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Presentation of a Neuroendocrine Tumor in a Child with Perforated Appendicitis: A Case Report

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ABSTRACT

Neuroendocrine tumors (NETs) are rare neoplasms that arise from neuroendocrine cells, with a significantly lower incidence in children compared to adults. In the pediatric population, appendiceal NETs are particularly notable, accounting for approximately 87.5% of cases, while the overall incidence of NETs in children is estimated at about 1.14 to 5.4 cases per million, compared to around 5.25 per 100,000 in adults. This illustrates the rarity of NETs in children and the difficulties in diagnosing them. These tumors often display symptoms similar to acute appendicitis, leading to their accidental detection during appendectomy. The vague clinical features of appendiceal NETs make preoperative diagnosis and management challenging. In children, these tumors are usually well-differentiated and have a positive outlook; however, the absence of specific pediatric guidelines has led to an excessive reliance on adult treatment protocols, which may be overly aggressive.

This case study recounts the experience of a 10-year-old girl who exhibited symptoms suggestive of acute perforating appendicitis. Surgical intervention revealed a neuroendocrine tumor within the mid-portion of the appendix lumen, highlighting the crucial role of histopathological examination in such instances. The findings underscore the need for increased awareness among clinicians about the potential for neuroendocrine tumors to imitate common surgical conditions like appendicitis. Additionally, this case emphasizes the importance of developing pediatric-specific management guidelines to enhance care and minimize unnecessary surgical interventions for this distinct patient group.

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Introduction

Neuroendocrine tumors (NETs) are more prevalent in adults, affecting around 5.25 individuals per 100,000, mainly impacting the lungs and gastrointestinal tract. In contrast, the occurrence of NETs in children is much lower, ranging from approximately 1.14 to 5.4 cases per million, with 87.5% of instances observed in the appendix, compared to just 3% in adults [1, 2]. The median age of the patients was 14 years, with an interquartile range of 12 to 16 years, and there was a predominance of females, accounting for 64.5% of the cases. In the majority of children, the tumor was found at the tip of the appendix (58.1%), and most tumors had a diameter of less than 1 cm (67.7%) [3]. This rarity presents unique diagnostic challenges, as these tumors often mimic the symptoms of acute appendicitis, leading to incidental discovery during surgical procedures [4, 5]. Although pediatric appendiceal NETs are typically well-differentiated and associated with a favorable prognosis, the lack of specific management guidelines can result in an over-reliance on adult treatment protocols, which may be unnecessarily aggressive [6]. This case report discusses a 10-year-old girl who presented with symptoms consistent with acute perforating appendicitis and was found to have a neuroendocrine tumor within the appendix post-surgery.

Case presentation

A 10-year-old girl presented at our hospital with severe abdominal pain, fever, and vomiting. Upon examination of her abdomen, there was generalized tenderness and guarding, indicating generalized peritonitis. Her laboratory results showed an increased white blood cell count and elevated C-reactive protein. The diagnosis was acute perforating appendicitis, and she was promptly taken to the operating room for an appendectomy. During the procedure, a perforated appendix was removed and sent for further examination. The gross pathology examination revealed a tan-necrotic appendix with a creamy-yellowish lesion in the mid-portion of the lumen, measuring 1 × 1 × 0.7 cm, along with pus and fecaloid material filling the rest of the lumen and the tip of the appendix (Figure 1). Microscopic analysis showed the presence of a neoplasm composed of neuroendocrine-like cells infiltrating the appendiceal wall. The neoplastic cells strongly expressed synaptophysin, with a mitotic rate of about 1/HPF and a Ki-67 activity index of 19% in hot spots (Figure 2). Additionally, there was neutrophilic infiltration in the muscularis propria and surrounding the appendix, along with moderate to severe fibrinoleukocytic exudate. The final diagnosis was a neuroendocrine tumor, grade 2, with perforated appendicitis and periappendicitis.

