CASE REPORT

APPENDICEAL NEUROENDOCRINE TUMOR IN A PEDIATRIC PATIENT – A CASE REPORT

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Abstract

Appendiceal neuroendocrine tumors (NETs) are the most common appendiceal neoplasms but remain rare in the pediatric population. They often present with clinical features mimicking acute appendicitis and are therefore usually diagnosed incidentally on histopathological examination after appendectomy. Prompt identification is crucial, as tumor characteristics determine management and prognosis. We report a case of a 13-year-old girl presenting with clinical and radiological signs of acute appendicitis due to fecalith obstruction and a right ovary cyst. Laparoscopic appendectomy and ovarian cystectomy were performed without intraoperative suspicion of malignancy. Histopathological analysis revealed a well-differentiated neuroendocrine tumor measuring 1.5 cm localized near the tip of the appendix. The lesion infiltrated all the layers of the appendix wall and penetrated the serosa, without lymphovascular invasion or mesoappendiceal extension. Immunohistochemical staining confirmed the diagnosis. Multidisciplinary tumor board decision guided further management and the patient remains disease-free at twelve-month follow-up. Outcome was promising with no recurrence, metastasis, complications, or need for additional therapy observed during follow-up. The discussion section emphasizes the critical role of histopathological analysis of appendectomy specimens and gives further recommendations for efficient management of appendiceal NETs in children, depending on tumor size and histological risk features. Appendectomy alone is usually sufficient for tumors less than 2 cm without invasion. This case underscores the critical role of histopathological vigilance and multidisciplinary care in pediatric surgical oncology.

ПРИКАЗ НА СЛУЧАЈ

НЕВРОЕНДОКРИН ТУМОР НА АПЕНДИКСОТ КАЈ ПЕДИЈАТРИСКИ ПАЦИЕНТ - ПРИКАЗ НА СЛУЧАЈ

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Клучни зборови: невроендокрин тумор на апендикс, детска хирургија, лапароскопска апендектомија, карциноиден тумор, педијатриска онкологија.

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Извадок

Невроендокрините тумори (НЕТ) претставуваат најчести неоплазми на апендиксот, но исклучително ретко се јавуваат во детска возраст. Се манифестираат со клиничка слика за акутен апендицит и најчесто се откриваат случајно при хистопатолошка анализа на отстранетиот апендикс. Навременото поставување на дијагнозата е од суштинско значење, бидејќи морфолошките и хистолошките карактеристики на туморот ги одредуваат понатамошниот третман и долгорочната прогноза. Во нашиот труд прикажуваме случај на 13-годишно девојче примено на нашата клиника со позитивни клинички и радиолошки знаци за акутен апендицит, предизвикан од опструкција со копролит, и придружна циста на десниот јајник. Беше направена лапароскопска апендектомија и цистектомија на јајникот, при тоа интраоперативно не се постави сомнение за постоење на малигна болест. Хистопатолошката анализа откри постоење на добро диференциран невроендокрин тумор со димензија од 1,5 см, локализиран во предел на дисталниот дел (врвот) на апендиксот. Туморот ги инфилтрира сите слоеви на апендикуларниот ѕид со зафаќање на серозата, но без присуство на лимфоваскуларна инвазија или екстензија во мезоапендиксот. Дијагнозата беше потврдена со имунохистохемиска анализа. Понатамошното лекување беше дефинирано врз основа на одлука на мултидисциплинарен онколошки конзилиум. По дванаесетмесечно следење, пациентката е без знаци за активна болест, рецидиви, метастатска дисеминација, постоперативни компликации или потреба од дополнителен третман. Во дискусијата се нагласува значењето на задолжителната хистопатолошка анализа на сите апендектомиски препарати; исто така се даваат и препораки за оптимален хируршки третман на невроендокрините тумори на апендикс кај деца, и тоа во зависност од големината на туморот и од присуството на хистолошки фактори на ризик. Кај тумори помали од 2 см, без високоризични карактеристики, апендектомијата вообичаено е доволна како единствен третман. Овој случај ја истакнува важноста на хистопатолошката анализа и мултидисциплинарниот пристап во третманот на невроендокрините тумори на апендиксот во педијатриската онколошка хирургија.

Introduction

Acute appendicitis remains one of the most common indications for emergency abdominal surgery in children[1]. While the overwhelming majority of appendectomy specimens confirm acute inflammation, a small percentage reveal unexpected findings, including neoplastic conditions2. Among these, appendiceal neuroendocrine tumors (NETs), previously referred to as carcinoid tumors, are the most frequently encountered neoplasms of the appendix[3,4].

Despite their relative frequency among appendiceal tumors, NETs remain rare in children, with an estimated incidence between 0.1% and 0.5% of pediatric appendectomies[2,4]. These tumors typically arise from enterochromaffin cells within the distal third of the appendix and are often small, slow-growing and asymptomatic[5].

