotam Gera [1]

Princess Margaret Hospital for Children, General Surgical Department, Roberts Road, Subiaco, Perth, Western Australia 6008

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ABSTRACT

Aim: The diagnosis of carcinoid tumour is a relatively rare one. Our surgical approach has changed over the last two decades from predominantly open to predominately laparoscopic with a tendency to leave the mesoappendix in situ. The aim of this audit was to identify how many cases we had at PMH and to see whether the shift in surgical approach allowed us to make prognostic decisions in keeping with current best practice and whether this made any difference in further surgery requirements or outcome for patients.
Methods: A retrospective review of all cases of carcinoid identified in our search of all appendicectomy histopathology results was conducted. Results were compared to those found in other studies. Duration of follow up and further investigations was reviewed, as was whether or not there was any recurrence.
Results: Our incidence of carcinoid tumours in patients undergoing appendicectomy since 1995 was 0.35%, similar to that in other centres. None of our patients had surgery beyond an appendicectomy and our active follow up varied from none to 6 months. There were no recurrences in this time.
Conclusions: The literature review carried out suggests further meta-analysis is needed including data on long term follow up before definitive guidelines regarding extent of surgical treatment and follow up based on histopathology are created. The condition is rare and the studies small, resulting in no clear consensus on the best practice for tumours measuring between 1 and 1.5 cm in diameter. Our surgical approach to appendicectomies has changed; it is unclear whether this has resulted in a change in outcome.

1. Background

Carcinoid tumours are found throughout the gastrointestinal and bronchopulmonary system and are the most common tumour of the appendix in adults and the gastrointestinal tract in children accounting for 0.1% of all paediatric malignancies. Despite this they are still relatively rare with a quoted incidence of 1 in 100,000 to 1 in 1 million per year [1–4]. Two distinct entities exist, the more common well differentiated endocrine tumour, and the poorly differentiated goblet cell tumour [5]. The majority of papers reviewed found the rate of carcinoid tumours in children to be approximately 0.3%–0.9% of those undergoing appendicectomies, in most cases for presumed appendicitis [6–8]. The classic carcinoid syndrome caused by serotonin release involving flushing, diarrhoea, bronchospasm, and valvular heart disease is rare [5,9]. In one analysis of all children presenting to paediatric oncology centre only one child presented with carcinoid syndrome; the majority present with symptoms suggestive of appendicitis.

The current practice at Princess Margaret hospital is to skeletonise the appendix and leave the mesoappendix in situ. The question regarding adequacy of surgical specimens, including nodal sampling, and analysis of size is an interesting one, and is integral in deciding whether we are able to make adequate prognostic decisions based on our current practice or whether we should adjust our practice to include resection of the mesoappendix.

2. Methods

A retrospective analysis of all histologically proven cases of carcinoid tumours of the appendix at PMH over the last 19 years using our histopathology laboratory online surgical specimen database was performed. This data base was also used to provide the total number of appendicectomies carried out. This initial search was followed by analysis of individual patient notes with data collected regarding presentation, surgical technique, and follow up. We then compared our results with those obtained at other centres through review of current literature. The literature review was carried out using PubMed and Ovid with the search terms 'carcinoid' and 'children' and restricted to full text articles published since 1960. We then limited this to articles concerning appendiceal carcinoid tumours only. Appendicectomies using both laparoscopic and open techniques were included.

3. Results

Since 1995 twenty two cases of carcinoid tumour were encountered in patients undergoing appendicectomy at PMH. Over this 18 year

* Corresponding author at: Royal Darwin Hospital, Rocklands Drive Tiwi, PO Box 41326 NT 0811. Tel.: +61 8 89228888; fax: +61 8 89228286.
E-mail addresses: alecia.vandevelde@health.wa.gov.au (A. Vandevelde), Parshotam.gera@health.wa.gov.au (P. Gera).
[1] GPO Box D184, Perth, Western Australia, 6840. Tel.: +61 8 9340822.

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Table 1
ENETS and AJCC carcinoid of appendix staging.

