A rare developmental anomaly associated with right colon cancer: appendix vermiformis agenesis

Mustafa Gok*, Erdogan Sozuer*///, Ugur Topal/**, Muhammet Akyuz*, Gamze K. Bozkurt*, Merve Hamurcu,* Kemal Deniz**

*Department of General Surgery, Erciyes University Medical Faculty, Melikgazi, Kayseri, Turkey
**Department of Pathology, Erciyes University Medical Faculty, Melikgazi, Kayseri, Turkey
***Department of Surgical Oncology, Erciyes University Medical Faculty, Melikgazi, Kayseri, Turkey

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Appendix vermiformis agenesis is quite rare. It is seen in 1/100,000 of patients undergoing laparotomy with an initial diagnosis of appendicitis. A 72-year-old woman who had not undergone any previous surgery was operated on for mechanical intestinal obstruction. Right hemicolectomy was performed due to obstructive tumoral mass in the hepatic flexure. There was no appendix vermiformis in exploration. Before deciding on the diagnosis of appendix agenesis, a thorough and rigorous exploration should be performed in ileocecal region and ascending colon. In our case, the diagnosis of appendix agenesis was incidental. However, it should be kept in mind that appendix agenesis may be present in patients undergoing surgery with a diagnosis of acute appendicitis.

KEY WORDS: Appendicular agenesis, Colorectal carcinoma, Congenital Abnormalities

Introduction

Appendix anomalies are very rare malformations. It usually occurs as an incidental finding in adult populations. Appendix agenesis was first described by Morgagni in 1718. Appendix vermiformis agenesis is quite rare. It is seen in 1/100,000 of patients undergoing laparotomy with an initial diagnosis of appendicitis. It was reported that appendix agenesis was present in 1/10,500 patients undergoing laparotomy and in 1/15,000 in autopsy series. In this paper, we aimed to present the appendix vermiformis agenesis found in a 72-year-old female patient undergoing surgery with a diagnosis of ileus and found to have obstruction due to colon cancer. We did not find appendix agenesis accompanied by colorectal carcinoma in the literature.

Case Report

A 72-year-old woman presented to the emergency department with nausea, vomiting and abdominal pain. Her medical history included Type 2 Diabetes Mellitus and essential hypertension. The patient had no previous history of abdominal surgery. Her physical examination showed blood pressure: 140/70 mmHg, respiratory rate: 24, pulse: 102/min, tythmic ritmik, and body temperature: 38.1°C. The general condition of the patient was mediocre, she was conscious, cooperative and orientated, and had abdominal distention and diffuse tenderness. In laboratory examination, the following results were obtained: Hgb: 13.6 g/dl, WBC: 13,9X10^3/µl, Platelets: 210x10^9/L and CRP:18 mg/L. Abdominal computed tomography showed thickening in the caecum and ascending colon wall and dilatation at its proximal (Fig. 1). Patient was admitted to intensive care...
care unit and intravenous fluid and antibiotherapy (piperacillin-tazobactam) were administered before the operation. Patient was operated in emergency conditions. After anesthetic clearance, emergency laparotomy was performed under general anesthesia. During exploration, there was a tumor extending from the colon to the hepatic flexure, causing tugging at the surrounding mesentery. Although there was no history of appendectomy, no appendix was observed. Extended right hemicolectomy and end ileostomy were performed in accordance with oncological procedures (Figs. 2, 3, 4).

There were no intraoperative complications. Postoperative follow-up was followed by administration of antibiotic therapy (piperacillin-tazobactam), intravenous fluid, analgesics (paracetamol), and anti-emetics and the oral intake was started after the gastrointestinal passage was achieved. The patient had no specific postoperative complications and wound complications did not develop. We did not need re-discovery/revision surgeries. We did not experience post-operative 30 day and long-term morbidity/mortality. The patient was followed up as an outpatient 10 days after discharge and they were well with no further complaints.

On pathological examination, moderately differentiated adenocarcinoma (2.9x2.5x1.2 cm, tumor invasion into subserosa, 13/13 reactive lymph nodes) was reported in the hepatic flexure. They were referred to medical oncology after discharge.
Discussion

Embryologically, the appendix arises from the caecum as an elongated tubular structure with a blind distal end, in the 10th week of gestation. Its base is constant in position and is 2-5 cm below the ileo-cecal valve on the posteromedial aspect of the cecum. The appendicular tip varies in position. Events that affect this process may lead to various positioning of the appendix and malformations. 4,5

Appendix vermiformis is usually found embedded in the lumen of the caecum. Appendix agenesis is thought to be the result of intrauterine vascular events, auto-amputation due to fibrous bands and appendicular atresia. 5,6

Intestinal malformations have previously been reported with appendix agenesis. 7 We did not detect intestinal malformations in our patient. History of previous abdominal surgery and appendectomy should be carefully investigated to avoid a wrong diagnosis. 2,4 In our patient, there was no history of previous surgery or intraabdominal infection that could lead to autoamputation.

In order to make this diagnosis in patients with previous abdominal surgery, the cecum and the ascending colon should be mobilized, the taenias should be followed up to the junction point. The entire ileocecal and retrocecal regions should be explored and the cecum should be carefully palpated to eliminate the possibility of intussusception of the appendix vermiformis into the caecum. 8

Appendix agenesis is a rare anomaly, and Collins classifies cecum and appendix anomalies into five classes (Table I). We classified our case as Type III. According to the Collins classification, the cecum was normal and the appendix vermiformis was absent.

Histopathological examinations should not be ignored due to the possibility that appendix vermiformis anomalies may be related to cancer. 9 Our case had accompanying colon cancer.

There are no specific symptoms and findings that may be used to diagnose appendix agenesis. Preoperative abdominal CT scanning may be helpful in some patients. A preoperative diagnosis could not be made in our case, agenesis of the appendix vermiformis was observed during surgical exploration.

In this case, we wanted to draw attention to the appendix agenesis because of the possibility of surgeons encountering a similar condition during surgery.

Table I - Collins classification

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>Type I</td>
<td>Absence of appendix and cecum</td>
</tr>
<tr>
<td>Type II</td>
<td>Rudimentary cecum and absence of appendix</td>
</tr>
<tr>
<td>Type III</td>
<td>The cecum was normal and the appendix vermiformis was absent</td>
</tr>
<tr>
<td>Type IV</td>
<td>Normal cecum and rudimentary appendix</td>
</tr>
<tr>
<td>Type V</td>
<td>Giant cecum without appendix</td>
</tr>
</tbody>
</table>

References

1. Morgagni G: *Adversaria anatomica all things (1-V1) of which the latter three are now for the first time will be betrayed*. Patavic J Cominus, 1719; 3:64.


Riassunto

La agenesia dell’appendice vermiforme è abbastanza rara, e si riscontra in 1 su 100.000 pazienti sottoposti a laparotomia con una diagnosi iniziale di appendicite. Una donna di 72 anni che non aveva subito alcun intervento chirurgico precedente è stata operata per ostruzione intestinale meccanica. L’emicolectomia destra è stata eseguita a causa della presenza di una massa tumoreale ostruttiva a livello della flessura epatica. All’esplorazione del pezzo operatorio non si è trovata l’appendice vermiforme.

Prima di concludere per una diagnosi di agenesia dell’appendice, deve essere eseguita un’esplorazione approfondita e rigorosa nella regione ileocecale e nel colon ascendente. Nel nostro caso, la diagnosi di agenesia dell’appendice è stata casuale. Tuttavia, va tenuto presente che l’agenesia dell’appendice può essere presente in pazienti sottoposti a chirurgia con diagnosi di appendicite acuta.


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