ANNALS OF GASTROENTEROLOGY 2004, 17(2):190-193

Original article

The incidence of primary appendiceal adenocarcinoma in Western Greece (prefecture of Aitoloakarnania): 13 years study of appendicectomy specimens

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SUMMARY

Aim of the study: **The primary adenocarcinoma of the appendix is a rare tumor. We describe three cases and register the tumor's annual incidence in our state.**

Methodology: **We studied 3596 appendicectomy specimens (1/1/1990 – 31/12/2002) from the two hospitals of the prefecture of Aitoloakarnania (Western Greece).**

Results: **We diagnosed 3 cases of the tumor (percentage 0.08%). The annual icidence in our state is 0.115 cases/ 100.000 persons/year (the estimated annual incidence for the total population of Greece is 11,5 cases/year). The mean age of our patients was 66.3 years. The preoperative diagnosis was acute appendicitis in two and caecum tumor in the other. In all, the definitive operation was a right hemicolectomy. Two patients had Dukes B tumors and the other, who died 15 days postoperatively, Dukes C. The two living patients are healthy without tumor reccurence, 15 and 60 months postoperatively.**

Conclusion: **The primary adenocarcinoma of the appendix: a. is very rare , b. affects both sexes in the fifth to sixth decade of their life, c. is diagnosed preoperatively with extreme difficulty, d. frequently gives ovarian metastases and coexists with other primary malignancy, and e. has better prognosis after right hemicolectomy. In our state (prefecture of Aitoloakarnania) we have registered very low rates of primary appendiceal adenocarcinoma (0.08% among appendicectomy specimens) and a very low annual incidence (0.115 cases/100.000 persons/year).**

Key words: primary appendiceal adenocarcinoma, appendicectomy, right hemicolectomy

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INTRODUCTION

The first report of primary adenocarcinoma of the vermiform appendix in the medical bibliography was by Berger in 1882[1]. Its rarity is manifested by the fact that approximately 500 cases only, have been reported to date. The incidence of the tumor in appendicectomy specimens is about 0.08%[2] and 0.2% among intestinal malignant tumors[1]. Most patients go to the operating theatre with symptoms of acute appendicitis and so, following the histologic confirmation, need to undergo more extensive preoperation.

In the present study we report the experience of the two hospitals of prefecture Aitoloakarnania-Western Greece, regarding the primary adenocarcinoma of appendix, during the last 13 years (1/1990 – 12/2002).

PATIENTS AND METHODS

During the last 13 years (1-1-1990 up to 31-12-2002) in the two hospitals of the prefecture of Aitoloakarnania, 3596 individuals underwent resection of the vermiform appendix. The preparations were studied by the routine method, which includes a longitudinal section of the tip and two transverse sections at the base and in the middle

of the appendix. In the case of detection of adenocarcinoma, repetitive sections were made, aiming at better identification of the histological type of the tumor and its stage, as well as estimation of possible malignant infiltration of the surgical margins.

RESULTS

From a total of 3596 appendicectomy specimens during the last 13 years, we diagnosed 3 cases of primary appendiceal adenocarcinoma (percentage 0.08 %) (Table 1). Based on official reports, the population of the prefecture of Aitoloakarnania that is served by the two hospitals is about 200.000 people, so the annual incidence of primary adenocarcinoma of the appendix, in our state, is 0.115cases/100000/year. In the total population of Greece (about 10.500.000 people) the annual incidence of the disease is estimated to be 11.5 cases/year.

Case 1: Female patient, 72 years old, underwent a right hemicolectomy (1995) due to a caecum tumor. Post-surgery microscopic examination revealed a primary appendiceal mucinous adenocarcinoma extending to the caecum. There was one lymph node with metastasis (Dukes C). The patient died 15 days after the operation from cardiac complication.

Case 2: Male patient 72 years old, underwent an urgent appendicectomy (1998) due to symptomatology of acute appendicitis (right lower quadrant pain, fever and leucocytosis). There was no suspicion of appendiceal cancer during the procedure. Microscopic evaluation revealed obstruction due to an intraluminaly growing adenocarcinoma (adenoma with malignant transformation), extending to serosa (Dukes B). The patient then underwent a right hemicolectomy (microscopic examination revealed the remaining tumor cells and 34 lymph nodes without metastases) (Dukes B). Sixty months after surgery there is no evidence of tumor reccurence.

Case 3: Female patient, 55 years old, with acute appendicitis symptomatology, underwent an urgent appendicectomy (2000). The preoperative laboratory examinations (blood test, radiographs and abdominal ultrasonography) was normal, except for mild leucocytosis. During the operation, an inflammed appendix was found without cancer suspicion, and we proceeded to a typical appendicectomy. The histologic examination revealed an intraluminal mucinous adenocarcinoma (extending to the muscularis propria, Dukes B), infiltrating the proximal surgical margin. The patient then underwent a right hemicolectomy (microscopic examination revealed the remaining tumor cells and 34 lymph nodes without metastases) (Dukes B). Fifteen months after the colectomy the patient is healthy, without evidence of tumor reccurence.

DISCUSSION

The incidence of primary adenocarcinoma of the vermiform appendix in the region of the prefecture Aitoloakarnania (Western Greece) during the last 13 years (1-1-1990 up to 31-12-2002) was 0.115 cases/100.000/year, while its frequency in the appendicectomy specimens was 0.08% (the estimated annual incidence for the total population of Greece is 11.5 cases/year). These percentages are among the lowest published worldwide.