Most pediatric cases present with symptoms indistinguishable from uncomplicated appendicitis, making preoperative diagnosis virtually impossible[6,7]. Imaging studies rarely detect these tumors, particularly if small and intraoperative findings are often non-suspicious2,6. As such, most NETs are identified only through postoperative histopathological examination. Because these tumors are rare in children, treatment recommendations are often based on adult data, highlighting the value of pediatric-specific case reports and registries[3,8]. This report presents a case of a 13-year-old girl whose appendiceal NET was discovered incidentally following appendectomy for suspected acute appendicitis. We discuss the clinical features, histopathology, surgical management, prognosis and the broader implications for pediatric surgical practice.

Case report

A 13-year-old girl presented to the emergency department of our clinic, complaining of right-sided lower abdominal pain that had started one day before, without associated nausea or vomiting. She was afebrile and her vitals were within normal limits. Abdominal examination revealed tenderness in the right iliac fossa. The pain was constant and progressing. Abdominal ultrasound showed a right ovarian cyst and small amount of free fluid in the pouch of Douglas. The patient was admitted to the hospital for further evaluation. Blood analysis were in normal range, except for an elevated C-reactive protein level (30.9 mg/L). Due to the non-conclusive ultrasound findings, a contrast-enhanced abdominal and pelvic computerized tomography (CT)-scan was indicated. The CT scan showed fecalith obstruction of the appendix and a right ovarian cyst. A clinical diagnosis of acute appendicitis and right ovarian cyst was made and she underwent an explorative laparoscopy, which revealed an inflamed appendix and a right ovarian cyst, with no other abnormalities and without any intraoperative suspicion of malignancy (Fig. 1). Laparoscopic appendectomy with right ovarian cystectomy was made. The resected specimens were sent for histopathological examination. Routine intravenous fluids, antibiotics and analgesics were given in the postoperative period. The patient was discharged from hospital

on postoperative day 3 with regular follow-up over the next two weeks.

The resected appendectomy specimen measured 6.5 cm in length. The cut surface revealed a yellow tumorous lesion of 1.5 cm size in its widest diameter and localized in the distal portion near the tip of the appendix (Fig. 2). The lesion infiltrated all the layers of the appendix wall and penetrated the serosa. Microscopy showed small sized round and oval tumor cells with a moderate amount of finely granular, eosinophilic cytoplasm and round nuclei with "salt and pepper" chromatin, arranged in nests. Mitotic rate was 0–1 per 10 high-power fields. A mild perilesional lymphocytic infiltrate was also described. The regional lymph nodes could not be assessed, as they were not submitted. Immunohistochemical staining was positive for neuroendocrine markers, including chromogranin, synaptophysin and neuron-specific enolase (NSE), with a Ki-67 proliferative index of < 2%. Based on these findings, a diagnosis of a well-differentiated Grade 1 neuroendocrine tumor of the appendix was reached. Follow up examination including PET scanning revealed normal results, with no regional or distant metastatic deposits detected.

The case was reviewed by a multidisciplinary tumor board, and given the favorable features, no further surgical intervention was recommended. The patient is currently undergoing monthly follow-up at the pediatric oncology department. At 12-month follow-up, the patient remains asymptomatic with no evidence of disease recurrence.

Figures

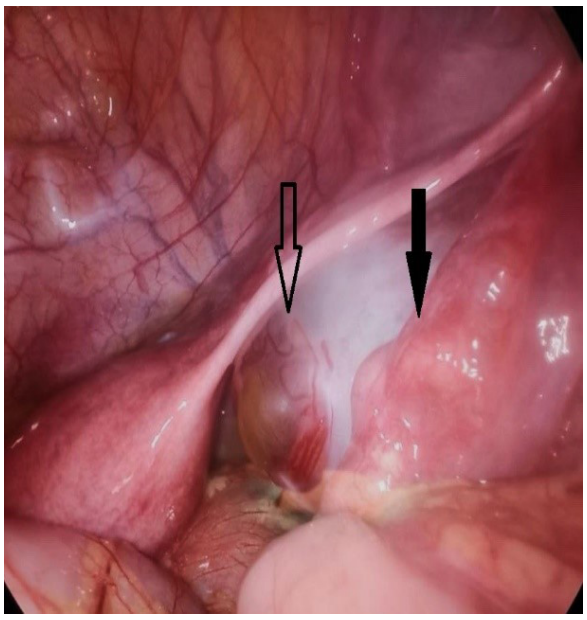

Figure 1:
Cyst of the right ovarium (hollow arrow); Inflamed vermiform appendix (solid arrow)

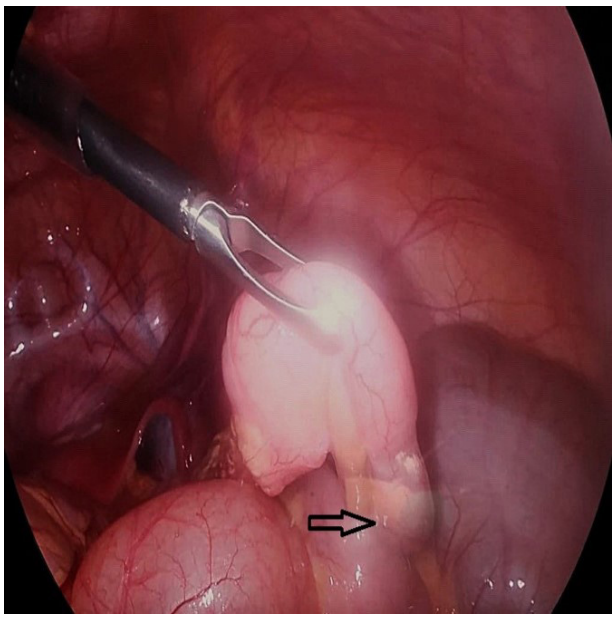

Figure 2:
Neuroendocrine tumor at the tip of the vermiform appendix (hollow arrow)

