ORIGINAL ARTICLE

Carcinoid tumor of appendix present as acute appendicitis: a single centre retrospective analysis

Irum Masood, Haris Rasheed, Anum Naz, Ahmed Raheem

Received:
5th May 2017

Accepted:
15th November 2017

Abstract

Background: Carcinoid tumors are neuro-endocrine origin and appendix is one of its common sites. Carcinoid tumor of the appendix still pursue with the curiosity in spite of their low occurrence rate. Nearly all appendiceal carcinoids pretty much always silent on clinical presentation and found incidentally at histopathology. Our study focus was to present our experience clinico-pathological behavior, dissemination of tumor and treatment protocols of carcinoid tumor of appendix as managed at university teaching hospital.

Material and Methods: A retrospective study of 1,406 appendectomies performed in tertiary care single centre in Ziauddin University and hospital Karachi, considered period from August 2005 to December 2016. Study of 11 patients with histo-pathologically established diagnosis of carcinoid tumor of the appendix. Reviewed the surgical records, analysis of demographic data, clinical presentation, histopathology, operative records and consequences was presented.

Result: Among the 1,406 appendectomies 11(0.78%) carcinoid tumor of appendix were identified, 7 (58.3%) male and 5 (41.6%) female patients. All the patients with a carcinoid tumor posed with the characteristics indicative of acute appendicitis, diagnosis established on histopathology after appendectomy. Additionally no more surgery was necessitated while all these tumors < 1cm and base of the appendix was not involved.

Conclusion: Our data confirmed the good prognosis as reported in literature. Nearly all carcinoid tumor of appendix frequently present as acute appendicitis. In most of the cases carcinoid found incidentally in histopathology and diagnosis is presumed seldom prior to histopathology. Carcinoid tumor of appendix can be cured by simple appendectomy if tumor size is less than 1 cm.

Keywords: Carcinoid tumors, appendectomy, neuro-endocrine tumors, appendiceal carcinoid, acute appendicitis

Ziauddin University and Hospital, Karachi.
I Masood
H Rasheed

Abbasi Shaheed Hospital, Karachi
A Naz

Agha khan University and Hospital Karachi
A Raheem

Correspondence:
Dr Irum Masood,
Department of General Surgery, Ziauddin University and Hospital Karachi.
Cell: + 92-
Email: drirum3@gmail.com

Introduction:

Carcinoid tumors are the most frequently occurring neuroendocrine tumors of the gastrointestinal tract and area of persistent curiosity in the surgery field. Carcinoid tumors are originated from primitive stem cells in the wall of gastrointestinal tract. Despite that it may be found in some other sites such as the lungs, mediastinum, liver, pancreas, thymus, bronchus, kidneys, ovaries, and prostate. Carcinoid tumor is most frequent malignancy affecting the appendix.[1-7] Usually arise from sub-epithelial endocrine cells in the lamina propria and submucosa.[8,9] Histopathologically carcinoid tumor of appendix is mostly entero-chromaffin type cells and dissimilar from neuro-endocrine tumor of other organs.[10] However it is occasional entity and incidentally detected in histo-pathology after appendectomy. It is observed the most frequent type of primary tumor of appendix and found in 0.3 to 0.9% of patients, experienced appendicectomy surgery.[11]

Material and Methods:

A retrospective study of 1,406 appendectomies performed in tertiary care single centre in Ziaud-

Table 1: Characteristics of Carcinoid Tumor of appendix

Carcinoid tumor of appendix	n=1406 appendectomies
Incidence Rate n (%)	11(0.78%)
Male	7 (58.3%)
Female	5 (41.6%)
Gender Ratio	
M/F	1.4:1
Mean age + SD	
Male	27.4 + 13.41
Female	37.8 + 9.36
Location	
Tip of the appendix	8(72.7%)
Middle third of appendix	3(27.7%)
Mean Tumor Size (diameter)/ Range	0.4 cm (0.2-1cm)
Less than 0.5 cm	5(45.4%)
Less than 1 cm	6(54.5%)
Clinical Presentation	
Acute Appendicitis	11(100%)
Other sign symptoms	0(0%)
Management/Surgery	
Appendectomy	11(100%)
Other Surgery/Further surgery	0(0%)
Histopathology (Tumor Infiltration)	
Submucosa and Muscularis propria	9(81.8%)
Serosa	2(18.18%)

din University and hospital Karachi, considered period from August 2005 to December 2016. Study of eleven patients with histopathologically established diagnosis of carcinoid tumor of the appendix. Reviewed the surgical records, analysis of demographic data, clinical presentation, histopathology, operative records and consequences was presented.

