Statement of Editorial Purpose

The Hospital Physician Oncology Board Review Manual is a study guide for fellows and practicing physicians preparing for board examinations in oncology. Each manual reviews a topic essential to the current practice of oncology.

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Management of Gastroenteropancreatic Neuroendocrine Tumors

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Note from the Publisher:
This publication has been developed without involvement of or review by the American Board of Internal Medicine.
Management of Gastroenteropancreatic Neuroendocrine Tumors

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INTRODUCTION

Neuroendocrine tumors (NETs) are a rare, heterogeneous group of neoplasms that arise from neuroendocrine cells located throughout the body. These tumors are characterized by variable but most often indolent biologic behavior. They are also classically characterized by their ability to secrete peptides, resulting in distinctive hormonal syndromes. Although NETs have been considered rare, recent studies suggest that they are more common than previously suspected. An analysis of the Surveillance, Epidemiology, and End Results (SEER) database demonstrated a significant increase in the incidence of NETs over time with an age-adjusted annual incidence in the United States of 5.25 cases per 100,000 population.¹ The increase in incidence is likely attributable to increasing awareness, improved diagnostic strategies, and possibly other undetermined environmental and genetic factors.

When NETs are diagnosed at an early stage, surgical resection is often curative. Unfortunately, curative surgery is rarely an option for patients with metastatic disease, and standard cytotoxic therapy for patients offers limited benefit. Treatment approaches with targeted therapy, including the use of agents targeting the vascular endothelial growth factor (VEGF) signaling pathway, the mammalian target of rapamycin (mTOR), and other pathways involved in neuroendocrine tumorigenesis, provide new therapeutic options for these patients. The aim of this review is to summarize advances in the diagnosis and management of well-differentiated, low-grade gastroenteropancreatic neuroendocrine tumors (GEP NETs). The management of poorly differentiated neuroendocrine carcinomas and mixed exocrine-endocrine tumors is beyond the scope of this review.

HISTOLOGIC CLASSIFICATION

NETs arising at different sites within the body are classified according to their histologic features. A number of histologic and anatomic classification systems have been proposed to describe these tumors (Table 1).²⁻⁴ Although there are differences in the specific criteria for grading tumors, the clas-