# Benigni in maligni tumorji pri 700 zaporednih apendektomijah

# Benign and malignant tumors in 700 consecutive appendectomies

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*Key words: appendix, appendicitis, appendiceal neoplasm, neuroendocrine tumour, mucinous tumour, GIST*

## POVZETEK

Vnetje slepiča (apendicitis) je najpogostejše akutno trebušno vnetje, ki potrebuje kirurško zdravljenje. Histološki pregled odstranjenega slepiča običajno potrdi klinični sum na akutno vnetno dogajanje, včasih pa pokaže tudi druge pomembne diagnoze. Namen raziskave je bil ugotoviti pojavnost in histološke značilnosti tumorjev slepiča v naši populaciji. V raziskavo smo vključili 728 slepičev, ki smo jih v obdobju sedmih let pregledali v naši ustanovi.

Primarni tumor smo ugotovili v 30 (4,1 %) primerih, kar je nekoliko večja pojavnost kot je opisano v literaturi. Med njimi je bilo osem nevroendokrinih tumorjev, pet klasičnih adenomov, 11 primerov mucinozne neoplazme nizke stopnje malignosti (v enem primeru v povezavi z psevdomiksomom peritoneja) in trije primeri karcinoma (po en mucinozni, intestinalni in nediferencirani). V enem primeru smo postavili tudi diagnozo multicističnega mezotelioma ter v slepiču redkega gastrointestinlnega stromalnega tumorja. Pri večini primerov je bila napotna diagnoza akutni apendicitis, le pri sedmih, pretežno velikih tumorjih je sum na tumorsko rast postavil že kirurg.

## ABSTRACT

Appendicitis is one of the most common abdominal inflammatory conditions requiring surgical intervention. Histology usually confirms the clinical suspicion of acute inflammation, but sometimes other clinically relevant diagnoses are made. The aim of our study was to analyse the appendectomies at our institution and to determine incidence and histological spectrum of appendiceal tumours in our population. We reviewed the histopathology results of 728 consecutive appendectomies received in a 7-year period.

Appendiceal tumours were found in 30 cases (4.1%), which is a higher rate than reported in the literature. There were eight endocrine tumours, all G1, five cases of classical adenomas, 11 cases of low-grade appendiceal mucinous neoplasms (one associated with pseudomyxoma peritonei) and three cases of carcinoma (one mucinous adenocarcinoma, one intestinal type, one undifferentiated carcinoma). We also diagnosed one case of multicystic mesothelioma and one gastrointestinal stromal tumour. Most patients presented with acute appendicitis, only in seven patients, mainly large tumours, the diagnosis was suspected already by surgeon.

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Histološki pregled odstranjenega slepiča je potrebno opraviti vedno, saj tudi v klinično nesumljivih primerih lahko odkrije številne klinično pomembne bolezni ter celo tumorje.

Histopathological examination of appendectomy specimens may reveal many different conditions not previously suspected; therefore, it should be performed in all cases.

## Introduction

Appendicitis is one of the most common abdominal conditions demanding surgical intervention. It affects about 10% of population and is thought to be initiated by progressive increases in intraluminal pressure that compromises venous outflow. In 50% to 80% of cases, acute appendicitis is associated with overt luminal obstruction, usually by lymphoid hyperplasia or fecalith (1). Less commonly, symptoms and signs result from other causes such as inflammation in the setting of Crohn's disease, endometriosis or even tumour. Primary appendiceal tumours are rare neoplasms accounting for 0.2–0.5 of all gastrointestinal tumours and < 2% of all appendectomies (1–7). Neoplastic processes in the appendix are histologically similar to their colonic counterparts. However, especially regarding epithelial neoplasm, the terminology is not standardised among pathologist and reading pathology reports can be a challenging task for the clinical doctors.

The aim of our study was to determine the incidence and histological spectrum of appendiceal tumours in our population.