Results:

Among the 1,406 appendectomies 646(45.94%) female and 760(54.05%) was male, 11(0.78%) appendiceal carcinoid tumors were identified, 7 (58.3%) male and 5 (41.6%) female patients with a mean age+SD is 27.4+13.41 and 37.8+9.36 years respectively, range 42(12-54) and 23(25-48) years respectively. Male and female ratio is 1.4:1. All the patients with a carcinoid tumor posed with the characteristics indicative of acute appendicitis, diagnosis established on histopathology after appendectomy. Additionally no more surgery was necessitated while all these tumors < 1cm and base of the appendix was not involved. In (8/11) patients the common site of tumor in appendix was tip of the appendix with a mean diameter of tumor was 0.4 cm (range 0.2-1.0 cm) and (3/11) located in the middle third. The tumor was less than 0.5 cm in diameter in five patients and less than 1 cm in diameter in six patients. The tumor infiltration in 9 patients was limited to the submucosa and muscularis propria, but serosa was involved in 2 patients. There was no proof of tumor invaded into the mesoappendicular fat, no lymph node involvement, no metastases seen in any patient. The 3-years follow up was accomplished in 2 patients, 3 were completed one years follow up and 6 patients were lost during follow up. All 5 patients those completed follow up were alive, well and disease free during a follow up.

Discussion:

Carcinoid tumor of appendix is a distinctive tumor and dissimilar from that are confront at a different place in the gastrointestinal tract distinguished as it is a quite frequent, mostly sluggish behavior, manifestation in adolescents and female supremacy tendency.[12,13] Carcinoid tumor of appendix regarded as 32 to 57% of all tumors of appendix.[12,14] It frequently find unexpectedly throughout the 4th or 5th decade of life even as an incidental intra-operatively but surgery carried out for some other intention.[15] One previous study reported, mean age of tumor is 42- year.[9] In literature reported a male preponderance with male to female ratio is 4:1.[16,17] Almost all patients are asymptomatic with carcinoid tumor of the appendix, probably due to the exact site of tumor in tie-up to the base of the appendix; tremendously 75% are situated in distal third of the appendix, there is danger of obstruction is less, rest of situated in the middle third, and < 10% situated at the base of the appendix.[15] This tumor had vague and clinically non-bspecific presentation, possibly not distinguishable from the features of acute appendicitis. This tumor mostly diagnosed incidentally intra-operatively or post-operatively in histopathology after surgery as treated for acute appendicitis.[17,18] Another study reported the same.[16] As in our study we had similar findings. Carcinoid syndrome occurs with carcinoid tumor in < 10% of patients.[19] In literature, carcinoid syndrome may be seen in patients with

liver metastases in < 2% of carcinoid tumor of appendix.[18] Carcinoid tumor manifested with minimal metastatic capability and for that reason it is found infrequently with metastases.[8,17] It is a feature characteristic of carcinoid tumor previse aggressive response comprises histological sub-type, size of tumor and tumor insertion in mesoappendix.[11] As stated in latest guidelines, only appendectomy is sufficient for the management of carcinoid tumor if tumor is less than l cm. Extensive surgery indicated if tumor size is greater than 2cm, involvement of lymph node or mesoappendix, if resection margins is tumor positive, and high mitosis with cellular pleomorphism.[20] Endanger of tumor metastasis is zero if tumor ≤ 1 cm, increase danger of metastasis if tumor ≥ 2 cm, metastasis set out from 20%-85%.[21] Almost 1/3 of carcinoid tumor if greater than 2 cm, presents with nodal and distant metastasis, and right hemicolectomy is indication as an adequate management[8,20,22] Treatment of the tumor if size is 1-2 cm still continue to talk over; so additionally extensive management with right hemicolectomy is frequently put into practice predominantly in adolescents.[22,23] Further prognostic characteristics like gender and depth of tumor infiltration may be employed into management as appendectomy or right hemicolectomy.[24] The enormous patients of carcinoid tumor of appendix no need of any more distant surgery or treatment after simple appendectomy. The prognosis of carcinoid tumor of appendix is considerably better than carcinoid tumor of midgut.[19] The reported calculated survival is five years for localized tumor, regional metastasis and distant metastasis to be 92%, 81% and 31% respectively.[7] In our study all the patients were alive and tumor free through a follow up of 36 months. Briefly carcinoid tumor of appendix appeared most frequently as sign and symptoms of acute appendicitis. In almost all cases it is found incidentally in histopathology and its diagnosis is hardly presumed prior to histopathology evaluation. In spite of the fact that carcinoid tumor of appendix has an extremely good prognosis but attention for regular screening should be given for these patients because frequency of correlated synchronous tumor and metachronous colorectal tumor is outrageous 13 to 33%.[7,8,12,14]

Conclusion:

Our data supported the good prognosis as reported in literature. Nearly all carcinoid tumor of appendix frequently presented as acute appendicitis. In most of the cases carcinoid found incidentally in histopathology and diagnosis is presumed seldom prior to histopathology. Carcinoid tumor of appendix can be managed by only appendectomy if tumor size is less than 01cm and base of the appendix was not engulfed.

Conflict of interest: None

Funding source: None

Role and contribution of authors:

Dr Irum Masood, Ziauddin University and hospital Karachi, design the study, data collection, tabulate and write-up introduction, discussion, result and conclusion.

