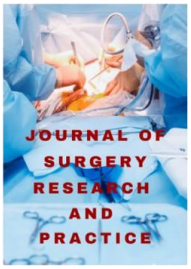

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Case Report

Clinical Approach to Incidentally Identified Appendiceal Neuroendocrine Neoplasms After Appendicectomy in Children

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Abstract

Neuroendocrine Tumours (NET) are the commonest type of tumours affecting the appendix. The majority of cases are diagnosed incidentally on histological examination of the resected appendix after Appendicectomy. There is debate about the optimal surgical management for localized appendiceal NETs that are impacted by many factors. The data to guide therapy in pediatric cases are limited due to the paucity of these tumors.

We present the case of 10 years-old boy with abdominal pain, fever and vomiting appeared for 2 days. On physical examination, a tenderness in the right lower quadrant of the abdomen. Laboratory tests showed an inflammatory syndrome. Ultrasonography revealed findings with acute appendicitis.

The patient underwent an uneventful appendectomy for an acutely phlegmoneous appendix. Histological examination showed a 0.3 cm well-differentiated neuroendocrine tumor grade 1 of the distal appendix with clean resection margins. The follow-up was uneventful and no further procedures were performed.

Appendicular NET are rare and usually asymptomatic and diagnosed incidentally on histological examination after appendectomy. Pediatric appendiceal NET tends to have a benign clinical course with excellent prognosis. Simple appendectomy is adequate treatment for NET less than one cm in diameter.

Keywords: Pediatric Surgery Department; Habib Thameur Hospital; Tunis; Tunisia; Neuroendocrine Tumor; Appendix; Children; Histology; Guideline

Introduction

Neuroendocrine Tumors (NET) are neoplasms that originates from the neuroendocrine cells that have properties of both neuronal and endocrine cells. NETs are most common in the small intestine, rectum and the appendix and frequently termed carcinoid [1]. NETs are considered rare tumor and the current incidence is reported to be about 2.8 cases per million in the pediatric age group [2]. Historically in 1907, they were considered benign and initially termed "little carcinomas" by Ordfer, specifically he termed them "karzinoide" and they were not considered a true neoplasm. The term carcinoid is often used for these NETs that are in the GI tract [3]. They have now considered true neoplasms and received the term gastroenteropancreatic NETs because of their immense heterogeneity [4]. The majority of cases are diagnosed incidentally on histological examination of the resected appendix. There is debate about the optimal surgical management for localized appendiceal NETs that are impacted by many factors including the tumor size, the extent of mesoappendiceal spread, lymphovascular invasion and perineural involvement. In addition, the data to guide therapy in metastatic disease are limited due to the paucity of these tumors.

Case Report

A 10 years-old boy presented to the emergency department with a chief complaint of abdominal pain localised in the right lower quadrant. He felt nauseous and had chills and fever appeared for 2 days. Physical examination revealed tenderness in the right

lower quadrant of the abdomen with a positive Rovsing's sign. Leucocytosis was 13.800 /mm^3 with neutrophilia, C-reactive protein was 68 mg/l. Abdominal ultrasound revealed findings consistent with early acute appendicitis, appendicolith at the appendiceal base with fluid filled appendix measuring up to 8 mm (Fig. 1,2). The patient was hospitalised for the diagnosis of acute no complicated appendicitis. Surgical consent was signed by his parents. The patient underwent a laparoscopic appendectomy. The per-operative exploration showed a phlegmonoeus appendix and an appendectomy was performed. No operative complications were noted. Intravenous antibiotics for 3 days was given with. Pain was controlled with analgesics. The patient did well post-operative and was discharged on third day. Histological exam showed a Grade one, well-differentiated NET at the tip of the appendix developed in the muscularis propia up to the serosal surface Histology with phlegmoneous appendicitis. The tumor was 0.5 cm in size and without mitosis figures. Special histochemical stains were not performed because of the small size of the tumor. Margin of resection was free of neoplasm (Fig. 3). The patient was doing well on routine follow up.

