

Case Report: Acute Appendicitis as Initial Presentation of an Appendiceal Neuroendocrine Tumor

Carolina Topete-Rodríguez¹, Eduardo Martín-De La Torre², María Contreras-Peregrina²

¹Instituto Mexicano del Seguro Social.

²Hospital San Javier.

ABSTRACT

Introduction: Acute appendicitis remains the most common cause of acute surgical abdomen worldwide. While typically regarded as an inflammatory condition, routine pathological analysis can reveal unexpected findings, including appendiceal neoplasms. Neuroendocrine tumors (NETs) of the appendix, though rare, represent the second most common gastrointestinal tumor in pediatric patients and the most frequent appendiceal neoplasm.

Case Presentation: We present the case of a 10-year-old male with clinical signs of acute appendicitis, confirmed by ultrasound and laboratory findings. Laparoscopic appendectomy was performed, and the specimen was submitted for pathological examination. Histopathological analysis revealed a well-differentiated grade 2 neuroendocrine tumor with infiltration into the internal muscular layer. Immunohistochemistry demonstrated diffuse chromogranin and synaptophysin positivity, with a Ki-67 index of less than 2%. Given the absence of metastasis or high-risk features, no further surgical or oncological intervention was required beyond regular follow-up.

Discussion: Appendiceal NETs are frequently discovered incidentally in appendectomy specimens performed for suspected acute appendicitis. Despite their rarity, they necessitate careful histopathological evaluation to determine tumor grade, depth of invasion, and proliferative index, which guide management. In pediatric patients, most cases are managed conservatively with appendectomy alone, given their favorable prognosis.

Conclusion: This case underscores the importance of routine histopathological examination of appendectomy specimens, as unsuspected neuroendocrine tumors may be discovered. Early detection and classification are crucial for appropriate management and long-term surveillance, ensuring optimal patient outcomes.

KEYWORDS: Neuroendocrine tumor, appendicitis, acute abdomen.

ARTICLE DETAILS

Published On:
08 April 2025

Available on:
<https://ijmscrs.com/>

INTRODUCTION

Acute appendicitis is the leading cause of acute surgical abdomen worldwide¹. Pathological classification is fundamental for the management of patients with appendicitis. It is traditionally described as a progressive inflammatory process that can generate an acute phase (edematous), a phlegmonous phase (producing pus), a gangrenous phase (necrosis in its walls), and a perforated phase (loss of continuity in its wall). In Mexico, the incidence of complicated appendicitis has not been reported, but worldwide, the percentages vary from 34 to 37% of the total of appendicitis cases¹.

Routine anatomopathological study of the cecal appendix may find unsuspected anomalies in about 5% of the cases, appendiceal tumors being an important part of these findings². Primary malignant neoplasms of the cecal appendix are infrequent; 1/10,000 cases are reported in the world casuistry and constitute 0.2 to 0.5% of all tumors of the digestive tract³. Their overall incidence is estimated to be between 0.3% and 0.9% of all appendectomies⁴.

The appendicular neuroendocrine tumors (NETs) are neoplasms resulting from neural crest cells and occupy the second place, after lymphoma, as the most frequent

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gastrointestinal tumor in the pediatric age and the first place at the appendicular level⁴.

In terms of differentiation, NETs can be divided into well-differentiated (nuclear monotony and “salt and pepper” granular chromatin with hardly any atypia) or poorly differentiated (vesicular nuclei with marked nucleoli and more atypia). On the other hand, the histological grade is defined according to the mitosis count and/or the proliferative index measured with Ki67, distinguishing 3 grades: G1, G2, and G3. In summary, 4 types of purely neuroendocrine tumors are established: well-differentiated neuroendocrine tumor grade G1, well-differentiated neuroendocrine tumor grade G2, well-differentiated neuroendocrine tumor grade G3, and poorly differentiated neuroendocrine carcinoma grade G3⁴.

The clinical presentation of NETs is variable, ranging from an asymptomatic patient, to a presentation of acute appendicitis⁵. Most of these tumors are operated with a diagnosis of acute appendicitis or acute abdomen and are later identified as an incidental finding in the histopathological study of the surgical specimen³.

In the absence of metastasis, the prognosis is very good, with 5-year survival rates close to 100%⁴.

CLINICAL CASE

A 10-year-old male patient with no important personal pathologic history.

The father states that the patient started 3 days before his admission with intermittent abdominal pain, accompanied by a diarrhea-like evacuation as well as a fever peak of 38.5 °C, nausea, and anorexia. 5 °C, nausea and anorexia, analgesia and probiotics were started in an external health center, without improvement, becoming a disabling pain so they decided to go to the emergency department of Hospital San Javier where he was evaluated; the physical examination showed a wooden board abdomen, decreased peristalsis, as well as positive signs of McBurney, Psoas, Obturator and Blumberg. The rest of the systems on physical examination were not altered.