Discussion

The case of the 10-year-old girl with a neuroendocrine tumor (NET) highlights the essential role of histopathological examination. In this particular case, the tumor was situated in the mid-lumen area, which is less frequently observed than the typical location for such tumors (tip of the appendix). This situation reinforces the need for careful and thorough assessments to ensure that similar cases are not missed. Accurate diagnosis is critical for differentiating these tumors from more common conditions like appendicitis. Pediatric NETs, while rare, can present similarly to acute surgical conditions, making precise diagnosis essential for effective management.

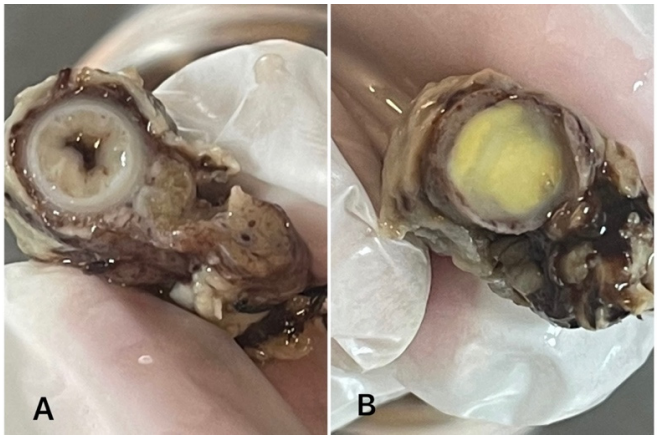

Fig. 1. Gross pathology of the appendix:
A: lumen of the appendix filled by pus, B: creamy-yellowish lesion in mid portion of the appendix

