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Neuroendocrine tumors of unknown primary site: gold dust or misdiagnosed neoplasms?

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ABSTRACT

Aims and background. Neuroendocrine tumors of an unknown primary site are rarer than other neuroendocrine tumors (0.6-2% of all neuroendocrine tumors) and have a poor prognosis. The aim of the study was to review the cases of unknown primary site neuroendocrine tumors encountered at the Istituto Nazionale Tumori of Milan between 1984 and 2008 in order to verify their incidence and evaluate their characteristics and prognosis.

Methods and study design. During the study period, 750 neuroendocrine tumor patients attended our Institute, 82 of whom (10.9%) were diagnosed as having neuroendocrine tumors of an unknown primary site. The data from their medical records were analyzed descriptively, and survival probabilities were calculated using the Kaplan-Meier method and the logrank test, considering patient, tumor and treatment-related characteristics.

Results. The 82 patients with neuroendocrine tumors of an unknown primary site (34 males) had a median age of 60 years; 57 (69.5%) had histologically well-differentiated tumors, 3 (3.7%) poorly differentiated tumors, and 22 (26.8%) had tumors that could not be classified. Of the 52 patients (62.2%) who underwent Octreoscan® (Bykgulden Italia SpA), 40 (78.4%) showed a pathological uptake and 11 (21.6%) were negative. Thirty-one patients (37.8%) underwent metastatic site surgery, which was radical in 11 cases (35.4%). Forty-eight patients (58.5%) received somatostatin analogues, and 41 (50.0%) underwent chemotherapy. At the end of the study period, 59 patients (72.0%) had died, 31 (53.0%) because of disease progression, and 23 (28.0%) were still alive.

Conclusions. Neuroendocrine tumors of an unknown primary site are difficult to identify but their incidence is higher than previously reported, and the prognosis remains unfavorable.

Key words: neuroendocrine tumors, Octreoscan, rare tumors, unknown primary site.

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