Neuroendocrine Tumors (NETs) of Unknown Primary: Is Early Surgical Exploration and Aggressive Debulking Justifiable?

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Background: Neuroendocrine Tumors (NETs) are rare tumors that often present with vague symptoms. Identification and localization of the primary NET can be challenging and up to 10% of patients are classified as having an unknown primary. These patients have been thought to have a poor prognosis compared to those patients with a known primary. Treatments for patients with unknown primaries are directed towards symptom control and/or cytoreduction of metastatic disease. We hypothesized that early surgical exploration and discovery of a previously unknown primary will increase long-term survival.

Methods: The charts for all 342 surgical patients, seen in our clinic between 1/2009-9/2012 were retrospectively reviewed to determine which patients had a pre-operative diagnosis of a “NET with unknown primary”. Twenty-six patients were identified. For these patients, the rate of successful surgical exploration in which a primary site was identified were recorded. Clinical outcomes for these “unknown primary” patients were compared to historical survival data for patients with unknown primaries that did not undergo a surgical exploration in search of a primary.

Results: Twenty-six (26/354, 7%) NET patients with a pre-operative diagnosis of an unknown primary were explored and cytoreduced. The primary tumor site was identified in all 26 patients (100%). The primary sites identified for these patients were 23 ileal (88.4%) and 3 pancreatic (11.6%). All twenty-six patients were still alive as of 9/2012 therefore, a survival curve could not be generated.

Conclusions: Unknown primary NETs are not associated with a poor prognosis should timely surgical exploration and debulking result in an identification of the primary and a maximum cytoreduction. Early surgical exploration with aggressive debulking is indicated for the treatment of these patients.