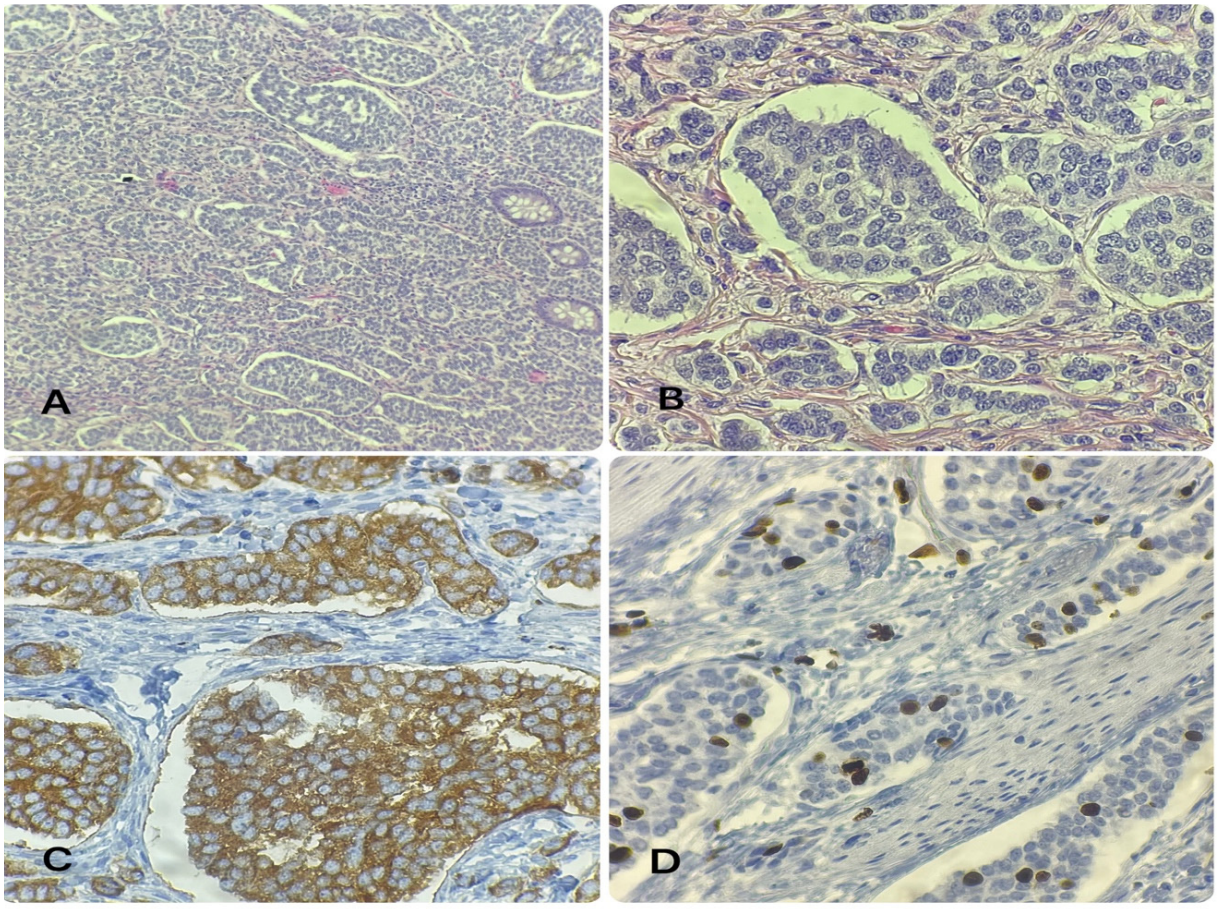

Fig. 2. Pathology slides of Neuroendocrine tumor in mid portion of the lumen of the appendix:
A,B: H&E-x4 and x40, C: Synaptophysin (positive), D: Ki67 (about 19% in hotspots)

Histopathological examination plays a pivotal role in the diagnosis of NETs. Differentiation between NETs and other conditions, such as appendicitis, is crucial, as misdiagnosis can lead to inappropriate surgical interventions. Studies indicate that early detection and accurate classification of NETs can significantly improve outcomes, with surgical resection often being curative for early-stage tumors. The histological grading of NETs, based on the 2017 WHO classification, allows for tailored treatment approaches that consider the tumor's biological behavior and histological characteristics [2, 7].

The management of pediatric NETs should reflect their unique characteristics. Current guidelines emphasize a multidisciplinary approach that includes surgery, medical therapy, and surveillance. For instance, peptide receptor radionuclide therapy (PRRT) has shown efficacy in managing advanced NETs, while somatostatin analogs are used to control hormone-related symptoms [8].

As awareness of pediatric NETs grows, there is a pressing need for specific clinical guidelines tailored to this demographic. The North American Neuroendocrine Tumor Society (NANETS) has been instrumental in developing consensus guidelines that address the management of various types of NETs. These guidelines are based on rigorous literature reviews and expert consensus, focusing on improving outcomes through standardized care protocols. Developing specific guidelines for pediatric NETs is essential to optimize patient care, reduce unnecessary surgeries, and improve overall outcomes [9]. Implementing such guidelines will enhance diagnostic accuracy and ensure that treatment strategies are aligned with the latest evidence-based practices.

Conclusion

The case of the 10-year-old girl underscores the necessity for clinicians to remain vigilant in recognizing neuroendocrine tumors as potential acute surgical conditions in children. As research continues and awareness increases, the development of tailored management strategies and specific guidelines will be crucial in optimizing care for pediatric patients diagnosed with these rare tumors. Enhanced

education and training for healthcare providers on the presentation and management of pediatric NETs will further contribute to improved patient outcomes.

Ethical Considerations

Ethics approval and consent to participate

This study was conducted in accordance with the ethical principles outlined in the Declaration of Helsinki. Ethical approval was obtained from the Ethics Committee of Tehran University of Medical Sciences.

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Conflict of Interests

The authors declare that they have no known competing financial interests or personal relationships that could have influenced the publication of this paper.

Authors' contributions

Aysan Nozheh: Conceptualization, Methodology, Project administration, Writing - original draft, Supervision

Seyed Alireza Hosseini: Investigation, Validation, Writing - review & editing, Resources

Maryam Sotoudeh Anvari: Writing - review & editing, Writing - original draft, Data curation, Software, Formal analysis

Competing interests

We declare that we have no known competing financial interests or personal relationships that could have influenced the work reported in this paper.

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