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A rare case of carcinoid tumor in a tailgut cyst

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Tailgut cysts are rare congenital lesions that arise from the failure of regression of the embryological tailgut. We report a case of neoplastic transformation of tailgut cyst to carcinoid tumor which is exceedingly uncommon.

Keywords: *carcinoid tumor*; *synaptophysin*; *neuron-specific enolase*; *chromogranin A*; *tailgut cyst*

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A 74-year-old male was found to have a large incidental cystic presacral lesion on computerized tomography (CT) of the abdomen. He underwent an elective transperineal resection with coccygectomy. Histologic examination showed a cystic structure with regions demonstrating a simple epithelial lining (Fig. 1) interlaced with areas characterized by intestinal-type epithelium with mucinous elements (Fig. 2). A 0.5 cm focus of glandular proliferation imparting an organoid appearance was seen (Fig. 3), with cells showing diffuse reactivity for markers of neuroendocrine differentiation, namely synaptophysin (Fig. 4) and neuron-specific enolase (Fig. 5), by immunohistochemistry. No reactivity for chromogranin was observed (Fig. 6). The morphological features, in correlation with immunophenotype, were consistent with carcinoid tumor arising within a tailgut cyst. Serum chromogranin was elevated at 271 ng/ml (normal < 93 ng/ml). Five years post-operatively, the patient remains asymptomatic with no evidence of recurrence on the most recent CT scan.

Tailgut cysts, also known as retrorectal cystic hamartomas, are rare congenital lesions that arise from failure of regression of the embryological tailgut (1, 2). Because of non-specific perianal symptoms, diagnosis can often be delayed (1, 3). Moreover, more than half of these cases may be asymptomatic and discovered incidentally (1). CT scan usually shows a well-marginated, presacral mass with water or soft-tissue density, but magnetic resonance

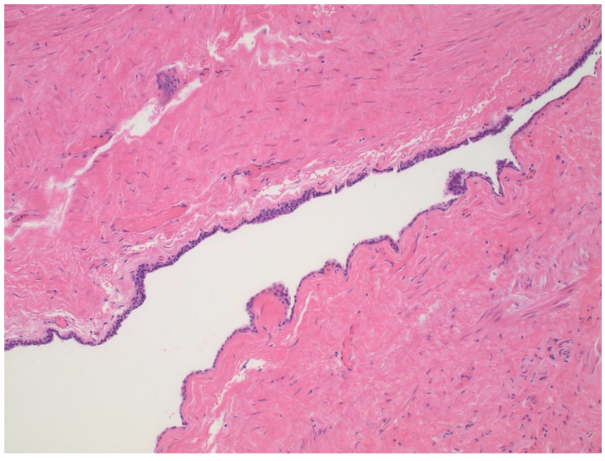

Fig. 1. A cystic structure within fibromuscular tissue showing a simple epithelial lining (H&E stain, 100 × original magnification).

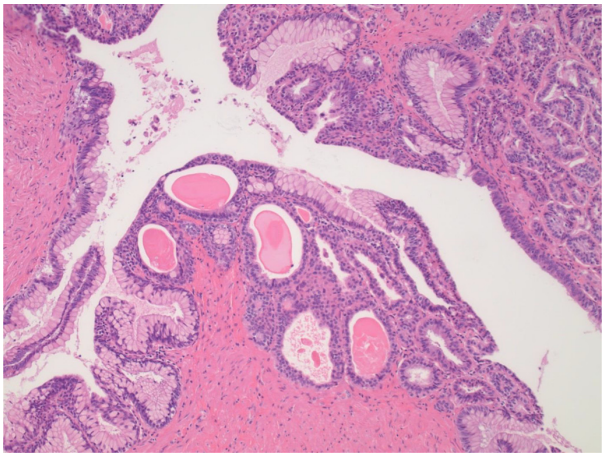

Fig. 2. Some areas within the cyst show intestinal-type glandular elements, consistent with a tailgut cyst (H&E stain, 100 × original magnification).

