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Short Communication

Carcinoid tumor of appendix; retrospective review of 1800 appendectomy patients

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ABSTRACT
Malignant tumors of the appendix are rare and the histopathological diagnosis can often be made in the postoperative period. The clinical records of 1800 patients who underwent surgery in emergency conditions with a preliminary diagnosis of acute appendicitis were reviewed retrospectively. Carcinoid tumor was detected in 7 patients (0.38%) on histopathological examination. Of these patients, 4 were male (57.1%), 3 were female (42.9) and the mean age was found to be 36 (22-49). Appendiceal carcinoid tumors are tumors with a good prognosis. Even though their prognosis is good, close monitoring of the patients is recommended due to the likelihood of the detection of a proportion (13-33%) of synchronous and/or metachronous colorectal cancer in the literature.

Keywords: appendix, carcinoid tumor, pregnancy, treatment, pathology

INTRODUCTION

Malignant tumors of the appendix are rare, and the histopathological diagnosis can often be made in the postoperative period. Carcinoid tumor, the most common tumor of the appendix, may be detected in 0.5% of appendectomy specimens (1, 2). Carcinoid tumors also composed 80% of the masses found in the appendix (2). The patients who underwent appendectomy in emergency conditions with a preliminary diagnosis of acute appendicitis and whose histopathological examination revealed a carcinoid tumor, were evaluated retrospectively in a period of 7 years and the diagnosis and treatment approaches in carcinoid tumor of the appendix are reviewed with current literature in this paper.

MATERIAL AND METHOD

The clinical records of 1800 patients who underwent surgery in emergency conditions with a preliminary diagnosis of acute appendicitis at Department of General Surgery of Sakarya University Training and Research Hospital between January 2008 and January 2015 were evaluated retrospectively. The clinical records of the patients in whom «carcinoid tumor of appendix» was detected on the histopathological examination were investigated regarding demographic data, preoperative process (clinical and radiological findings, laboratory tests), operative findings, and follow-up results.

RESULTS

One thousand eight hundred patients underwent surgery in emergency conditions with a preliminary diagnosis of acute appendicitis in the specified 7-year period. While laparoscopic appendectomy was performed in three hundred eighty-six (21.3%) patients, open method was preferred in 1419 patients (78.6%). Carcinoid tumor was detected in 7 patients (0.38%) on histopathological examination. Of these patients, 4 were male (57.1%), 3 were female (42.9) and the mean age was found to be 36 (22-49).

Common presenting symptom was abdominal pain in all patients (100%), loss of appetite and nausea, vomiting accompanied the abdominal pain in three patients (50%). The mean time between the onset of the complaints and the hospital admission was 2(1-4) days. None of the patients had symptoms suggestive of carcinoid syndrome (including flushing, diarrhea, asthma, and cyanosis). Laboratory examination in 5 patients (66.6%) showed leukocytes (average 13000/mm3). While abdominal ultrasonography (USG) was performed in 3 patients (43%), abdominal computed tomography (CT) in 3 patients (43%) during the preoperative period at the time of diagnosis, 1 patient underwent surgery as a result of physical examination and laboratory investigations. One of the patients (14.2%) was operated under spinal anesthesia, 1 (14.2%) under epidural anesthesia, 5 (71%) under general anesthesia. Six (71%) patients underwent open appendectomy, 1 patient (14.2%) laparoscopic appendectomy. The classical Mc Burney incision was preferred in patients who underwent open appendectomy.

A perforated appendix was observed in none of the patients. All tumors were located in the tip of the appendix and the mean tumor diameter was 1.11 cm (0.3-2.5 cm). No patient had serosa, appendiceal mesentery or lymphatic invasion on detailed histopathological examination. While the tumor size was 2.5 cm in 1 patient (14.2%), 1.5 cm in 1 patient (14.2%), it was < 1 cm in others.

There was a pregnancy of 16 weeks in one patient among those with tumor diameter of<1cm and oophorectomy was added to the appendectomy procedure because a cystic mass of 4cm was detected in the right ovary during the operation. The pregnancy progressed uneventfully to term after the operation. Histopathological examination of the oophorectomy material demonstrated a mucinous cystadenoma.

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The decision of hemicolectomy was made in the patient with tumor size of 2.5 cm, but the patient refused to be operated.

The patients who had a trouble-free postoperative period were discharged on an average day 2 (1-4). The average 38 (9-74) -month follow-up of all patients including the patient with a tumor diameter of 2.5 cm continue to be disease-free.

