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Pathology Page

# Acute appendicitis associated with an appendiceal carcinoid tumor

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A 28-year-old man presented with acute periumbilical pain for 1 day. The pain had started in the periumbilical region and then shifted to the right lower quadrant with intermittent abdominal cramps. He visited the hospital and was admitted with the suspicion of acute appendicitis. He was afebrile and hemodynamically stable. No rebound tenderness or abdominal guarding was noted during the physical examination. The appendix could not be visualized by ultrasonography. A computed tomographic scan showed an edematous and dilated appendix with fecolith and one nodular lesion over the proximal part of the appendix (Fig. 1A, arrow). The patient underwent appendectomy, and the gross picture of the appendix showed a white solid mass (1.2 cm in greatest diameter; Fig. 1B, arrow). The postoperative histological examination (Figs. 1C and 2A–D) revealed a Grade 1 neuroendocrine tumor (NET G1; carcinoid).

Over the past 2 decades the term “carcinoid tumor” has commonly been used for well-differentiated neuroendocrine neoplasms that have a relatively benign prognosis. According to the newest World Health Organization nomenclature and classification of neuroendocrine neoplasms of the digestive system, a typical carcinoid is classified as a Grade 1 neuroendocrine tumor (NET G1). Appendiceal neuroendocrine tumors are rare and one of more unusual histopathological findings (about 0.37%) when examining appendectomy specimens. Nonetheless, they are the most common primary neoplasm of the appendix. The peak incidence is observed in the 3rd decade and 4th decade of life, but these tumors can occur in any age group, including children. In most of such cases, they are incidental findings, but they have also been found to be associated with acute appendicitis, probably as a result of luminal obstruction.

Carcinoid tumors tend to be small and asymptomatic, and therefore they are rarely seen on imaging. Luckily, when our patient presented, it was possible to identify the problem preoperatively by computed tomographic scan. The postoperative histological examination revealed uniform cells with round nuclei that were growing in a trabecular or ribbon-like arrangement (Fig. 2A–C; hematoxylin and eosin 100–400×); the cells were immunohistochemically positive for synaptophysin (Fig. 2D), chromogranin A, and CD56. The Ki-67 proliferation index was <1%. Such cases have a relatively good 5-year survival rate (>90%), and the disease is most frequently seen in young adults, especially women, with a mean age of presentation of 42 years. For most patients, simple appendectomy is an adequate treatment. With this patient, the appendiceal carcinoid was an incidental finding and was probably associated with the acute episode of abdominal discomfort. He was discharged 3 days after the operation and remains well after 34 months of clinical follow-up.

Conflicts of interest: none.

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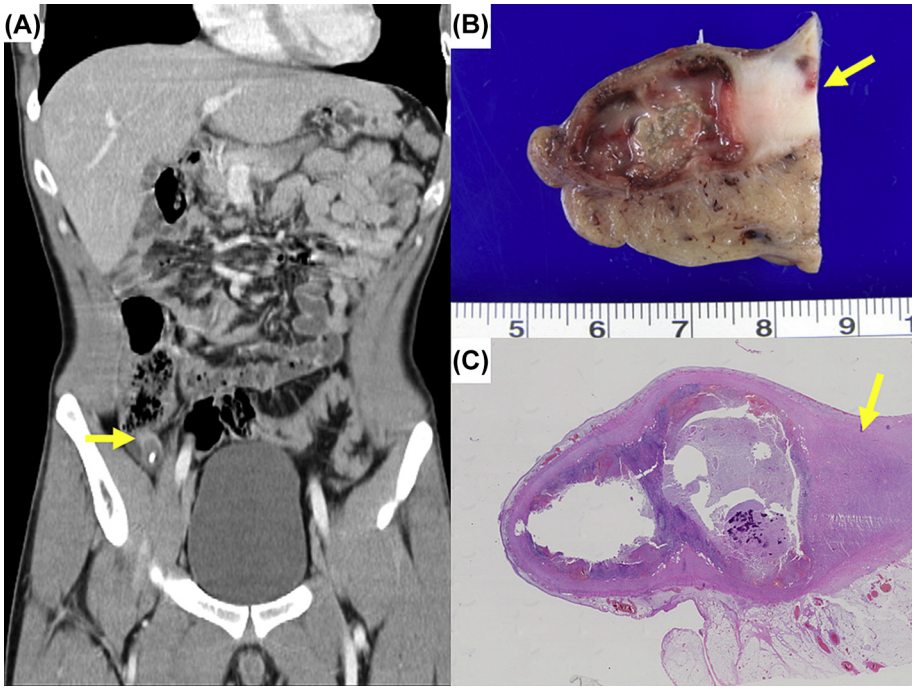


