Neuroendocrine Tumors (NETs) and Gastroenteropancreatic Neuroendocrine Tumors (GEP-NETs) Disease Backgrounder

Neuroendocrine tumors, also known as NETs, are a rare group of tumors originating in the neuroendocrine cells of numerous organs including, but not limited to the lungs, gastrointestinal tract (including the stomach, small intestine, appendix, colon, and rectum), and pancreas. The term neuroendocrine refers to the dual features of these cells which are a cross between nerve cells and hormone-producing endocrine cells.

The estimated incidence, or rate of new cases of NETs in the United States is approximately 6.98/100,000 per year, while the estimated prevalence for 2014, based on the National Cancer Institute's Surveillance, Epidemiology, and End Results (SEER) database, was 171,321.[1] Even though NETs are considered to be rare (orphan disease), their incidence has grown over 500% over the last 3 decades.[1,2,3,4] This increase is probably due to improved diagnostic tools and disease awareness.

Gastroenteropancreatic neuroendocrine tumors (GEP-NETs) are subdivided into two categories: tumors of the gastrointestinal (GI) tract and pancreatic NETs. Characteristics of GI NETs can also vary, depending on where in the GI tract they are located (foregut, midgut or hindgut).

Some patients with NETs develop symptoms arising from the excessive production of hormones by neuroendocrine tumor cells. Common symptoms include diarrhea, abdominal pain, flushing, fatigue, wheezing, skin irritation, shortness of breath, and heart problems. Symptomatology can vary widely, however, complicating accurate diagnosis. NETs are generally slow growing, and in many cases, non-functioning (not secreting hormones). As a result, they can remain clinically silent in the early years of the disease process, often delaying the diagnosis in many patients.

Optimal management of NETs usually involves a multidisciplinary effort including medical, surgical, imaging and pathology specialties. Surgery is often the first line therapy for treating early-stage NETs. Many patients with NETs are diagnosed once metastases have already occurred, limiting the curative ability of surgical approaches. Survival of patients with advanced GEP-NETs depends on stage and histology. Patients with well- and moderately-differentiated distant metastases have a 5-year survival probability of 35%.[3]

There is a need for improved and effective treatment of inoperable, advanced NETs in patients who are progressive under somatostatin analogs. NET treatments have only recently benefited from therapeutic advances. Radiolabeled somatostatin analogs can target well differentiated gastroenteropancreatic NETs, relieving symptoms and suppressing tumor growth and spread.

References:

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[4] Frilling A, et al. Neuroendocrine tumor disease: an evolving landscape. *Endoc Related Cancer*. 2012, 19: R163-815.