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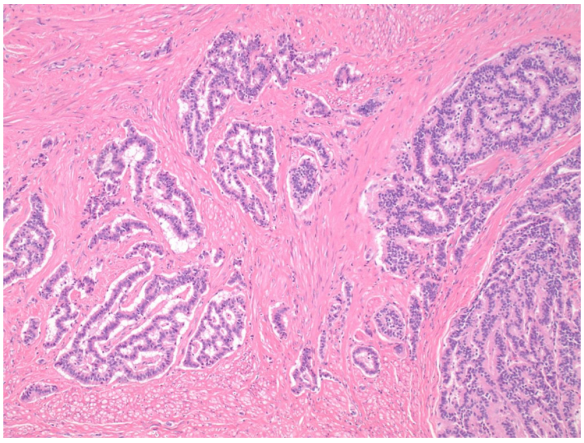

Fig. 3. Within the cyst, there is a nested proliferation of glands imparting an organoid appearance, with cells showing a stippled chromatin pattern (H&E stain, 100 × original magnification).

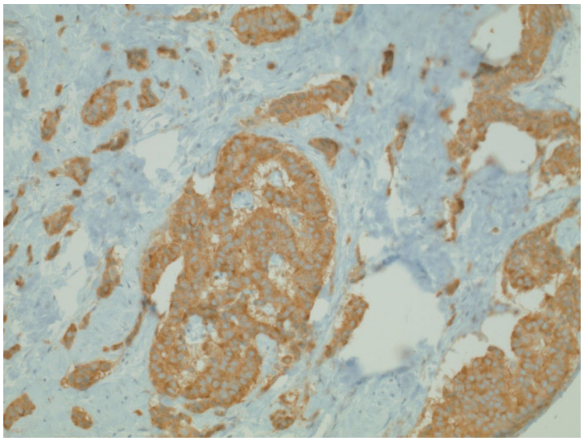

Fig. 4. Synaptophysin immunohistochemistry, showing diffuse reactivity within the nested cellular proliferation, supporting neuroendocrine differentiation (100 × original magnification).

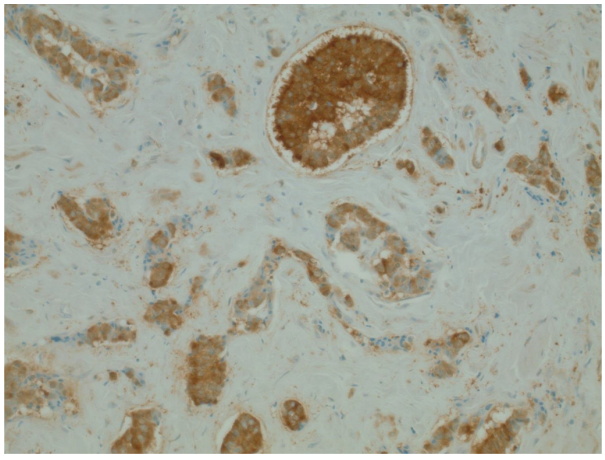

Fig. 5. Diffuse immunoreactivity for neuron-specific enolase within the nested cellular proliferation, also serving as evidence of neuroendocrine differentiation (100 × original magnification).

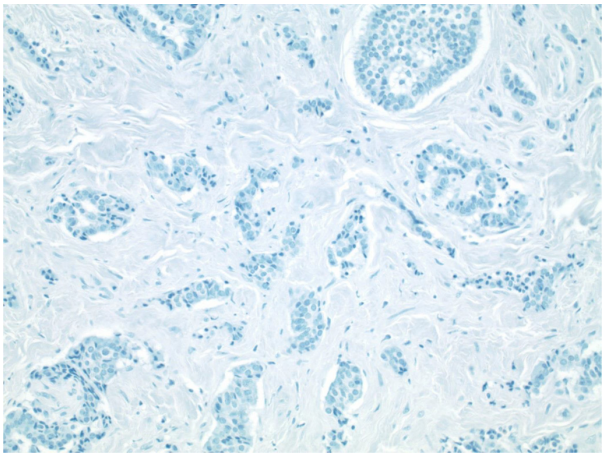

Fig. 6. There is absence of immunoreactivity for chromogranin (100 × original magnification), although the serum chromogranin level was elevated.

imaging helps in better visualization of the morphology (1). Pathologically, these cysts are lined by various types of epithelia (3). Neoplastic transformation of tailgut cysts to carcinoid tumors is rare, with only 17 cases (four males) reported through 2014 (4). Surgical excision is the definite treatment; coccygectomy is generally recommended to decrease the chances of recurrence, which may be as high as 16% (1, 3).

Conflict of interest and funding

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