

Case Report

A neuroendocrine tumor in a macroscopically unchanged appendix

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ABSTRACT

Introduction: The intraoperative failure to find a macroscopically visible pathological condition within the appendix or other abnormalities of abdominal organs that could explain clinical symptoms in patients eligible for appendectomy does not prejudice the absence of underlying disease in an apparently healthy appendix.

Aim: The aim of the study is to highlight the problem of possible hidden pathological conditions in a macroscopically unchanged appendix in patients scheduled for appendectomy.

Case study: In this article, we present a case of female patient who was qualified to laparoscopic appendectomy due to symptoms of acute appendicitis. Although intra-operatively appendix did not show any pathological signs, histopathological examination revealed neuroendocrine tumor in the organ.

Results and discussion: This publication describes a case of the removal of a macroscopically unchanged appendix in a patient with clinical and laboratory signs of acute appendicitis. A neuroendocrine neoplasm was histopathologically revealed in the removed appendix. The correct eligibility for appendectomy can present great difficulties even for experienced surgeons. Failure to identify visible abnormalities in the appendix after ruling out other possible causes of clinical symptoms does not prejudice the absence of pathological conditions in this organ.

Conclusions: This clinical case shows that the removal of a morphologically normal appendix in a patient eligible for appendectomy, in the absence of other intra-abdominal pathological conditions explaining the clinical signs, is not necessarily a mistake.

1. INTRODUCTION

According to available data appendicitis has been reported one of the most common reasons of acute abdominal pain with lifetime risk of 8.6% in males and 6.7% in females.¹ However, accurate diagnosis of this disease is still a serious medical problem.^{2,3} For example it is assessed that more than 300,000 appendectomies are performed each year in the United States, and less than 10% result in removal of normal appendix.^{4–7}

Despite the progress that has been made in the diagnostic capabilities of acute abdominal diseases,⁸ it is still the case that patients are operated on in whom surgery proves to be unnecessary. The operating surgeon, often without the assistance of more experienced colleagues, is faced with a serious dilemma as to whether a macroscopically healthy appendix should be removed if other intraoperatively identifiable causes may cause clinical symptoms (prompting the surgeon to consider the patient eligible for surgery) have been ruled out.

The following case study does not provide a clear answer to this question but may help many operators facing such a challenge to make a decision.

2. AIM

This report presents the diagnosis of a macroscopically unchanged appendix that contained a neuroendocrine tumour in a patient who was eligible for appendectomy, but during the operation, no special pathology in the intraperitoneal organs was found.

3. CASE STUDY

This paper concerns the case of a 26-year-old female patient treated in the Department of General Surgery at the District Hospital in Biskupiec, Poland.

The patient was admitted for complaints of right lower abdominal pain lasting for 4 days. The pain was located in the same area from the beginning. It was accompanied by malaise and an elevated body temperature of 38.7°C. The patient denied vomiting, diarrhoea, and dysuric symptoms. A laboratory test ruled out pregnancy. Clinically, the patient's condition was good.

Physical examination revealed regular heart rate of 70 bpm, and clear, loud heart sounds. An audible alveolar murmur over the lungs on both sides of the chest in auscultation.

Abdomen was soft on palpation; preserved, quiet peristalsis. Tenderness on palpation was marked in the right lower abdomen. Doubts were raised about the presence of peritoneal signs. The patient reports moderate tenderness on deeper palpation, but no increased muscle tone, marked muscle defence or significant tenderness upon release of pressure from the described area was noted.

Due to the frequent occurrence of pathological conditions within the reproductive organs that might cause complaints at this site, the patient was referred for a gynaecological consultation. The examining gynaecologist found no abnormalities in the reproductive system.

Laboratory tests showed elevated inflammatory parameters: CRP 53.3 mg/L (against a hospital laboratory standard of up to 5 mg/L), with no elevation of the leukocytosis and procalcitonin values. In the abdominal US scan performed, no appendix could be found. The sonographer described the presence of a small amount of free fluid in the right iliac fossa.

Bearing in mind the serious complications that can be associated with delayed surgical treatment of acute appendicitis and the occasionally atypical position of the appendix that can affect symptomatology, the patient was considered eligible for an on-call laparoscopic appendectomy.

The decision to perform laparoscopy was prompted by the patient's young age, the better cosmetic effect of the post-operative scar, the usually smaller number of peritoneal adhesions formed in the abdominal cavity following the procedure, and the possibility for a thorough evaluation of the entire abdominal cavity if no pathological conditions have been found within the appendix.

