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Neuroendocrine Tumor of Appendix Presenting as Acute Appendicitis : A Case Report

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1. Abstract

Appendicitis is a common cause of abdominal pain that demands immediate surgical intervention to avert complications such as rupture and peritonitis. Case reports about patients with appendicitis, who had an appendiceal neuroendocrine tumors (NETs) post-appendectomy, are scarce. Diagnosis at early stage and speedy treatment is necessary to reduce morbidity and mortality rates. In this case, a 41-year-old male presented with two days of sharp non-radiating right iliac fossa pain with no fever, nausea, vomiting or change in bowel movements. Physical examination revealed abdominal tenderness localized at the right lower quadrant accompanied by positive McBurney's sign. In imaging he was indicated to be having acute appendicitis while laparoscopy showed acutely inflamed gangrenous appendix with appendicular adhesion to the abdominal wall. Patient made an uneventful recovery and was discharged the following day. Histopathological examination revealed well-differentiated neuroendocrine tumor measuring 0.3 cm at the organ of appendix without lymphovascular invasion and negative margins were recorded. A follow-up multi-disciplinary team confirmed low risk for recurrence on post-surgical colonoscopy and further laboratory tests recommended. Preoperatively diagnosing appendiceal neuroendocrine neoplasms is difficult since they are rarely symptomatic prior to their incidental identification through appendicitis management. A concomitant detection of a primary neuroendocrine neoplasm with acute appendicitis is an accidental diagnosis that emphasizes the need of an extensive diagnostic work-up and a multidisciplinary approach to have better results for patients.

2. Introduction

Neuroendocrine tumors (NETs) are a rare and heterogeneous group of neoplasms that display aggressive behaviour, originating from neuroendocrine cells; which are distributed throughout the body being mostly common in the gastrointestinal tract and localized in appendix [1-3]. Appendiceal NETs are uncommonly encountered neoplasms that can either be an incidental finding or mimic symptoms of acute appendicitis [2]. When a patient is operated with clinical suspicion for acute appendicitis, special care must be taken during histopathological examination to look for NETs [3]. This unforeseen result would require further investigation and management using tumor-specific modalities, thus underscoring the need for awareness and multi-disciplinary collaboration in managing these cases [4-5]. Appendiceal neuroendocrine tumors (ANETs) comprise three major types: Appendiceal Neuroendocrine Tumors (ANETs), Appendiceal Neuroendocrine Carcinomas (ANECs), and Mixed Neuroendocrine-Non-Neuroendocrine Neoplasms (MiNENs) [3]. For instance, ANETs are often found incidentally during appendectomy for acute appendicitis and managed by simple appendectomy for tumors ≤2 cm [2]. On the other hand, larger tumors or those with high-risk features require right hemicolectomy with lymph node dissection [2]. Rare but aggressive, the poorly differentiated ANECs should be surgically removed before being exposed to chemo based on platinum compounds [2]. Others, like MiNENs contain both non-neuroendocrine as well as neuro-endocrine portions of which the later is more aggressive and

requires combined surgical intervention plus chemotherapy 3. The proper management of these conditions depends on accurate histopathological typing onto which appropriate surgical excision treatments may be grounded on subsequent adjuvant therapy [2].

3. Case Presentation

A 41-year-old adult male patient presented to us complaining of pain in the right iliac fossa over two days starting from 20th March, 2024. He described this as an irritating stab which had evolved into a more serious state and brought him here. There was no accompanying nausea, vomiting or fever. On examination, there was tenderness in the right lower quadrant with a pain score of 7/10 suggestive of acute appendicitis. Laboratory work-up showed minor rise of serum creatinine at 1.33 mg/dL and normal GFR necessitating further search for renal function. However, all the inflammatory, septic markers, and urine analysis were unremarkable. As per Dubai Hospital policies, it is necessary to perform an abdominal CT scan for any patient who presents with appendicitis and is more than 40 years of age to rule out malignancy. Importantly, CT scan revealed an enlarged inflamed appendix measuring about thirteen to fourteen centimeters that showed fat stranding (Figure 1). Moreover, intraluminal appendicoliths served as additional evidence towards clinical diagnosis of acute appendicitis (Figure 2). Importantly, no specific alternate layover for intraabdominal pathology was recognized. An early laparoscopic appendectomy was carried out based on clinical and radiological evidence. Acute inflammation of the appendix with minimal adhesions with the abdominal wall and surrounding structures were noted. The patient had uneventful postoperative period and discharged on second postoperative day. Histopathology showed acute appendicitis, as well as an incidental finding of NET within the submucosal layer. Further examination revealed that the NET was a well-differentiated neoplasm designated low grade 1 (G1) (Ki67 index <3%). With no lymphovascular invasion or metastasis, this tumor measured 0.3 cm; however, there was penetration into the submucosal layer. This case was discussed by a multidisciplinary team and the decision was that surgical intervention is not required, and colonoscopy appointment was scheduled to follow up the patient. Unfortunately, we tried calling the patient several times but he didn't pick up and patient lost to follow up regarding colonoscopy.

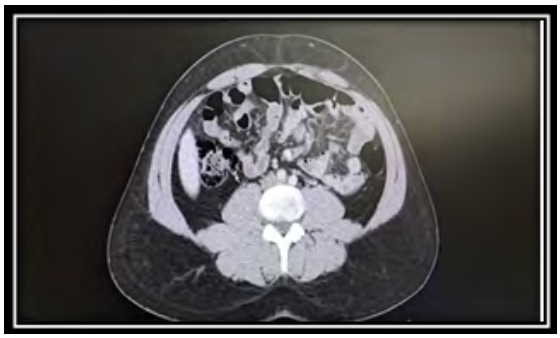

Figure 1: CT scan revealed an enlarged inflamed appendix measuring about thirteen to fourteen centimeters that showed fat stranding.

