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Bening behavior of pediatric appendiceal NETs: is there a need for additional surgery?

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INTRODUCTION

Neuroendocrine tumors of the appendix (aNET) are rare in children even though they are considered one of the most common gastrointestinal pediatric neoplasms.

Due to their rarity in pediatric population, there is no consensus on treatment and follow-up.

We aimed to discuss the management of two patients who were incidentally diagnosed with aNET following histopathological examination after appendectomy.

CASE REPORTS

Case 1 : A 12-year-old female presented with generalized abdominal pain and signs of acute abdomen.

Abdominal CT revealed peri-appendiceal and perihepatic fluid and free air. (Figure 1)

Due to intra-abdominal adhesions and extensive abscess formation, laparoscopy was converted to laparotomy, which revealed a perforated appendix, and an appendectomy was performed.

Postoperatively, the patient developed renal dysfunction requiring two sessions of hemodialysis, pleural effusion that necessitated drainage, and elevated pancreatic enzymes consistent with pancreatitis. Subsequently, a widespread rash developed, suggesting a possible drug allergy or carcinoid syndrome. (Figure 2)

Histolopathological examination revealed a 1.2x0.6×0.7 cm **WHO grade 2 aNET** infiltrating into the subserosa, without lymphovascular or perineural invasion. Margins were clear. (Figure 3)

A PET-CT scan was performed for metastatic evaluation, revealing no evidence of metastasis. No additional surgery or chemotherapy was planned, and she remains free of recurrens at 9-month follow-up.

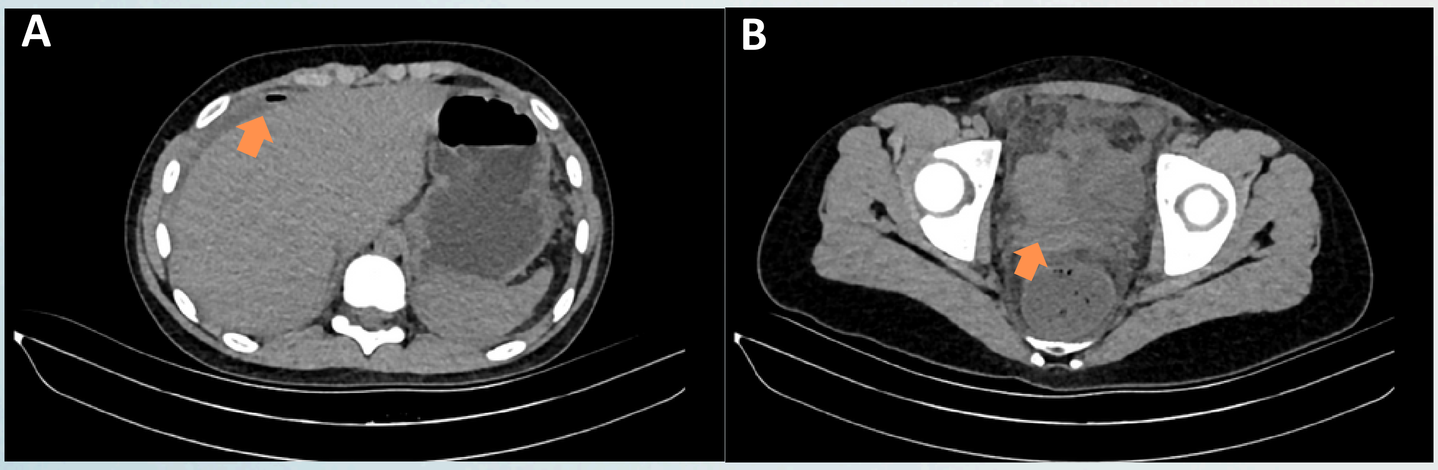

Figure 1. Findings of perihepatic fluid and free air on CT scan (A) and peri-appendiceal fluid (B). (arrows)

Figure 2. Postoperative rash

Case 2: A 16-year-old male presented with right lower quadrant pain.

Abdominal CT confirmed acute appendicitis, and laparoscopic appendectomy was performed.

The postoperative course was uneventful.

Histopathological examination revealed a 1 mm **WHO grade 1 aNET** in the distal appendix, invading the submucosa. No lymphovascular or mesoappendiceal invasion was identified, and resection margins were clear.

No further intervention was required. The patient remains recurrence-free at 8 months of follow-up.

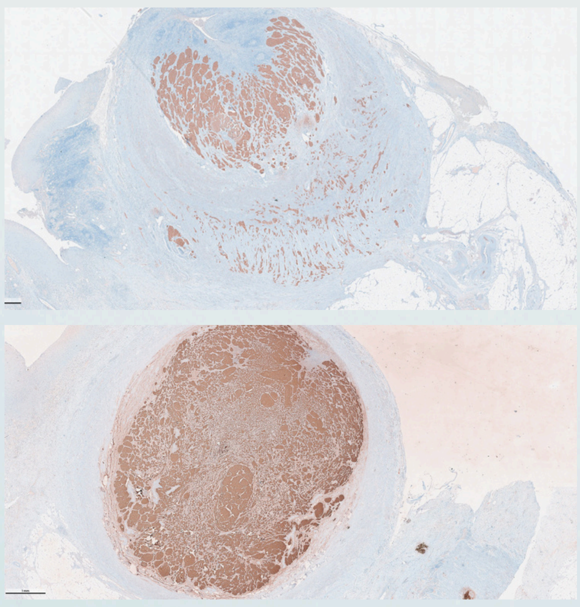

Figure 3. Synaptophysin and Chromogranin A immunostaining showing cytoplasmic positivity in tumor cells, supporting neuroendocrine differentiation

CONCLUSION

ANETs in children have an excellent prognosis and rarely require surgery beyond appendectomy.

Our findings support the benign nature of this tumor and demonstrate that appendectomy alone is sufficient in most pediatric cases. Surgery remains the mainstay of management, with tumor size being the most critical factor in determining the extent of surgical intervention.

Given the lack of international consensus in pediatric cases, collaborative studies and guidelines are warranted.