**Fig. 1.** (A) Computed tomographic scan showing an edematous and dilated appendix with fecolith and one nodular lesion over the proximal part of appendix (arrow). (B) The gross picture of the appendix shows a white solid mass (diameter being 1.2 cm in its greatest diameter; arrow). (C) Microscopic examination showing the tumor (arrow; hematoxylin and eosin 4×).

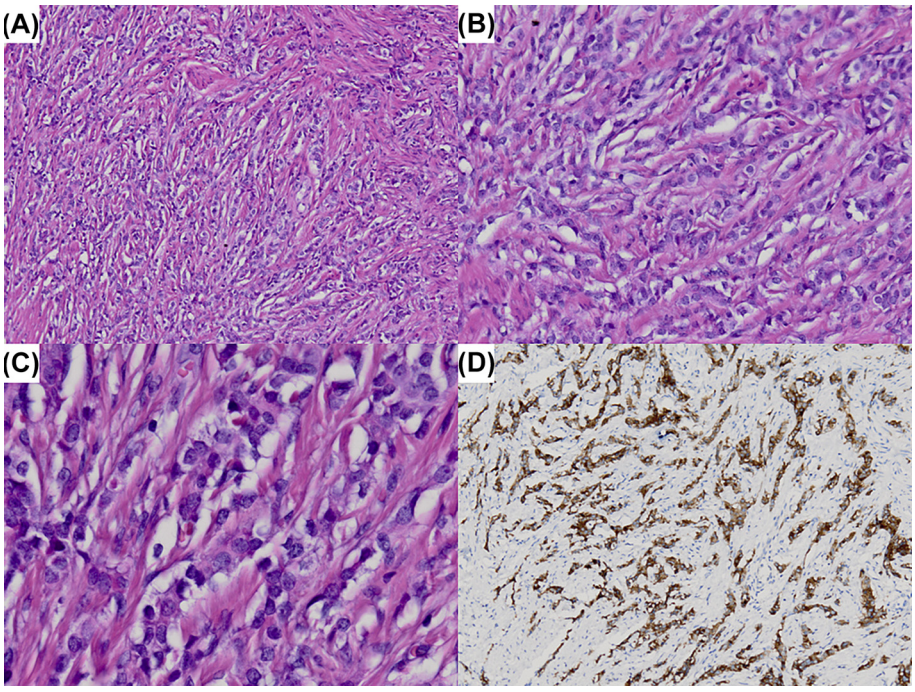


**Fig. 2.** (A, B) Microscopically, the appendiceal tumor revealed diffuse infiltration of uniform cells with round nuclei that grew in a trabecular or ribbon-like arrangement (hematoxylin and eosin, 100× and 200×, respectively). (C) Cytomorphology of the neoplastic cells revealed uniform nuclei, salt-and-pepper chromatin, finely granular cytoplasm, and rare mitotic events (hematoxylin and eosin, 400×). (D) Immunohistochemically, the neoplastic cells were found to be diffusely positive for synaptophysin, which highlights their ribbon-like arrangements (synaptophysin, 100×).

## Further reading

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