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Clinical characteristics of mixed neuroendocrine non-neuroendocrine neoplasm (MiNEN): a single institution case series

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BACKGROUND

Gastroenteropancreatic mixed neuroendocrine non-neuroendocrine neoplasm (MiNEN) is a rare entity consisting of two morphologically distinct populations of cells, one of which is neuroendocrine and one which is non-neuroendocrine. MiNENs typically contain high grade neuroendocrine carcinoma, conferring a poor prognosis. Due to the rarity of this diagnosis, data is limited and therefore, we sought to add to the current clinical understanding of this diagnosis.

METHODS

We conducted a single-institution, multisite survey of the electronic medical record using key search words to identify patients with a diagnosis of MiNEN with a pathological specimen reviewed within our institution. Data are presented descriptively, and recurrence free survival (RFS) and overall survival (OS) are reported using the Kaplan-Meier method.

RESULTS

Fifty-seven patients with a pathological diagnosis of MiNEN were included. Median age was 63.5 years, 60% were male, and 79% were white. The location of the primary malignancy included colorectal (CRC, 47%), esophagus/stomach (EGA, 18%), appendix, pancreas/ampulla (11% each), gallbladder, and small intestine (7% each). Fifty-one percent had stage 1-3 disease at diagnosis and 47% had stage 4 disease. Forty-six patients received systemic chemotherapy, including 12 in the neoadjuvant setting and 12 in the adjuvant setting as part of curative intent therapy, as well as 22 in the palliative setting. Twenty-eight patients underwent curative intent therapy with follow-up data available and manifested a median recurrence-free survival (mRFS) of 12 months. Patients with CRC had equivalent mRFS when compared to other primaries ($p=0.93$). Median overall survival (mOS) for the entire cohort was 22.0 months, with similar survival for CRC, EGA, and other primaries ($p=0.965$). Patients with synchronous metastatic disease had mOS of 13.0 months vs 53.0 months in stage 1-3 disease ($p=1.12e-05$). Patients who received first-line systemic therapy with platinum plus etoposide manifested a mOS of 23.0 months versus 22.0 months in those who received some other form of systemic therapy ($p=0.84$). However, 71% of patients receiving platinum plus etoposide did so in the palliative setting, whereas only 38% of those receiving some other chemotherapy did so in the palliative setting.

CONCLUSIONS

MiNEN remains a rare diagnosis, with poor prognosis. Primary location of the tumor was not predictive of survival, but metastatic disease conveyed a poorer prognosis. MiNENs are often underdiagnosed, and it is essential that oncologists and pathologists be aware of this rare entity.

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