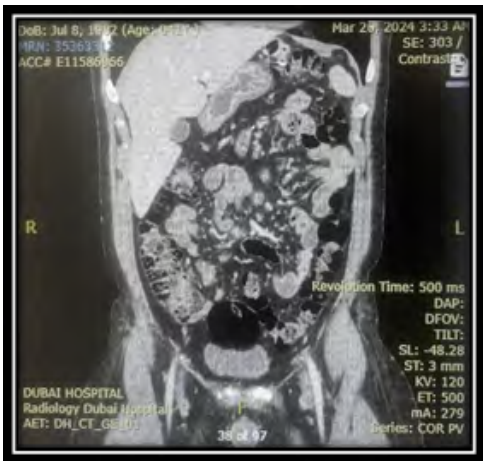

Figure 2: Moreover, intraluminal appendicoliths served as additional evidence towards clinical diagnosis of acute appendicitis.

4. Discussion

Appendiceal neuroendocrine tumors (NETs) are a category of appendiceal epithelial neoplasms that are believed to derive from neuroendocrine cells in the appendix [2]. They are ordinarily sporadic and can be grouped as well-differentiated neuroendocrine tumors (NETs), poorly differentiated neuroendocrine carcinomas (NECs) or combined neuroendocrine-non-neuroendocrine neoplasms (MiNENs) [1]. Additionally, goblet cell adenocarcinoma which was previously believed to be mixed form of aNENs is now placed among adenocarcinomas [4]. The majority of appendiceal carcinoid neoplasms present as incidental findings on pathology examination specimens following appendectomy accounting for almost 80% cases [5]. Though rare, it is the fifth most common NET in the gastrointestinal tract with incidences ranging between 0.15 and 0.6 per 100000 person years 5. It has been proposed that pathophysiological causes of appendiceal NETS involve enterochromaffin cell NETS particularly for those originating from subepithelial neuroendocrine cells lining mucosal crypts or submucosal glands [3]. However, what leads to these abnormal growths remains largely unknown [3]. Somatostatin receptor imaging currently represents a worldwide diagnostic test for neuroendocrine neoplasms; however, diagnosis of appendiceal NETs is also limited by diagnostic modalities [2]. In some cases, increased concentrations of chromogranin A, 5-HIAA, neurokinin A or pancreatic polypeptide may be detected using laboratory testing [2]. Due to their relatively small size, appendiceal NETs are typically not visible on medical imaging devices and they are common characteristics of acute appendicitis familiar presentations from ultrasound to CT imaging [5]. Nevertheless appendix cancers have an exceptionally optimistic prognosis even though their diagnosis poses diagnostic challenges with survival rates exceeding 92% for up to 10 years in some cases [5]. Majority of these metastasize at most regional lymph nodes at worst and fiver year survival is greater than 85% for all patients including those with regional metastatic disease [5]. However a decrease in survival has

been observed in cases involving more than 3 mm invasion and mesoappendiceal invasion [1]. The proper management depends on the extent of differentiation, aggression and metastasis in the organ under attack 1. Even though many controversies still exist about surgical treatment options for localized appendiceal NETs, some of the factors that can come into play include size of tumor, mesoappendix tissues spreading, perineural participation as well as lymphovascular invasion among others 1. Currently, patients with >2cm aNENs should undergo right complete hemicolectomy because their high risks of getting lymph node involvement while simple appendectomy is curative and requires no further action for <1 cm aNENs [4]. Our 41 year old patient's management aligns with the latter option since his histopathology result revealed a well-differentiated, low-grade 1 (G1) neuroendocrine tumor (NET) with a Ki67 index of less than 3% and measuring just 0.3 cm in size. After the multidisciplinary tumor board reviewed his case, they determined that no further intervention is required. Instead, they recommended a follow-up colonoscopy to monitor his condition. The patient failed to respond to multiple phone calls from the medical team, resulting in a missed opportunity for the scheduled colonoscopy follow-up. These types of neoplasms are significant mainly due to the diagnostic challenge presented by incidental finding of appendiceal NETs in cases of acute appendicitis 3. While these tumors are generally benign, their discovery demands thorough assessment by multidisciplinary teams and long term follow up plan so as to optimize patient care 2. Given insights from this case; we therefore stress on increasing surveillance and teamwork across disciplines when dealing with the perplexing world of appendiceal NET's 1.

5. Conclusion

The incidental finding of a neuroendocrine tumor (NET) in the appendix is illustrative of diagnostic challenges in surgical practice. The incident finding of a neuroendocrine tumor (NET) in the appendix is an example of how hard it can be to diagnose what goes on during surgery. Careful histopathologic analysis and investigation revealed well-differentiated NET, underscoring the need for careful examination of appendectomy specimens. This case highlights the need for constant recognition that appendiceal NETs may be associated with acute appendicitis and that more extensive pathology must be undertaken to ensure correct diagnoses, as well as proper management based on histopathology. Concomitant discovery of an acute appendicitis with an appendix neuroendocrine tumor is a coincidental diagnosis which stresses the importance of thorough evaluation of histopathology specimen post appendectomy, and a multidisciplinary approach to achieve better outcomes for patients.

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