

BACKGROUND

- Neuroendocrine tumor (NET) of the appendix is the most common gastrointestinal epithelial tumor in children.
- Neither the utility of serum markers nor the indication for hemicolectomy have been established in children.

PURPOSE

- To describe the clinical features, treatment and outcome of children with NET of appendix.

METHODS

- Retrospective analysis of clinical features and outcome of patients with NET of appendix diagnosed at Texas Children's Hospital between 1995 and 2014.

RESULTS

- Forty-five children with a median age of 12 years (range 6-17 years) were identified (Table 1).
- The pathology slides of 35 patients were reviewed (Table 2).

Table 1. Patient Characteristics (n=45)

Age at diagnosis (years)	
Median (range)	12 (6-17)
Gender	
Male	20
Female	25
Reason for appendectomy	
Acute appendicitis	40
Incidental appendectomy	5
Somatostatin scan	
Performed	5
Abnormal	0
Chromogranin A	
Performed	4
Abnormal	0
5-HIAA	
Performed	9
Abnormal	0
Serotonin	
Performed	1
Abnormal	0
Right hemicolectomy	7

Table 2. Histopathologic Characteristics (n=35)

Size (cm)	
Median (range)	0.5 (0.1 – 1.2)
AJCC T1a	30
T1b	5
Location	
Body of Appendix	19
Tip of Appendix	16
Depth of Invasion	
Submucosa	8
Muscularis Propria	9
Serosa	8
Mesoappendix	10
Nuclear Grade	
Low	23
Intermediate	2
Mitotic Activity	
Present	7
Absent	28
Vascular invasion	2
Perineural invasion	23
Acute appendicitis extending into the tumor	13
Acute inflammation in background	28

Table 3. Characteristics in Patients Undergoing Hemicolectomy

Pt.	Location	Size (cm)	Depth of invasion	Lympho-vascular invasion	Indication for hemicolectomy	Hemicolectomy findings
1	Body	0.7	Mesoappendix	Present	Large size (1.5 cm by gross examination measurement) and vascular involvement	No tumor identified
2	Body	0.8	Serosa	Absent	Tumor near resection margin	micrometastasis in 1/43 LNs
3	Mesoappendix	0.1	Appendiceal Soft Tissue	Absent	Invasion of mesoappendix	No tumor identified
4	Tip	0.7	Mesoappendix	Absent	Invasion of mesoappendix	No tumor identified
5	Tip	0.9	Mesoappendix	Absent	Invasion of mesoappendix	No tumor identified
6	Tip	1.0	Mesoappendix	Absent	Invasion of mesoappendix	Tumor nests in cecal/appendiceal mesentery
7	Tip	1.2	Mesoappendix	Absent	Invasion of mesoappendix	No tumor identified

RESULTS

- The median tumor size was 0.5 cm, and 46 % of tumors were located in the tip of the appendix. Invasion of mesoappendix was seen in 29%.
- Seven patients (16%) underwent hemicolectomy (Table 3).
- Only two hemicolectomy specimens showed disease, with one in the appendiceal mesentery, and the other as a micrometastasis in a mesenteric lymph node.
- Only 4 of the 10 patients with confirmed mesoappendix invasion underwent hemicolectomy.
- There were no recurrences and all patients were alive without evidence of disease at last follow-up (median 3.7 months; range 0-150 months)

CONCLUSION

- NET of the appendix in children tends to have a benign clinical course with excellent prognosis.
- In the absence of carcinoid syndrome, postoperative scans and serum biomarkers do not appear to be useful.
- In completely resected tumors, the indication of hemicolectomy is unclear.