

Neuroendocrine Tumors

A Rare Finding: Part I

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Neuroendocrine tumors (NET) are very rare and many oncology nurses have never cared for a patient with the disease. NET arise from neuroendocrine cells dispersed throughout the body and are characterized by their ability to synthesize and secrete hormones and amines (Yao, 2007). NET can be subdivided into carcinoid and pancreatic islet cell tumors, which are difficult to distinguish by histology. Carcinoid tumors are derived from embryonic gut cells (Modlin, Lye, & Kidd, 2003) (see Figure 1). Although they can develop in any location, about 60% occur in the intestine and about 25% occur in the bronchopulmonary system. Islet cell tumors originate in the pancreas (Yao).

According to the Surveillance, Epidemiology, and End Results (SEER) 9 database, the incidence of carcinoid tumors, the most common form of NET, is about 3 cases per 100,000 per year and has increased in the last 30 years (Modlin et al., 2003; Talamonti, Stuart, & Yao, 2004). For carcinoid tumors, the average age at diagnosis is 61.4 years (Modlin et al.). Incidence varies with gender, age, and race. Slightly more women (55.1%) are diagnosed with carcinoid tumors; however, age-adjusted incidence rates are highest in African American men. Pancreatic islet cell tumors are much less common than carcinoids, and epidemiologic data are sparse. A population-based study by Yao et al. (2007) of the SEER 9, 13, and 17 databases indicated that the median age at diagnosis is 59 years. In the latest SEER database (SEER 17), the incidence of islet cell carcinomas is 0.12 per 100,000 and represents 1.3% of cancers arising in the pancreas, which is relatively rare as well.

Diagnosis

Although NET are characterized by the ability to secrete hormones and amines, hormonal syndromes resulting from this secretion usually occur only after the tumor has metastasized (Yao, 2007). At presentation, the signs and symptoms of NET are varied, making diagnosis difficult. Patients with carcinoid tumors may present with asymptomatic hepatomegaly or bowel obstruction or their tumors may be found incidentally at surgery for unrelated reasons (Talamonti et al., 2004). Patients also may present with symptoms such as flushing, diarrhea, and asthma (Talamonti et al.; Yao). Pancreatic islet cell tumors are similarly difficult to diagnose. They are classified as functioning (up to 50% of tumors) if they secrete peptides that cause systemic clinical symptoms; nonfunctioning if they do not. Patients with nonfunctioning tumors may present with pain or a mass causing biliary or bowel obstruction (Talamonti et al.).

Clinical Course

The clinical course of NET often is indolent, but the tumors also can be aggressive and resistant to therapy (Yao, 2007). NET usually are classified as low grade or high grade based on histologic differentiation, tumor size, angioinvasion, infiltrative growth, and presence

of metastasis. Clinical diagnosis is based on hormonal symptoms (if present); urinary 5-hydroxyindoleacetic acid (the breakdown product of serotonin); serum chromogranin A (a glycoprotein secreted by NET); and radiologic imaging, including OctreoScan® (Mallinckrodt Inc.) or positron-emission tomography scan (Robertson, Geiger, & Davis, 2006; Talamonti et al., 2004).

For localized primary tumors, surgical excision is the mainstay of therapy; NET are incurable in the metastatic setting (Talamonti et al., 2004). The liver is a frequent metastatic site. An estimated 50%–90% of patients with NET will develop liver metastases, and 80% of patients with advanced liver disease will die within five years of diagnosis. Liver-directed therapies, such as chemoembolization and radiofrequency ablation, are used to treat liver metastases (Talamonti et al.).

Hormonal Symptoms and Management

In the metastatic setting, carcinoids can secrete serotonin and other bioactive amines, causing diarrhea, carcinoid syndrome (diarrhea, flushing, and, less frequently, wheezing), carcinoid crisis (severe flushing and diarrhea leading to dehydration, hypotension, and arrhythmias), and carcinoid heart disease (a late complication) (Yao, 2007). Functional

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Digital Object Identifier: 10.1188/09.CJON.21-23