

Neuroendocrine Tumor of the Appendix in Children

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Background: Neuroendocrine tumor (NET) of the appendix is the most common gastrointestinal epithelial tumor in children. It is usually diagnosed as an incidental finding in appendectomy specimens. The utility of serum markers or the indication for hemicolectomy have not been established in children.

Methods: The presentation, treatment and outcome of all children <18 years of age diagnosed at Texas Children's Hospital between 1995 and 2014 were retrospectively reviewed. Pathology slides from the appendectomy and hemicolectomy, when available, were reviewed to determine tumor size, location, invasiveness and presence of metastasis.

Results: Forty five children with a median age of 12 years (range 6-17 years) were identified. The majority (56%) were female. NET was an incidental finding following appendectomy performed for suspected acute appendicitis (89%) or incidental appendectomy during other surgical procedures (11%). Somatostatin scan (n=5), serum chromogranin A (n=4), and Urine 5-HIAA (n=9) were obtained post-operatively and were within normal limits. The pathology slides of 43 patients were reviewed. The median tumor size was 0.5 cm, and 42% of tumors were located in the tip of the appendix. Invasion of mesoappendix was seen in 26%. Perineural invasion and vascular invasion were seen in 63% and 12% respectively. Seven patients (16%) underwent hemicolectomy. The indications for hemicolectomy were invasion of mesoappendix

(4), tumor infiltration near the serosal margin (1), tumor size 1.5 cm with vascular invasion (1) and persistence of previously unrecognized carcinoid syndrome post-surgery (1). Only two hemicolectomy specimens showed disease; one in the appendiceal mesentery and other a micro metastasis in a mesenteric lymph node. There were no recurrences and all patients were alive at last follow-up.

Conclusions: NET of the appendix in children is a benign disease with excellent prognosis. In the absence of carcinoid syndrome, post-operative scans and serum biomarkers are not useful. In completely resected tumors, the indication for hemicolectomy is unclear.