## Materials and methods

The study included all appendices received for histological evaluation at the Institute of Pathology of the Medical Faculty at the University of Ljubljana in a seven year period. Appendectomies performed as an incidental procedure during some other operation were excluded.

The histology of the appendiceal tumours was evaluated again by experienced pathologists. Demographical data were collected from the patients' charts.

## Results

During the study period, 728 patients had an appendectomy for appendiceal pathology. Appendiceal neoplasm was diagnosed in 31 patients (4.3%). The largest single group of tumours were low grade appendiceal mucinous neoplasms (LAMN) comprising 35.5% of all tumours (Fig. 1A and 1B), followed by neuroendocrine tumours (25.8%) (Fig. 2). The incidence of different histological entities is presented in Table 1.

Most of the patients presented with acute appendicitis with or without abscess formation, some with chronic appendicitis. Only occasionally tumour was

Tabela 1. Histološke diagnoze tumorjev slepiča pri 728 zaporednih apendektomijah
*Table 1. Histological diagnoses of the appendiceal tumours in 728 consecutive appendectomies*

| Type of tumour | No. of cases (%) | |
|---|---|---|
| Adenoma | 5 (0.7%) | |
| Low-grade appendiceal mucinous neoplasm (LAMN) | 11 (1.5%) | 1 associated with pseudomyxoma peritonei |
| Adenocarcinoma | 3 (0.4%) | 1 intestinal type<br>1 mucinous<br>1 undifferentiated |
| Neuroendocrine tumour | 8 (1.1%) | all G1 according to WHO<br>2 with lymph node metastasis |
| GIST | 1 (0.1%) | |
| Multicystic mesothelioma | 1 (0.1%) | |
| Folicular lymphoma | 1 (0.1%) | |
| Metastasis (urothelial carcinoma) | 1 (0.1%) | |
| Total | 31 (4.2%) | |

Figure1. Low-grade appendiceal mucinous neoplasm (LAMN). Abundant mucin causing appendiceal distention (A).

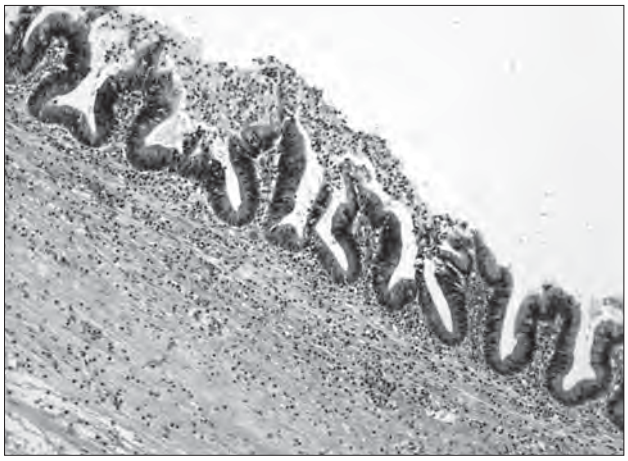

Figure1. Histology shows filiform villi lined by tall mucinous epithelium displaying low-grade cytologic atypia (B).

suspected already by surgeon: in one of eight neuroendocrine tumours (the largest one), two out of 11 LAMNs and in two out of three carcinomas.

In this series, the mean age of patients with neuroendocrine tumours was 35.4 years with a male-to-female ratio 1:7. The size of tumours ranged from