An abdominal ultrasound was performed, where conclusive data of acute appendicitis was reported. Hematological biometry was requested, and leukocytosis of 14,000 mm³ with predominance of neutrophils and bandemia was reported.

Laparoscopic appendectomy was performed with findings of free fluid in the cavity of approximately 50ml of serohematic characteristics, appendix in suppurative phase with fibrinopurulent exudate, and fibrin creams. An appendectomy was performed without complications.

The specimen was sent to pathological anatomy (Figure 1). The patient had a favorable postoperative course and was discharged after 24 hours.



Figure 1: Tissue belonging to appendectomy.

Anatomic Pathology reports the following: Tissue belonging to appendectomy. Increased volume corresponding to distal third, with congestive changes and deposit of fibrin creams; second tubular fragment, fibrous appearance with opaque serous surface and faint congestive changes. Altogether, they show a length of 8 cm, with larger diameters of 2.3x1.6 cm.

Microscopic description: Histopathological elements corresponding to cecal appendix, neoplastic affection in the middle portion, represented by cellular elements of small to intermediate size, which are arranged in syncytia of organoid aspect at central level, with signs of infiltration to the internal muscular wall, adopting an architectural pattern in rows and cellular cords (Figure 2). The characteristics of the cell population show regular nuclei with chromatin in fine clumps in a “salt and pepper” image, with inconspicuous nucleoli, as well as the presence of 3 mitotic figures in a scan of 50 high-power fields. This lesion infiltrates up to the internal muscular wall without overgrowth. It shows inflammatory histopathological changes associated with acute suppurative appendicitis.

Interpretation: Well-differentiated neuroendocrine tumor grade 2.

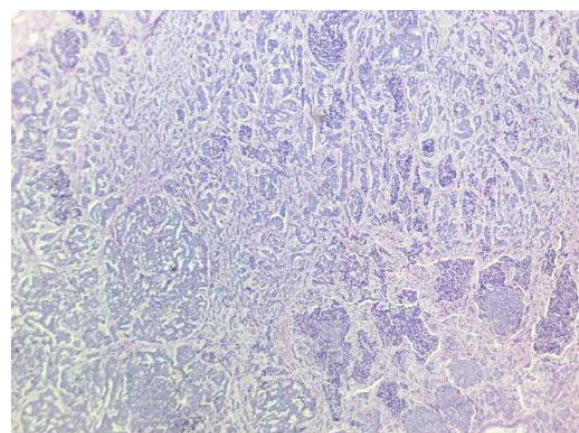


Figure 2: Histopathological elements corresponding to cecal appendix, neoplastic affection in the middle portion.

It was decided to complement the study with immunohistochemical analysis for neuroendocrine markers:

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chromogranin, synaptophysin, and Ki57 to assess the cell proliferation index.

The immunohistochemistry analysis reports: “Complementary study is performed in paraffin-embedded tissue”. Diffuse positive Chromogranin in neoplastic cells, intense positive Synaptophysin in neoplastic cells, positive Ki 67 (MIB-I clone) in less than 2% of neoplastic cells.

The patient consulted with pediatric oncology who ruled out the need for management with chemotherapy, radiotherapy, or right hemicolectomy, however, it was suggested to continue with oncological surveillance during the following years.

DISCUSSION

Neuroendocrine tumors are distributed throughout the digestive system, 19% are located in the cecal appendix³, they are infrequent in pediatric age, although they are a common group of gastrointestinal tumors⁴; most of these tumors are operated with a diagnosis of acute appendicitis or acute abdomen and are later identified as an incidental finding in the histopathological study of the surgical specimen³, in a study performed by Álvarez-Álvarez et al. with a total of 105 patients diagnosed with appendicular tumors, which were classified as benign and malignant: 10 cases of malignant tumors (9.52%) and 95 of benign tumors (90.47%) were obtained. Although there are currently no guidelines to guide us in the diagnosis and management of these tumors in the pediatric age, in most cases, appendectomy is curative, and the prognosis is excellent⁴. The presented case is consistent with findings in the international literature, representing a typical clinical manifestation of this pathology. While the diagnosis aligns with established criteria, it was not initially suspected and was only confirmed through histopathological analysis, as documented in similar case report and studies.

CONCLUSION

It is essential to emphasize the need to perform a histopathological study in all patients undergoing appendectomy. As previously mentioned, it is rare to suspect

an appendiceal neoplasm before or even during surgery when acute appendicitis is present, which may result in a delayed diagnosis and a less favorable prognosis.

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