Clinical Surgery Journal

Case Report | Vol 4 Iss 3

Acute Appendicitis Mask Serious Diagnostic in a Child: Neuroendocrinien Tumor and Leukemia

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Received: February 15, 2021; Accepted: March 20, 2021; Published Date: March 27, 2021

ABSTRACT

Acute appendicitis is common surgical emergency among children (1% - 2% in pediatrics surgical admissions) However, despite wide spread availability of sophisticated diagnostic imaging, acute appendicitis may mask severe diagnostic. We report the cases of 3 children's how were diagnosticated as acute appendicitis to fall on appendicular neuroendocrine tumor NET of appendix in 2 cases and one case of an acute myelogenous leukemia AML. The anatomopathology study was the key of diagnostics.

KEYWORDS

Neuroendocrine tumor; Appendix; Myelogenuis leukemia; Children

INTRODUCTION

Acute appendicitis is one of the frequent causes of abdominal pain in children [1]. The lifetime risk of developing acute appendicitis among males and females is 8.6% and 6.7%, respectively, however it can hide serious pathology such as appendiceal cancer and leukemia.

Tumor of appendix and acute leukemia are a rare malignancy. There most often diagnosed following appendectomy for suspected acute appendicitis [2]. In the pediatric aged group, there are very little cases being published of appendix tumor and the association of acute appendicitis and leukemia; the histopathological study have a crucial role for diagnostics [3].

CASE REPORT

The two cases of tumor had similar clinical presentation: 13-years old and 10-years old child. They had repeated appearance of the abdominal pain and spontaneous disappearance of the symptom for 1 month, but abdominal pain gradually got worse, in the admission, they had fever and Mc Burney defenses. Biological examination and imaging study showed acute appendicitis. Appendectomies were performed founding appendiceal tip mass. Although the body of the appendix looked intact as well as the ileum and the cecum, the histopathological examination revealed neuroendocrine process us in the tip of the resected appendix. Conservative treatment was realized. The patients are doing well till today on follow-up without progression of the disease 2 years after the operation (Figure 1).

Citation: Zaari N, Acute Appendicitis Mask Serious Diagnostic in a Child: Neuroendocrinien Tumor and Leukemia. Clin Surg J 4(3): 1-4.

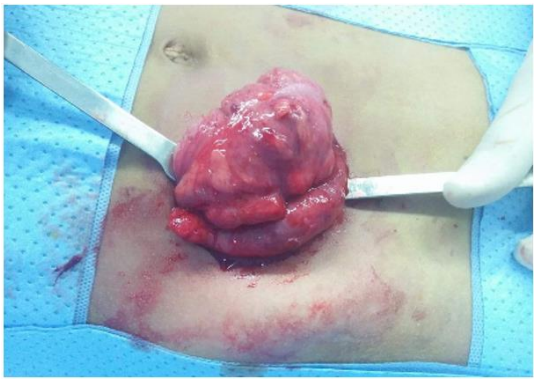

Figure 1: Preoperative view of appendiceal mass in the NET case.

The third case is a 9-years old girl, presented in emergency for abdominal pain and fever for 2 weeks getting more and more severe, physical examination found abdominal guarding especially in the right iliac fossa, with conservative general state; blood analysed show important inflammatory syndrome and abdominal ultrasound was in favor of a appendicitis with a thickened appendix around 8 mm. appendectomy was performed immediately. Showing a normal macroscopic aspect, cecum and small bowel were unroll did not reflect both abnormality, the histopathological and immunohistochemistry illustrate profuse blastic cell infiltration in the appendiceal with atypical mononuclear cell which are myeloid origin with immunoreactivity to CD45 and Bcl2.

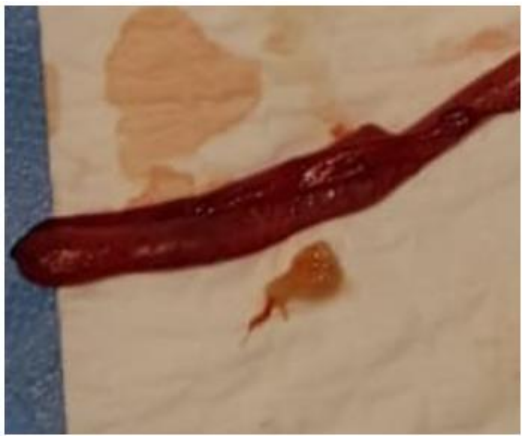

Figure 2: Macroscopic aspect of the appendix in the AML case.