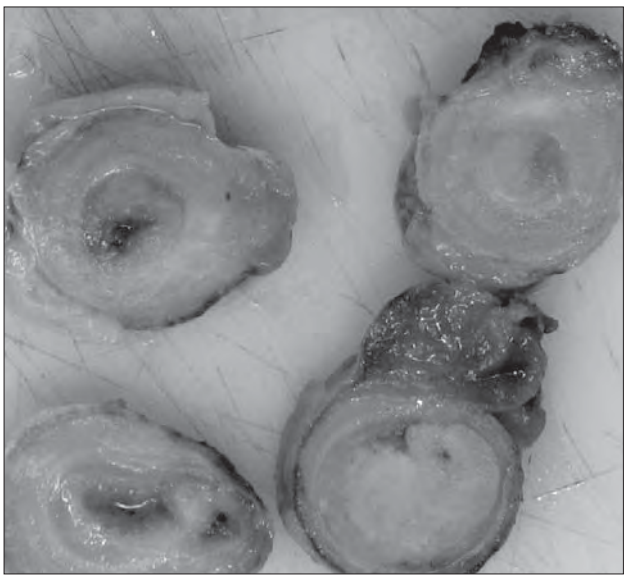

Figure 2. Neuroendocrine tumour of the appendix with a characteristic yellow colour.

0.35 to 3.5 cm (mean 1.42 cm), in five patients tumour was less than 1 cm in greatest diameter. In two cases, metastases in lymph nodes were detected.

The mean age of patients with colonic-type adenoma was 59.2 years. There were three tubular adenomas and, one villous and tubulo-villous adenoma each. None of them was associated with mucocele and none of the patients had familial adenomatous polyposis.

Among 11 patients with LAMN there were six women and five men, mean age of 58.4 years. In six cases herniation of mucin with or without neoplastic epithelium into the muscularis propria was present, and one of them was associated with pseudomyxoma peritonei.

GIST measuring 1.0 cm was diagnosed in a 11-year-old girl, multicystic mesothelioma was found in a 20-year-old male with clinical signs of acute appendicitis, but without histological signs of inflammation in the appendix.

## Discussion

Primary appendiceal neoplasms are uncommon, being found in approximately 0.5%–1.0% of appendectomy specimens at pathologic evaluation (1–3). In this study the incidence of primary appendiceal tumours was 4.3%. This rate is higher compared to

most similar studies (2–3, 7–8). Most of previous studies did not include benign and rare types of primary appendiceal tumours such as classical (colonic type) adenomas, lymphomas and tumours of mesothelial origin neither did they include metastatic processes. The number of examined appendices in our study increased from 37 in the first year of the study to 266 in the last year. Since the population covered by out pathology department remained the same during that period we believe that, in the past, not all appendices were sent for the histological evaluation, making the calculated incidence higher.

Although the incidence of appendiceal tumours in our study was higher, the distribution of histological entities was similar to previous studies with LAMNs being the most frequent and accounting for more than one third of cases, followed by neuroendocrine tumours representing about one quarter of appendiceal neoplasms (7). On the other hand, according to some authors. neuroendocrine tumours are the most common neoplasms of appendix (9–10). Interestingly, we have not diagnosed a single case of goblet cell carcinoid although we have encountered a few cases in the past. Neuroendocrine tumours tend to be diagnosed at an earlier age than other neoplasms and women predominate (1, 10–11), as was the case in our series.

Appendiceal neoplasm is typically associated with acute appendicitis and the diagnosis of tumour is usually difficult to make preoperatively. In our series, a diagnosis was suspected prior to histology report only in a few patients, all of them had large tumours or tumours associated with mucocele. On the other hand, benign processes associated with mucocele can sometimes be misinterpreted as tumours by surgeon.

One of three patients with appendiceal carcinoma (undifferentiated carcinoma) also had a moderately differentiated adenocarcinoma of the sigmoid colon. A higher rate of associated cancer in patients with appendiceal neoplasm, especially of synchronous colon carcinoma has previously been reported (6).

Gastrointestinal stromal tumors (GIST) are rare in the vermiform appendix. Less than 10 cases have been reported so far, all in patients over 50 years of age (12). Interestingly, our patient with appendiceal GIST was an 11-year-old girl.

In conclusion, appendiceal neoplasm is a rare gastrointestinal malignancy. Most of them are diagnosed only after histological examination of the resected specimen. Therefore the histological examination of the resected appendix is necessary for establishing the correct diagnosis and can influence further clinical decisions. Especially with appendiceal mucinous neoplasm it is important to remove appendix intact, trying to avoid trauma and possible rupture of the appendix, as this can affect long term outcome.

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