CASE REPORT

Primary Adenocarcinoma of the Appendix

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Summary
Primary adenocarcinoma of the appendix is exceedingly rare type of malignancy with the incidence of 0.4 cases per 100,000. It composes less than 0.5% of gastrointestinal malignant neoplasms. Frequently, the diagnosis is reached only after histological examination of surgically excised appendix due to suspected inflammation. Despite the low survival rate in case of primary appendix tumours aggressive therapy is necessary to obtain long-term survival. We present a well-documented case of instant neoplasm evaluated at our institution. To the best of our knowledge, no such detailed studies have been carried out before in Latvia.

Key words: adenocarcinoma, vermiform appendix, mucinous neoplasm.

AIM OF THE DEMONSTRATION
We present thoroughly documented case of primary adenocarcinoma of the appendix along with imaging studies in order to demonstrate an exceedingly rare case of malignant tumour.

CASE REPORT
Seventy nine-years-old female was admitted to the hospital due to dull pain in the right flank and febrile temperature. During her disease history, this was the third hospitalization within a year. Six month ago she was admitted to urology department with the same clinical symptoms and signs including blunt pain in the right side and fever. Right side lumbotomy with drainage was performed then due to retroperitoneal abscess. Two weeks later, re-operation was needed along with drainage.

During the third hospitalization the computed tomography (CT) scan revealed 9.5x3.0x3.0 cm large abscess cavity in the retroperitoneal space along with periappendicular infiltration. The laboratory investigations demonstrated haemoglobin level as low as 86 g/L and markedly increased C-reactive protein (CRP) level up to 269.9 mg/L. No other abnormalities were found by the laboratory tests. At first, relumbotomy was done and abscess cavity was opened. Bacteriological examination disclosed Enterococcus spp. sensitive to Ampicillin, Vancomycin and Linezolid. The relumbotomy wound healed secondarily. A week later appendix was surgically removed through lower median laparotomy access. During the operation it was observed that the appendix was placed close to retroperitoneal space, it had unusual high density on palpation and lacked gross signs of acute inflammation. Fistula tunnel was found at the end of the apex. The postoperative recovery was slack. The CT of the abdominal cavity and retroperitoneal space was repeated on the 7th postoperative day and showed no fluid collection. The laboratory tests demonstrated only slightly elevated CRP level up to 50.6 mg/L. Histological examination of the appendix revealed invasive high-grade mucinous adenocarcinoma (Figure 1). The tumour invaded through the serosa at the apex. The final estimate of the cancer spread was pT4G3R0. The appendicular mucosa was completely replaced by villous adenoma extending to the resection line and showing high-grade dysplasia. The presence of precursor lesion confirmed the true cancer origin in the appendix. Oncologic council concluded any chemotherapy or extended surgery is not necessary. The dynamic surveillance was required instead. No metastases were observed during the hospitalization and surveillance of 1 year. The colonoscopy and CT scan were performed year later and did not reveal any malignant tumour.

DISCUSSION
Primary appendicular neoplasm was the first described by Berger in 1882 (5). The primary carcinoma of the appendix is a rare malignancy accounting for 0.4-1.0% of all gastrointestinal tumours, while primary adenocarcinoma of the colon is widespread (13). The rarity of the instant neoplasm has also embarrassed the scientific studies (1,2). To meet the requirements of evidence-based research, in all cases the diagnosis of appendicular adenocarcinoma has been verified by the pathologists performing formal histology after the appendectomy (14). Mucinous adenocarcinoma is one of the three histological subtypes of the primary appendicular adenocarcinoma including also colonic type and signet ring cell cancers (13). This tumour arises in pre-existing adenomas as in our case (3). In contrast, the carcinoid (called also gastrointestinal neuroendocrine tumour) is the most common neoplasm of the appendix constituting 90% of all appendicular primary tumours.

Patients with appendicular malignancies usually present with the symptoms of acute appendicitis (9). There are no specific symptoms of the appendicular tumour (4). In accordance with the published studies, the appendicular neoplasm was established preoperatively in none of
cases (6,12). The sex distribution is usually equal (7). Appendectomy is appropriate for the small tumours (pT1), usually found incidentally, measuring less than 2 cm and lacking mesoappendiceal involvement (4). On the other hand, several clinical studies have demonstrated better survival rate after the right hemicolectomy in comparison with appendectomy (9). For primary appendicular tumours greater than 2 cm, involving the mesoappendix or base of the appendix, right hemicolectomy should be considered (5,11). Overall 5-year survival is 44% for appendicular mucinous tumours, 52% and 20% for the colonic subtype and for the signet ring cell subtype, respectively (10). The general prognosis is comparable to the colonic cancer (8).

In conclusion, primary adenocarcinoma of the appendix has diverse clinical manifestations. Surgeons should keep in mind the possibility of primary appendicular malignancy when managing patients with suspected acute appendicitis. To improve the outcome, right hemicolectomy should be recommended.

Conflict of interest: None

REFERENCES

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Fig. 1. The histologic structure of the appendicular adenocarcinoma. A, Invasive growth. Haematoxylin-eosin (HE), original magnification (OM) 100x. B, Marked nuclear atypia in the tumour cells. HE, OM 100x. C, Nuclear expression of CDX2 in the neoplastic cells. Immunoperoxidase, anti-CDX2, OM 100x. D, Villous adenoma replacing the appendicular mucosa. HE, OM 100x.