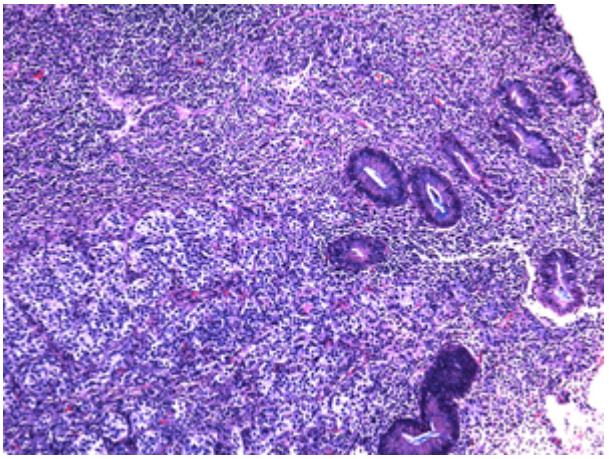
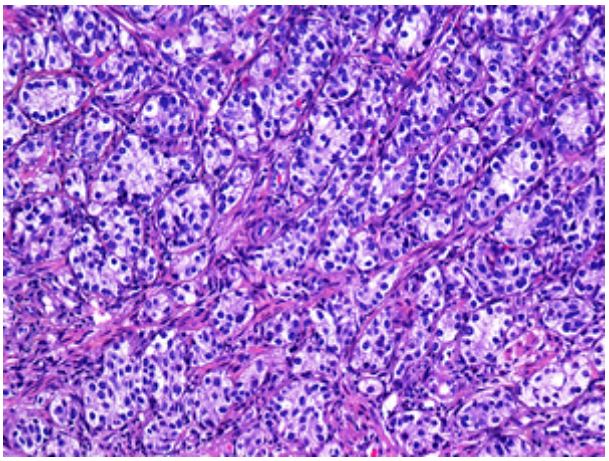
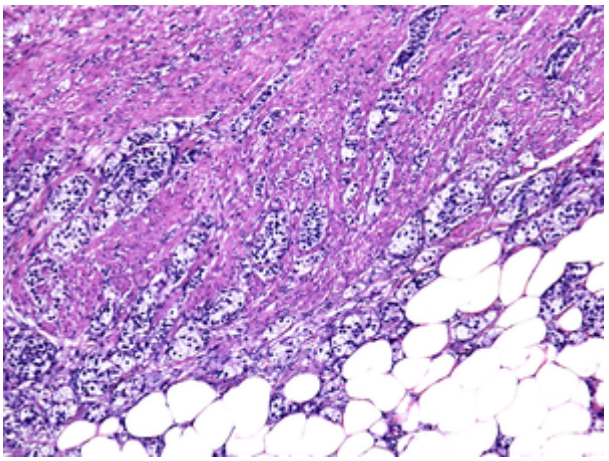
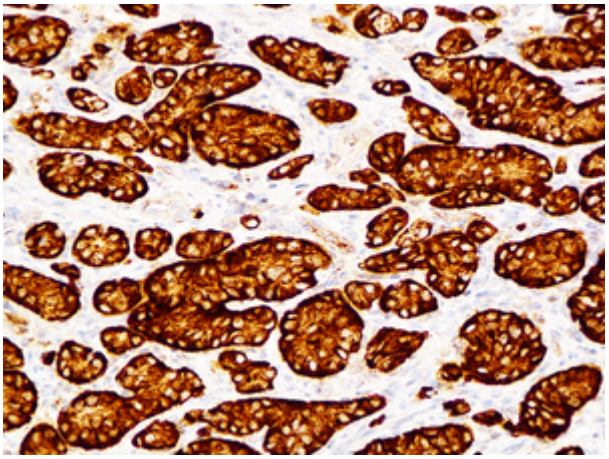
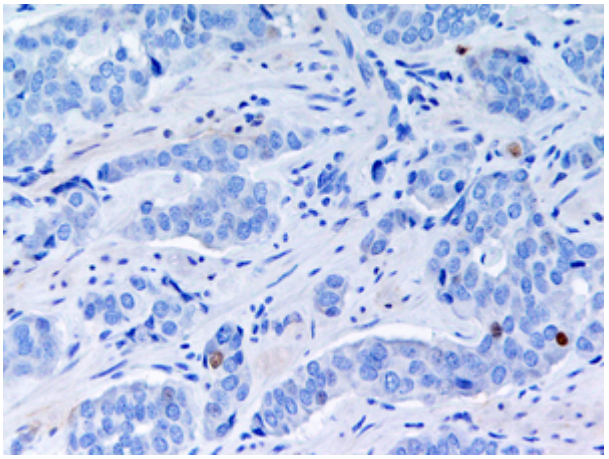
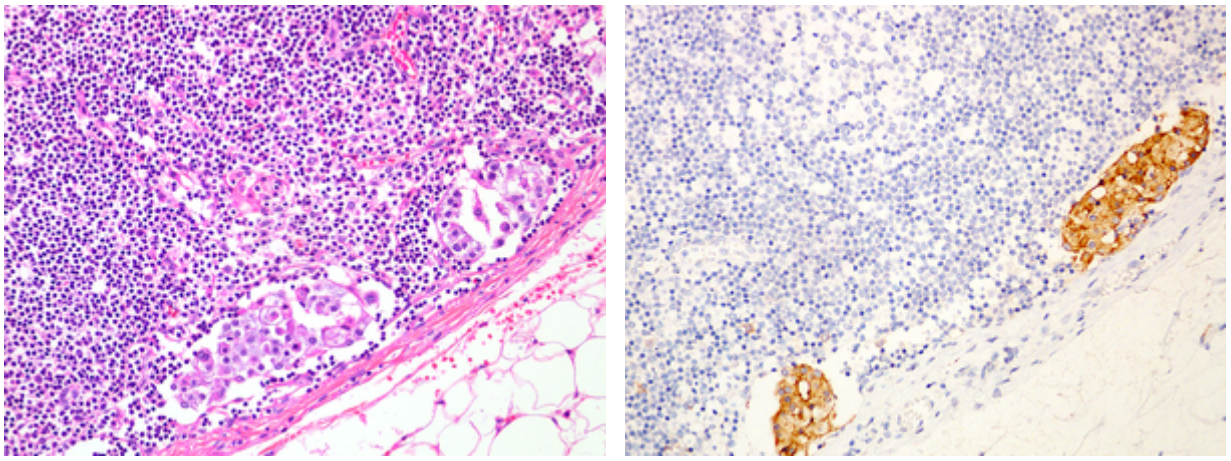
June 2016