ENETS	UICC/AJCC	
T	Primary tumour	
X	Primary tumour cannot be assessed	
0	No evidence of primary tumour	
1	tumour </= 1 cm invading submucosa and muscularis propria	
1a	tumour </= 1 cm in greatest dimension	
1b	tumour > 1 cm but </= 2 cm in greatest dimension	
2	tumour </= 2 cm invading submucosa, muscularis propr. and/or minimally (up to 3 mm) invading subserosa/mesoappendix	tumour > 2 cm but </= 4 cm or with extension to the caecum
3	tumour > 2 cm and/or extensive (>3 mm) invasion of subserosa/mesoappendix	tumour > 4 cm or with extension to the ileum
4	tumour invades peritoneum/other organs	tumour perforates peritoneum or invades other adjacent organs or structures e.g. abdominal wall and skeletal muscle.
N	regional lymph node metastasis	
X	regional lymph nodes cannot be assessed	
0	no regional lymph node metastasis	
1	regional lymph node metastasis	
M	distant metastasis	
X	distant metastasis cannot be assessed	
0	no distant metastasis	
1	distant metastasis	

ENETS Stage			
stage 0	Tis	N0	M0
stage I	T1	N0	M0
stage IIa	T2	N0	M0
stage IIb	T3	N0	M0
stage IIIa	T4	N0	M0
stage IIIb	any T	N1	M0
stage IV	any T	any N	M1
UICC/AJCC Stage			
stage I	T1	N0	M0
stage II	T2–3	N0	M0
stage III	T4	N0	M0
	any T	N1	M0
stage IV	any T	any N	M1

period 6236 appendicectomies were carried out. The resulting incidence during this time was 0.35% of those undergoing appendicectomies. In all except two cases the presumed diagnosis preoperatively was appendicitis. The other two were thought to be most likely gynaecological in nature. One case was excluded as there were no notes electronic or otherwise regarding the patient available.

All tumours were less than 2 cm with the median size being 6.5 mm. Two of the cases did not have a size recorded in the report and instead were described as being microscopic and less than 2 cm. 23.8% were larger than 1 cm in size, with only one larger than 1.5 cm. All had clear proximal surgical margins. The mesoappendix was removed in 10 cases, all of which were carried out in an open manner. In 5 cases there was mesoappendix involvement but no comment on any nodes in the histopathology reports on these patients.

In all cases the presenting symptom was abdominal pain, and the vast majority were acute or less than 1 week in duration. Intraoperatively the presumed diagnosis was appendicitis 75% of the time. None of our patients underwent a right hemicolectomy. The patient was seen by or discussed with the oncologists 47% of the time. The mean age of our patients was 12.5 years.

Intraoperatively a tumour was suspected in one case, and on one occasion enlarged nodes were noted intraoperatively however not excised. This patient had no appendiceal inflammation on histopathology but only a 'microscopic' sized tumour. The majority of cases were diagnosed as appendicitis intraoperatively, and in 80% of cases there was appendiceal inflammation histologically.

Our active follow up time varied from no follow up at all post discharge to 6 months, with an average period of 4 weeks. There were no recurrences during this time. PMH is however the only tertiary paediatric hospital in Western Australia. And given that all public hospital admissions are accessible online with the same unique medical record number for each patient it is reasonable to assume that no further presentations to hospital imply that there was no recurrence. If the date the histology was obtained until the date of the audit was carried out is considered then it could be assumed that follow up for purposes of this study was much longer, with a mean of 95 months, or eight years, and a median of 87 months, or 7 years. During this time none of the cases had a recurrence.

4. Discussion

The low incidence of carcinoid tumours in children means that the formulation of definitive management guidelines is difficult thereby creating a surgical management dilemma. The diagnosis of carcinoid tumour of the appendix carries a good prognosis generally [1,3]. A large study by SEER (Surveillance, Epidemiology, and End Results program) found an 84% five year survival rate in those with the condition however this particular study included adults [1]. In children the tumours most frequently involve the tip of the appendix; survival rates in this situation are higher at 92%, and studies specific to the paediatric population report the five year survival rate to be closer to 100% [2,10]. Where the rate is less than 100% it has not been made clear by the authors whether the deaths reported have been due to metastasis from the appendiceal carcinoid itself or from concomitant neoplasms and other causes. Most studies conclude that important risk factors for recurrence and metastasis are size and infiltration of the mesoappendix [10,11]. Other factors that have been found to be of prognostic significance include age, the presence of hepatic metastases, and lymph node involvement [3]. It is recommended that screening for distant metastases be undertaken in those with tumours larger than 2 cm [12].

The changing TNM and WHO classifications over the last decade reflects the persistent uncertainty and the increasing knowledge