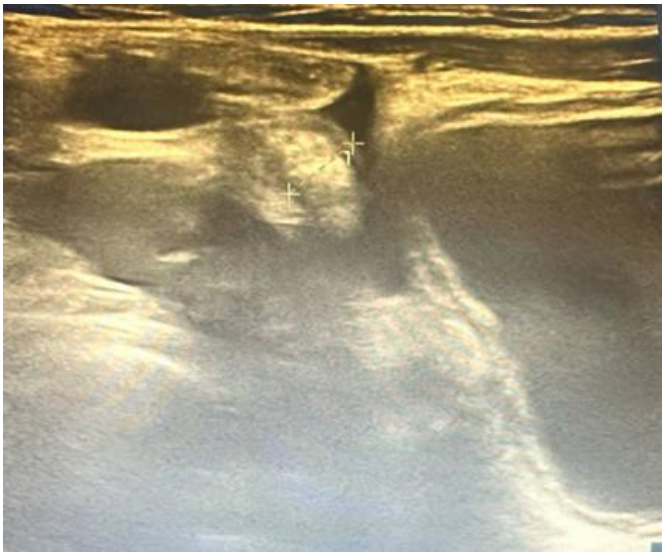

Figure 1: Ultrasound image showing an appendiceal neuroendocrine tumor.

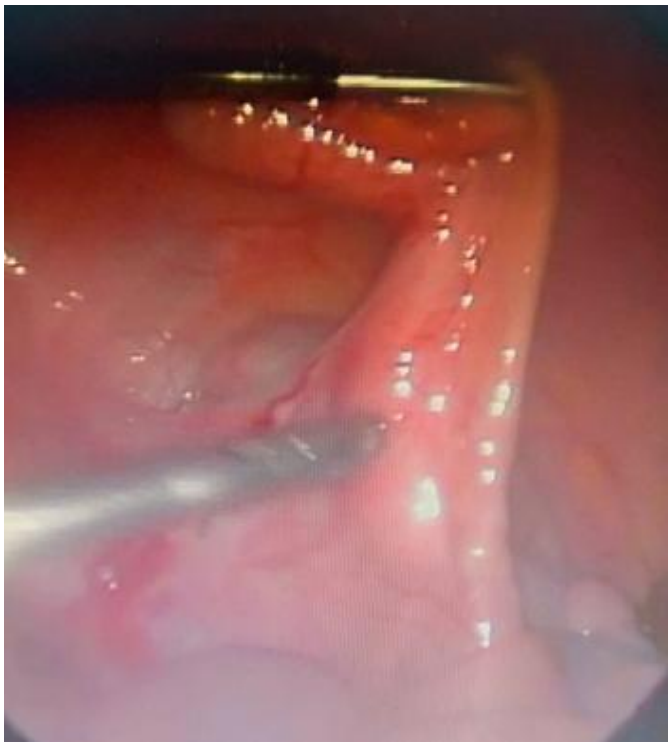

Figure 2: Intraoperative view of the appendicectomy.

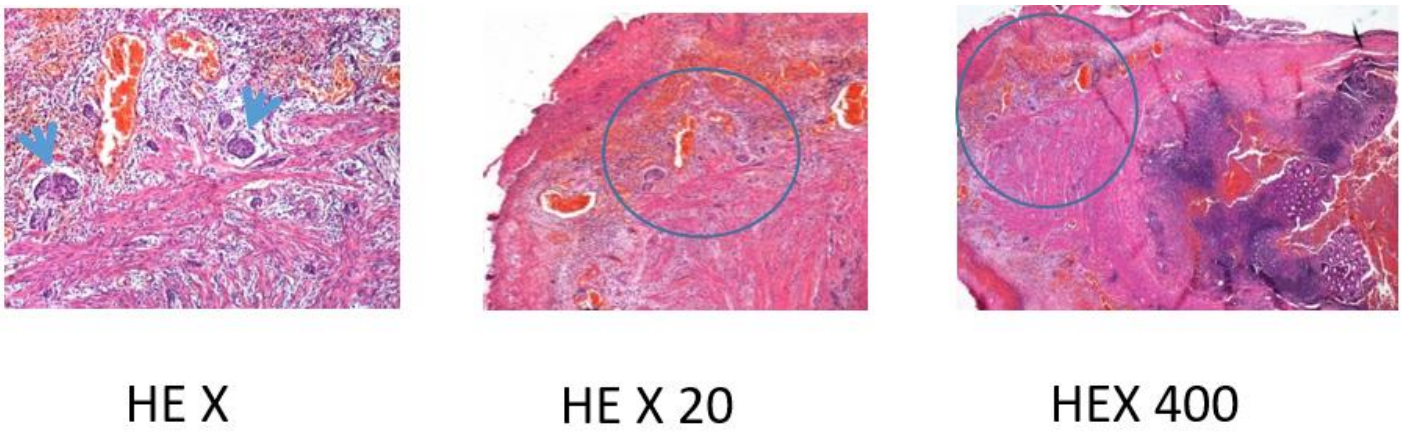

Figure 3: Histological section of the appendiceal neuroendocrine tumor.