Dr Haris Rasheed, Consultant and laparoscopic General surgeon, Ziauddin University and hospital Karachi, supervise, design the study, initial methodology and review.

Ahmed Raheem Statistician Department of Pathology & Laboratory Medicine Agha khan University and Hospital Karachi, Statistical analysis

Dr Anum Naz, General Surgery Department Abbasi Shaheed Hospital Karachi, Data collection, write up results and Review.

References:

1. Broaddus RR, Herzog CE, Hicks MJ. Neuroendocrine tumors (carcinoid and neuroendocrine carcinoma) presenting at extra-appendiceal sites in childhood and adolescence. Arch Pathol Lab Med. 2003;127(9):1200-3.
2. Moraes TJ, Langer JC, Forte V, Shayan K, Sweezey N. Pediatric pulmonary carcinoid: a case report and review of the literature. Pediatr Pulmonol. 2003;35(4):318-22.
3. Soga J, Yakuwa Y, Osaka M. Evaluation of 342 cases of mediastinal/thymic carcinoids collected from literature: a comparative study between typical carcinoids and atypical varieties. Ann Thorac Cardiovasc Surg. 1999;5(5):285-92.
4. Piura B, Dgani R, Zalel Y, Nemet D, Yanai-Inbar I, Cohen Y, et al. Malignant germ cell tumors of the ovary: a study of 20 cases. J Surg Oncol. 1995;59(3):155-61.
5. Murali R, Kneale K, Lalak N, Delprado W. Carcinoid tumors of the urinary tract and prostate. Arch Pathol Lab Med 2006;130(11):1693-706.

6. Romero FR, Rais-Bahrami S, Permpongkosol S, Fine SW, Kohanim S, Jarrett TW. Primary carcinoid tumors of the kidney. J Urol. 2006;176(6):2359-66.
7. Modlin IM, Lye KD, Kidd M. A 5-decade analysis of 13,715 carcinoid tumors. Cancer. 2003;97(4):934-59.
8. Moertel CG, Dockerty MB, Judd ES. Carcinoid tumors of the vermiform appendix. Cancer. 1968;21(2):270-8.
9. Lundqvist M, Wilander E. Subepithelial neuroendocrine cells and carcinoid tumours of the human small intestine and appendix. A comparative immunohistochemical study with regard to serotonin, neuron-specific enolase and S-100 protein reactivity. J Pathol 1986;148(2):141-7.
10. Stinner B, Rothmund M. Neuroendocrine tumours (carcinoids) of the appendix. Best Pract Res Clin Gastroenterol 2005;19(5):729-38.
11. Goede A, Caplin M, Winslet M. Carcinoid tumour of the appendix. Br J Surg 2003;90(11):1317-22.
12. Sandor A, Modlin IM. A retrospective analysis of 1570 appendiceal carcinoids. Am J Gastroenterol. 1998;93(3):422-8.
13. Carr NJ, Sobin LH, editors. Neuroendocrine tumors of the appendix. Semin Diagn Pathol 2004: Elsevier.
14. Connor S, Hanna G, Frizelle F. Appendiceal tumors. Dis Colon Rectum 1998;41(1):75-80.
15. Kulke MH, Mayer RJ. Carcinoid tumors. N Engl J Med. 1999;340(11):858-68.
16. Tchana-Sato V, Detry O, Polus M, Thiry A, Detroz B, Maweja S, et al. Carcinoid tumor of the appendix: a consecutive series from 1237 appendectomies. World J Gastroenterol 2006;12(41):6699-701.
17. Roggo A, Wood WC, Ottinger LW. Carcinoid tumors of the appendix. Ann Surg 1993;217(4):385.
18. Spallitta S, Termine G, Stella M, Calistro V, Marozzi P. [Carcinoid of the appendix. A case report]. Minerva Chir. 1999;55(1-2):77-87.
19. Sweeney JF, Rosemurgy AS. Carcinoid tumors of the gut. Cancer control. 1997;4(1):18-24.
20. Fornaro R, Frascio M, Sticchi C, De Salvo L, Stabilini C, Mandolfino F, et al. Appendectomy or right hemicolectomy in the treatment of appendiceal carcinoid tumors? Tumori. 2007;93(6):587.
21. Thompson G, Van Heerden J, Martin Jr J, Schutt A, Ilstrup D, Carney J. Carcinoid tumors of the gastrointestinal tract: presentation, management, and prognosis. Surgery. 1985;98(6):1054-63.
22. McGory ML, Maggard MA, Kang H, O'Connell JB, Ko CY. Malignancies of the appendix: beyond case series reports. Diseases of the colon & rectum. 2005;48(12):2264-71.
23. MacGillivray DC, Heaton RB, Rushin JM, Cruess DF. Distant metastasis from a carcinoid tumor of the appendix less than one centimeter in size. Surgery 1992;111(4):466-71.
24. Shebani KO, Souba WW, Finkelstein DM, Stark PC, Elgadi KM, Tanabe KK, et al. Prognosis and survival in patients with gastrointestinal tract carcinoid tumors. Ann Surg 1999;229(6):815.