DISCUSSION

Carcinoid tumors are classified in the group of neuroendocrine tumors (NET) and it is considered to be arisen primarily from enterochromaffin cells which are found in the submucosa of the gastrointestinal tract and main bronchi. Carcinoid tumors of Midgut which also involves appendix are different from foregut and hindgut equivalents by positive argentaffin staining histochemically with silver staining, containing a higher proportion of serotonin, causing metastasis and carcinoid syndrome more frequently, and leading to less bone metastases (3).

Despite the carcinoid tumors may be observed at any age, it is usually diagnosed in adult hood (average age of 29.8) and more common in females (2-4/1) (4-7). Although they are the most common malignancy of appendix, detection rate in the appendectomy materials is reported to be 0.3-0.9% (8). Even though the detection rate of the disease in our study is compatible with the literature, we thought that the differences in age and gender ratio with the literature are due to our small number of patients.

Carcinoid tumors of the appendix are tumors whose 80% generally may be smaller than 1 cm, 15% 1 to 2 cm and 5% greater than 2 cm (9). Given the localization at the appendix, 75% are located in the tip of the appendix, 20% in the middle, and 5% in the appendiceal root (10). While the diameters of the tumors in our study were found to be consistent with the literature data, the tumor was located in the tip of appendix in all of our patients.

Appendectomy continues to be 55% of the most commonly performed surgical interventions in emergency operations performed in the United States (11, 12). Nevertheless, since the carcinoid tumors are in the group of rare tumors, it may lead to confusion about the future approach for the patient as the histopathological examination after the operation resulted in the appendiceal carcinoid tumor. In general, appendiceal carcinoid tumors smaller than 1 cm are considered not to metastasize, and appendectomy is an adequate treatment (7, 13). There is a possibility of metastasis for tumors of 1 to 2 cm; however, it is usually contented with only appendectomy in this group of patients because of low rates (2%). The possibility of metastasis is up to about 20% for tumors greater than two cm (1, 14-16). From another perspective, more than 99% of metastatic carcinoid tumors of appendix are tumors of > 2cm. Therefore, traditionally more radical surgical interventions are recommended for tumors of >2cm (15, 16). Complementary radical surgical intervention, apart from the tumor diameter, is recommended in the presence of poor prognostic criteria such as the existence of positive surgical margin, involvement of appendiceal mesentery, involvement of cecum, a high mitotic index, and the elevation of Ki-67 (7). We performed a complementary radical surgical intervention in just one patient.

Carcinoid syndrome is a clinical picture caused by the release of the vasoactive substances including 5-hydroxytryptamine and histamine in to the systemic circulation and characterized by flushing, diarrhea, bronchospasm, and peripheral vasomotor symptoms. Although the carcinoid syndrome is one of the first conditions that come to the mind of all physicians when carcinoid tumor is mentioned, actually the incidence of this syndrome is quite low in carcinoid tumors (10%) (17), because the gastrointestinal system carcinoid tumors may cause this syndrome just when they metastasize to the liver (because released vasoactive substances escape the first–pass hepatic elimination) (9, 17, 18). Treatment options includes to reductive chemotherapy or pharmacological control of vasoactive substances in the presence of metastatic disease or carcinoid syndrome. Cytoreductive chemotherapy response rates are low (30-40%) (19). Octreotide, a somatostatin analog, is the most effective pharmacological agents for the control of symptoms associated with carcinoid syndrome. None of the patients had symptoms of carcinoid syndrome in our study.

The co-occurrence of appendiceal carcinoid tumor and the pregnancy is extremely and the literature is limited to case reports. Although the relationship between appendiceal carcinoid tumor and the pregnancy was not able to be demonstrated, it was reported that it may be accompanied by fetal malformations (10). It was shown that serotonin may cause growth retardation, fetal malformation and abortion in animal experiment performed in case of carcinoid syndrome which may possibly develop as a result of metastasis. Patients were recommended to be done an emergency appendectomy when the incidental carcinoid tumor was detected radiologically during pregnancy (20).

Appendiceal carcinoid tumors are tumors with a good prognosis and 5- year survival rates are reported to be 92%, 81% and 31% respectively in the presence of local disease, regional metastasis and distant metastasis (4). Even though their prognosis is good, close monitoring of the patients is recommended due to the likelihood of the detection of a proportion (13-33%) of synchronous and/or metachronous colorectal cancer in the literature. No local or systemic problem was seen during the 3 years of follow-up period, in our study.

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