Discussion

Appendiceal NETs account for nearly 80% of all appendiceal tumors but occur infrequently in children[3,4]. Their estimated incidence in pediatric appendectomy specimens ranges from 0.1% to 0.5%, with a slight female predominance and a peak incidence in early adolescence[4]. Unlike many pediatric malignancies, there are no known genetic or hereditary syndromes commonly associated with appendiceal NETs in children. The tumors are usually sporadic and risk factors remain undefined[9]. Pediatric-specific data remain limited and treatment recommendations often reflect adult protocols[3].

Surgical excision remains the primary and definitive treatment for appendiceal neuroendocrine tumors (NETs) in pediatric patients[1,5]. Because these tumors are most often discovered incidentally during appendectomy performed for suspected acute appendicitis, as in our case, the initial surgical approach depends on the acute clinical presentation rather than oncologic suspicion[2,6]. Laparoscopic appendectomy is the preferred method in children, offering minimal invasiveness, reduced postoperative pain, shorter stay in the hospital and faster recovery. Improved visualization and the routine use of specimen retrieval bags contribute to safe tumor removal, even when malignancy is not suspected intraoperatively[1].

Intraoperative identification of appendiceal NETs is uncommon; however, features such as focal thickening, firm nodules at the appendiceal tip, or regional lymphadenopathy may raise suspicion. When available, intraoperative frozen section analysis can assist in immediate surgical decision-making, although definitive management is typically guided by postoperative histopathological findings[5]. Current recommendations stratify treatment according to tumor size and histologic risk factors. Appendectomy alone is considered curative for tumors smaller than 1 cm. For tumors measuring 1–2 cm, additional surgery may be considered in the presence of high-risk features, including mesoappendiceal invasion greater than 3 mm, lymphovascular invasion, elevated Ki-67 index, or positive surgical margins. Tumors larger than 2 cm generally warrant right hemicolectomy due to an increased risk of lymph node and distant metastasis[3,5,8]. In the present case, despite a tumor size of 1.5 cm, the absence of unfavorable histopathological and immunohistochemical features supported appendectomy as the sole definitive treatment.

Similar pediatric cases have been published recently, including a 16-year-old adolescent who presented with signs of non-typical appendicitis and was later found to have an appendiceal NET; a 14-year-old male with a well differentiated NET who required subsequent hemicolectomy after staging; and a 13-year-old female with an inflamed but not perforated appendix and a NET of 7 mm size located at its tip[2,4,7].

When indicated, right hemicolectomy involves resection of the terminal ileum, cecum, ascending colon and associated lymphatic drainage. In pediatric patients, the surgical approach, laparoscopic or open, is individualized based on tumor characteristics and surgical expertise,

with the aim of balancing oncologic clearance while minimizing long-term impact on overall growth and gastrointestinal function[1,3,5].

Postoperative surveillance is tailored to tumor risk profile and typically includes regular clinical evaluation, abdominal imaging and selective biomarker monitoring. Follow-up intervals generally range from every 6 to 12 months, particularly in intermediate risk cases and may extend for at least five years[10]. Prognosis for pediatric appendiceal NETs is excellent when management guidelines are followed, with recurrence being exceedingly rare[7,9]. Despite favorable outcomes, the psychological burden of an unexpected tumor diagnosis in a child should not be underestimated. Families may experience significant anxiety even in the setting of low-grade disease. A multidisciplinary approach involving pediatric surgeons, oncologists, radiologists and psychosocial support services is therefore essential to ensure comprehensive care[9,10]. This case highlights how a routine appendectomy can uncover a clinically silent malignancy and underscores the importance of meticulous histopathological examination and coordinated, patient-centered management to optimize both medical and emotional outcomes.

Conclusion

Appendiceal neuroendocrine tumors in pediatric patients, although rare, are clinically significant and are frequently discovered incidentally. They often mimic acute appendicitis, making preoperative diagnosis unlikely. This case reinforces the necessity of routine histopathological evaluation of all appendectomy specimens. Appendectomy alone is curative in most children with tumors smaller than 2 cm and without high-risk features. Surgical management should be guided by tumor pathology and aim to balance oncologic safety with preservation of quality of life. With multidisciplinary care, pediatric patients can achieve excellent long-term outcomes.

This case of a young patient reminds us that rare but significant conditions may lie quietly beneath common presentations, reinforcing the need for sustained clinical vigilance. It is our clinical attentiveness, precision and compassion that ensure these silent sentinels are not missed and that each child receives care as unique and resilient as they are.

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