Nielsen et al[3] reported an annual incidence of appendiceal adenocarcinoma almost twice ours (approximately 0.2 cases/100.000/year, Island 1974-1989). In the more extensive relative study of Collins et al[2], on 50.000 appendicectomy specimens, the reported incidence of primary adenocarcinoma was 0.08%. This

Table 1. Clinical details of patients with primary appendiceal adenocarcinoma

Case	Age	Sex	Preoperative diagnosis	Inflammation	Initial operation	Definitive treatment	Stage (Dukes)	Follow-up (months)
1	72	F	Caecum tumor	No	Right hemicolectomy		C	Died 15 days after the operation
2	72	M	Acute appendicitis	Yes	Right hemicolectomy	Right hemicolectomy	B	60 (without tumor recurrence)
3	55	F	Acute appendicitis	Yes	Right hemicolectomy	Right hemicolectomy	B	15 (without tumor recurrence)

percentage is the lowest reported prior to ours. On the other hand, greater incidences was reported by Rutlegde et al[4] in 3474 and Hananel et al[1] in 2520 appendectomies (incidences 0,2%), and by Gilhome et al[5] in 9380 and Burgess and Done[6] in 10.526 appendectomies (incidences 0,1%). The usual age of diagnosis is the fifth to sixth decade of life.

The preoperative diagnosis of the appendiceal adenocarcinoma is very difficult. Most cases are diagnosed during emergency appendicectomy for suspected appendicitis and the others are incidental findings during laparatomy performed for some other disease. In our study, two of the patients (cases 2 and 3), were operated on an emergency basis for suspected acute appendicitis. The other patient (case 1) underwent a right hemicolectomy due to a caecum – appendix tumor. In the study of Hananel et al[1], only one of the eight patients had a correct preorerative diagnosis, while the most common indication for laparatomy was acute appendicitis or periappendicular abscess. Nielsen et al[3] reported seven cases of adenocarcinoma of the appendix. Four patients presented with symptoms of acute appendicitis and in the other three cases the clinical presentation was that of peritoneal carcinomatosis of unknown origin. In the most extensive relevant study from Mayo Clinic[7], the authors could not make the correct preoperative diagnosis in any patient as well as in the study of Cortina et al[8].

As a result of the difficulty in preoperative diagnosis, in most cases the initial operation is limited to appendicectomy alone. In the two hospitals of the prefecture of Aitoloakarnania we follow the routine method (as described in the Patients and Methods section) for the microscopic evaluation of the appendicectomy specimens. This method increases the possibility of identification of such tumors, without significantly increasing the cost of the examination and pathologist's work time. Hananel et al[1] in their study, use a slight modification of this routine protocol, including one to three sections when the removed appendix looks normal. They consider that their relatively high rate of the adenocarcinomas of the appendix (0.2%), is due partly to the change in the routine pathological examination. Based on our results, we do not support this modification in the routine protocol, because the possibility of appendiceal adenocarcinoma identification is minimal and the only consequence is the probable increase in the cost and work time.

Staging of the primary adenocarcinoma of the appendix is based upon the same microscopic findings as in colon cancer. Mucinous adenocarcinoma is the principal histological type[1,4,9]. In our study two patients (cases 1 and 3) had mucinous tumors, Dukes C and B respectively, and the other (case 2), Dukes B colorectal-type tumor that developed from a tubular adenoma of the appendix. In Hananel's[1] study, seven from the eight patients had Dukes C and only one had a stage B tumor. Cortina et al[8], report that 75% percent of patients had metastatic disease at the initial operation, 15% had second primary malignancy while 38% of female patients had synchronous ovarian lesions. The authors suggest a complete work-up of the patient for a synchronous malignancy and possibly bilateral oophorectomy.

Undeniably, the second operation should be an oncological procedure, which is achieved via resection of the right colon along with the drainage lymph nodes. Two of our patients with primary appendiceal adenocarcinoma underwent a right hemicolectomy about one month after the appendicectomy. The proposal for right hemicolectomy has only supporters in the literature, whereas there are no controlled prospective studies that compare the procedure with the simple appendicectomy[1,8,9]. For lesions confined to the mucosa, some suggest that there is no survival advantage in right hemicolectomy over appendicectomy alone[1]. In our study two patients (cases 2 and 3), live without evidence of tumor recurrence, 60 and 15 months respectively after right hemicolectomy, while patients died postoperatively from cardiac complication. The long-term prognosis for primary appendiceal adenocarcinoma is proportionate to the tumor stage, as in colon adenocarcinoma. The mucinous histologic type associates with a better survival[9] while the prognosis is poor if the tumor is perforated[1].

In conclusion, the primary adenocarcinoma of the appendix: a. is a very rare tumor (usually mucinous adenocarcinoma), b. affects equally both sexes in the fifth to sixth decade of life, c. is diagnosed preoperatively with extreme difficulty, so most patients should undergo right hemicolectomy, shortly after the initial appendicectomy, d. frequently gives ovarian metastases and coexists with other primary malignancy, and e. has better prognosis after right hemicolectomy. Finally, in our state (prefecture of Aitoloakarnania – Western Greece) we have registered very low rates of primary appendiceal adenocarcinoma (0.08% among appendicectomy specimens) and very low annual incidence (0.115 cases/100.000 persons/year).

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