Appendectomy specimen from a 13-year-old girl with clinical diagnosis of acute appendicitis.

What is your diagnosis?



Diagnosis:

Well-differentiated neuroendocrine tumor (NET G1) at the base of appendix, with metastasis in a regional lymph node.

Comment:

A 13-year-old girl with clinical symptoms of acute appendicitis underwent appendix resection. Grossly, appendix measured 5 x 1.5 cm and exhibited signs of acute inflammation. At the base of appendix, there was a yellowish tumor (Panel A) measuring 1.5 cm in diameter. The tumor grew close to the resection margins, but the margins were free of the tumor. Microscopically, tumor exhibited characteristic features of a well differentiated neuroendocrine tumor (NET) (Panel B and C), with focal invasion of the mesoappendix (Panel D) and positive immunohistochemistry for chromogranine (Panel E) and synaptophysin and Ki67 index <2 % (Panel F). No lymphovascular invasion was found. Further clinical examination revealed enlarged ileocolic lymph nodes by ultrasound, and ileocecal resection was performed. No residual tumor was found in the resection specimen, but there was metastasis in one of the ilecolic lymph nodes (Panel G), confirmed by immunohistochemistry for chromogranine (Panel H).

Neuroendocrine tumors (NETs) are the most frequent tumors of appendix. Most are asymptomatic and occur at the tip of appendix. Approximately 10% are located at the base of appendix, frequently causing obstruction and acute appendicitis. In contrast to NETs at other locations, appendiceal NETs behave less aggressively and tumor size is considered more important than depth of invasion. Separate staging criteria have therefore been proposed for appendiceal NETs, with tumor size as a major criterion for aggressiveness for localized tumors. A risk for metastasis is approximately 1% for NETs smaller than 2 cm, and 20-40% for those larger than 2 cm. There is general agreement that a right-sided hemicolectomy should be performed in NETs >2 cm. However, it is controversial whether radical surgery is indicated in tumors measuring <2 cm. Possible additional risk factors include deep invasion into the mesoappendiceal fatty tissue, the presence of vascular invasion, location at the base of appendix and high proliferation rate. The presented case is one of the very rare small appendiceal NETs that metastasized to the lymph nodes. Even in these cases, an excellent prognosis can be expected, as suggested by recent studies.

For further reading:

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- Pape UF, Niederle B, Costa F, Gross D, Kelestimur F, Kianmanesh R, Knigge U, Öberg K, Pavel M, Perren A, Toumpanakis C, O'Connor J, Krenning E, Reed N, O'Toole D; all other Vienna Consensus Conference participants. Consensus guidelines for neuroendocrine neoplasms of the appendix (excluding goblet cell carcinomas). Neuroendocrinology. 2016
- De Lambert G, Lardy H, Martelli H, Orbach D, Gauthier F, Guérin F. Surgical management of neuroendocrine tumors of the appendix in children and adolescents: a retrospective French multicenter study of 114 cases. Pediatr Blood Cancer. 2016; 63: 598-603.

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