

Neuroendocrine tumor

- [Left Renal Vein Transection and Reconstruction for Functional Paraganglioma Resection: A Case Report](#)
- [Electrochemical Signal-Off Competitive Immunoassay of Chromogranin A toward a Sandwiched Graphene Oxide Structure for Neuroendocrine Tumor Detection](#)
- [All-cause mortality in patients with medullary thyroid carcinoma of different ages: an inverse L-curve analysis study](#)
- [Consensus guideline for the management of peritoneal metastases from neuroendocrine neoplasms](#)
- [Cholecystokinin: Clinical aspects of the new biology](#)
- [A Neuropsychiatric Prelude to Unveiling Small Cell Lung Cancer with Suspected Paraneoplastic Limbic Encephalitis: A Case Report](#)
- [Consensus guideline for the management of patients with appendiceal tumors, part 2: Appendiceal tumors with peritoneal involvement](#)
- [Downregulated ALDH2 Contributes to Tumor Progression and Targeted Therapy Resistance in Human Metastatic Melanoma Cells](#)

[Small-cell lung cancer](#) AKA [Oat cell carcinoma](#) is a neuroendocrine [tumor](#).

There have been significant developments in diagnostic and therapeutic options for patients with neuroendocrine tumors (NETs). Key phase 3 studies include the CLARINET trial, which evaluated lanreotide in patients with nonfunctioning enteropancreatic NETs; the RADIANT-2 and RADIANT-4 studies, which evaluated everolimus in functioning and nonfunctioning NETs of the gastrointestinal tract and lungs; the TELESTAR study, which evaluated telotristat ethyl in patients with refractory carcinoid syndrome; and the NETTER-1 trial, which evaluated Lu-DOTATATE in NETs of the small intestine and proximal colon (midgut). Based on these and other advances, the North American Neuroendocrine Tumor Society convened a multidisciplinary panel of experts with the goal of updating consensus-based guidelines for evaluation and treatment of midgut NETs. The medical aspects of these guidelines (focusing on systemic treatment, nonsurgical liver-directed therapy, and postoperative surveillance) are summarized in this article. Surgical guidelines are described in a companion article ¹⁾.

From [tailgut cysts](#) are rare; only 15 cases have been reported until now. A tailgut cyst with [tethered spinal cord](#) has not been previously reported, although both diseases are congenital anomalies in the early stage of gestation.

A 53-year-old man, who presented with gluteal pain and bladder dysfunction. MR images showed that a tumor of the sacral spinal canal extended into the retrorectal space and connected to a thickened fatty filum terminale, which was tethering the spinal cord.

Because of tumor malignancy on a CT-guided biopsy and the imaging data for involvement of presacral lymph nodes on imagings, Mitsuyama et al. performed total removal of the tumor. Pathological examination revealed neuroendocrine tumor (Grade 2) arising from a tailgut cyst. The patient received somatostatin analogue therapy after surgery, followed by local radiation because of the further enlargement of the lymph nodes. Later, the authors started everolimus therapy for the metastases to the retroperitoneal lymph nodes. He presented with no local recurrence or further disease progression at 28 months after surgery. The review indicated that tumors in grade 2 or 3

showed progressive clinical course after surgery and three of seven patients with biopsy were misdiagnosed.

The correct preoperative diagnosis of neuroendocrine tumors from tailgut cysts is difficult, but extremely important because grade 2 or 3 tumors show disease progression even after surgery. Presacral congenital tumors, such as tailgut cysts, have the potential of malignant transformation into neuroendocrine tumors or adenocarcinomas. Comorbidity of spinal cord tethering and tailgut cyst suggests some relationship to common developmental errors in embryogenesis ²⁾.

Pituitary neuroendocrine tumor

see [Pituitary neuroendocrine tumor](#).

¹⁾

Strosberg JR, Halfdanarson TR, Bellizzi AM, Chan JA, Dillon JS, Heaney AP, Kunz PL, O'Dorisio TM, Salem R, Segelov E, Howe JR, Pommier RF, Brendtro K, Bashir MA, Singh S, Soulen MC, Tang L, Zacks JS, Yao JC, Bergsland EK. The North American Neuroendocrine Tumor Society Consensus Guidelines for Surveillance and Medical Management of Midgut Neuroendocrine Tumors. *Pancreas*. 2017 Jul;46(6):707-714. doi: 10.1097/MPA.0000000000000850. PubMed PMID: 28609356.

²⁾

Mitsuyama T, Kubota M, Nakamura Y, Yuzurihara M, Hoshi K, Okada Y. Neuroendocrine Tumor Arising from Tailgut Cyst with Spinal Cord Tethering: Case Report and Literature Review. *Spine J*. 2014 Oct 8. pii: S1529-9430(14)01552-6. doi: 10.1016/j.spinee.2014.09.027. [Epub ahead of print] PubMed PMID: 25305642.

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