Postoperative course was uneventful; complementary examination was mad, therapeutic protocol of leukemia was initiate. Following some cycle of therapy, her performance status had declined, after one year of follow up she passed away (Figure 2 and Figure 3).

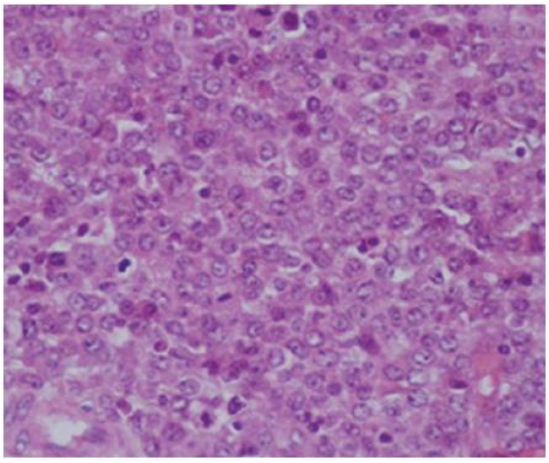

Figure 3: Infiltration of the appendix by monomorphic leukemic cells.

DISCUSSION

Tumor of appendix in children is uncommon with an incidence of around 0.1 in 1,000,000, making up only 0.5% of all gastrointestinal malignancies [2]. Delay in the diagnosis and management predominantly result from poor communication skill, failure to elicit physical signs in irritable children, atypical presentation, and overlap of symptoms with other disorders. Pediatric neuroendocrine tumors (NETs) of the appendix are mostly detected incidentally after appendectomy for acute appendicitis [4].

Neuroendocrine tumors (NETs), which were first described by Oberndorfer in 1907, arise from enterochromaffin cells found throughout the gastrointestinal tract and the bronchopulmonary system [5]. The etiology of NETs is not well understood. The tumor itself is not aggressive but has the potential for rupture and spread throughout the peritoneum [2], there is a known correlation between tumor size and NET localization in the appendix: small tumors are more often situated at the tip, whereas larger tumors tend to be at the base. NET management is a matter of controversy but the review of the literature proof that two criteria may point to the need for additional surgery, the possibility of regional

lymph node involvement: Tumor size >20 mm and incomplete surgical resection margins. Childhood NET of the appendix has an excellent prognosis [4].

According to Boxberger et al. (prospective analysis of 237 patients), a right hemicolectomy is recommended in all patients with a primary tumor size of >15 mm because of significant risk for developing a local lymph node metastasis [6]. Lambert et al., affirm that the appendectomy alone, in all cases even if incomplete resection, seems to be curative for appendiceal NETs in children, with no influence on life expectancy [7].

In our clinical cases, the tumor was located at the tip of the appendix and the patients experienced acute right lower abdominal pain witch lead to the clinic diagnosis of acute appendicitis. The surgical procedure was performed for the treatment of appendicitis, and the NET was identified incidentally during the histological examination. In the 2 present patients, the tumor was under 1 cm in diameter and therefore, in the light of current knowledge of ANETs treatment, no additional treatment was needed.

For the cases of acute myeloid leukemia AML, Appendicitis in leukemic patients is uncommon but associated with increased mortality. Additionally, leukemic cell infiltration of the appendix is extremely rare [2]. Leukemic cell infiltration of the appendix, first reported by Rappaport in 1967, acute appendicitis has infrequently been described in the setting of known acute leukemia and is generally associated with patients receiving active chemotherapy [8]. Acute appendicitis as the initial manifestation of AML, as in this case, has only been described in 3 other cases where the patients were found to have AML. This case illustrates the importance of maintaining a high suspicion for acute leukemia in the setting of a significant leukemoid reaction even if a clear acute process such as appendicitis is present [9].

CONCLUSION

When encountering atypical cases of the appendicitis, neuroendocrine tumors of the appendix or AML should be considered as a cause of acute appendicitis during appendectomy, therefore the histopathological examination of all the specimens' appendix are essential.

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