No pathological condition was found intra-operatively within the appendix that was of an average length, not swollen, with no inflammatory infiltration or nodular lesions, with a normal diameter. The small intestine was also examined, with no Meckel's diverticulum found. What drew attention, however, was the redness and slight swelling of a significant part of the small intestine. A slightly cloudy, yellow and green fluid was present in the pelvis minor, with the image being inconsistent with the content typical of purulent abdominal inflammation. The fluid was aspirated. Material for culture was collected. The reproductive organ was macroscopically normal. Accessible sections of the large bowel and the upper abdominal area were also examined, with no pathological lesions found.

Considering the patient's age, the adverse effect of any lower abdominal surgery on fertility, the possibility of the development of appendicitis in the future, and the suspicion of acute appendicitis frequently raised by doctors of other specialisations, a decision was made to remove the macroscopically unchanged appendix. The specimen was sent for histopathological examination.

Cefuroxime and metronidazole, antibiotics frequently used at the Department in similar cases, were empirically included in the treatment. The patient's postoperative course was uncomplicated.

The patient was discharged home with normal healing wounds on the 2nd day after surgery.

The result of the peritoneal fluid culture was only obtained after the patient was discharged home. Cefuroxime-sensitive *Escherichia coli* was cultured from the specimen.

The histopathological specimen showed a well differentiated neuroendocrine tumour NET G1 with a diameter of less than 1 cm, confined to the appendicular wall, with extensive

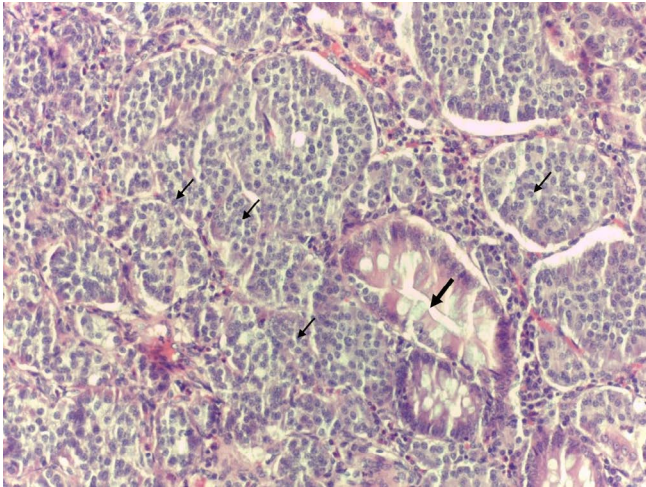


Figure. Histopathological examination of the neuroendocrine tumor. Small arrows indicate tumor cells while large arrow shows intestinal crypt (HE staining; magnification 200×).

infiltration of the muscle layer and focal penetration into the periappendiceal fat tissue without vascular invasion (Figure).

The following parameters were determined: panCK⁺, Ki67 < 2%, CD56⁺, Chromogranin⁺, Synaptophysin[±].

Once the result was received and communicated to the patient, she was referred for further follow-up at the regional oncology centre.

4. RESULTS AND DISCUSSION

Having retrospectively analysed the above case, it can be concluded that the clinical signs presented by the patient may have been related to the presence of a neuroendocrine tumour.⁹ Non-specific abdominal pain, moderately elevated inflammatory parameters with a rather long duration of complaints, good general condition, and the absence of symptoms typical of local or diffuse peritonitis may be due to the release of biologically active substances, e.g. serotonin, by the tumour.^{10,11} Since its concentration is not routinely determined as part of standard hospital diagnostics, it was not possible to determine whether it was responsible for causing the above symptoms in this case.

An additional conundrum is the fact of obtaining a positive result of the culture of the fluid collected intra-operatively from the peritoneal cavity.

The circumstance that the appendix was removed along with the tumour was a lucky coincidence.

To summarise the above case, the decision taken intuitively by the operator, contrary to the suggestions of van Dalen et al.¹² to remove a normal-looking appendix enabled an early diagnosis of a potentially fatal disease being fully treatable at an early stage.¹³ This may have helped the patient avoid the need to receive chemotherapy or undergo a major mutilating surgery.

Similar cases can occur at any facility regardless of the size and geographical location.¹⁴ The above study, being a

description of a single clinical case, must not be used as a basis for producing valid recommendations. It can, however, help the surgeon make a difficult decision to remove a normal appendix after ruling out other intra-operatively identifiable intra-abdominal pathological conditions explaining the clinical symptoms.

6. CONCLUSIONS

- (1) The lack of visible macroscopic signs of disease in the appendix does not prejudice it is free of pathology.
- (2) Removing an unchanged appendix associated with abdominal clinical symptoms, and intraoperatively excluded other abdominal pathology, does not have to be a mistake.
- (3) Diagnosing acute appendicitis and differentiating it from other acute abdominal diseases still remains a significant clinical problem.

Conflict of interest

The author declares that he has no conflict of interest.

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