Discussion

Neuroendocrine tumors are often considered benign but carry a definite potential for malignant transformation. Due to the indolent nature of the disease, diagnosis is often delayed. It is reported that 10% to 20% of children and young adolescents present with metastatic disease at presentation [5]. There has been a 70-133% rise in the incidence of appendiceal NET's in the last 10 years [6]. It is observed that there is female dominance in pediatric reports of NETs. A short review of 14 appendiceal carcinoid tumors found a female preponderance (64.3%) [7]. Diagnosis of appendiceal NETs is usually established histologically after routine appendectomy, like in our case and occurs in 0.3-0.9% of appendectomies [8]. These tumors were categorized into grade I, grade II or grade III based on mitotic count and/or Ki-67 proliferation index. The Ki-67 index of grade I tumor is ≤2%, for grade II it is 3% to 20% and for grade III it is ≥20% [9]. Grade III poorly differentiated carcinomas were recognized to be highly aggressive with unfavorable outcomes. Studies showed that the TNM staging is not as helpful as is histopathological features in prognosis for these neoplasms [10]. NETs of the appendix are most often located on the tip, do not commonly affect regional lymph nodes and uncommonly metastasize to the liver and they are incidentally diagnosed during appendectomies [11]. Clinical management of NETs largely depends on tumor location, tumor grade, tumor growth rate and the extent of disease and symptoms. Surgical resection is conventionally considered first-line therapy in early-stage disease due to its excellent long-term outcomes [12]. Simple appendectomy is adequate treatment for NET less than 1 cm in diameter. Fortunately, for our patient, surgery was both curative and palliative of symptomatology and is associated with decreased risk of overall and disease-specific death. A review conducted by Murray S, et al., on 50 cases of NETs revealed that all patients included in their review for appendiceal NETs were suspected of acute appendicitis. The absence of tumor recurrence in this study after resection of small appendiceal carcinoids leads us to conclude that post-resection surveillance may not be necessary for tumors ≤1 cm after an R0 resection [13]. another review contacted by Henderson L, et al., on 27 patients at two UK centres with a confirmed histological diagnosis of appendix NET lesions during January 1997-January 2013. All 27 cases are alive and well [14]. This study confirms that paediatric patients with 'incidental' NET tumours of the appendix have an excellent prognosis. In the present case, the patient's tumor was 0.3 cm in size. For tumors of this size, previous studies have indicated that appendectomy is sufficient. The guidelines set by The European Neuroendocrine Tumor Society reveal that in cases of curative resection of appendiceal NEN ≤1cn by simple appendectomy, no specific follow-up strategy has been recommended [14]. Appendiceal NET (G1/2) >2 cm should be treated with oncological right-sided hemicolectomy. Right-sided hemicolectomy is justified only in those rare tumors measuring 1-2 cm but with positive or un- clear margins or with deep mesoappendiceal invasion (ENETS T2), higher proliferation rate (G2) and/or vascular invasion. Tumors with a diameter >2 cm should be treated by right-sided hemicolectomy. In pediatric patients, outcome after appendiceal NET resection has been extremely favorable in the group with tumors between 1 and 2 cm and even in those with tumors >2 cm and thus these guidelines explicitly do not apply to this specific population. The reason for the even better outcome in the pediatric population is currently not understood. For cases with involvement of the lymph nodes or any cases with resected distant metastases, however, long-term follow-up is advised because of the proven invasiveness of the tumor. NET is a rare disease and with reported good prognosis, but it can still be fatal if not handled appropriately. We present the case of a 10-year old boy that presented with an appendiceal G1, well-differentiated neuroendocrine tumor that manifested as acute appendicitis. An appendectomy was performed. Histopathology report revealed the presence of an appendiceal neuroendocrine tumor at the tip of the appendix with clear margins and absence of tumor recurrence.

Conclusion

This case presents the increasing incidence of neuroendocrine tumors, the importance of histopathological examination of the appendectomy specimens and the necessity of more studies focusing on the management. NET is a rare disease and with reported good prognosis, but it can still be fatal if not handled appropriately.

Conflict of Interest

The authors declare no conflict of interest.

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