Patients with neuroendocrine neoplasms: the experience of a referral center in Greece

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BACKGROUND

Neuroendocrine neoplasms (NENs) are rare and heterogeneous neoplasms with variable biological behavior but generally slow progression.

AIMS

Registration of demographics, clinical, imaging, pathological features, therapeutic options, response to treatment, overall survival of patients with NENs

METHODS

• 355 patients with NENs from our database registered during the period 2004-2014 have been retrospectively studied
• 166 females with mean age 52 years and range: 11-88
• TNM classification system has been used for staging
• Proliferation index Ki-67 has been used for grading
• Registered: primary site, presence of secretory/functional syndrome, metastatic deposits, presence of familiar syndrome
• Therapeutic management and outcome were registered

RESULTS

• 35 (10%) had neoplasms in the context of familiar disorders: MEN-1 (n=32) and Von Hippel Lindau (n=2).
• 54 (15.3%) had functional syndrome
• Metastatic deposits were found in 103 (29%) patients: bones: 16 (4.5%), lung: 5 (1.4%), liver: 94 (22.5%), peritoneum: 7 (2%), pancreatic: 2 (0.6%), brain: 2 (0.6%), omentum: 2 (0.6%), ovaries/ kidney/spleen: 1 (0.28%).

Initial diagnosis:
135 (38.8%) Stage 1
63 (18.1%) Stage 2
48 (13.8%) Stage 3
102 (29.3%) Stage 4

LUNG NEN
4 (1.4%) typical
10 (3.4%) atypical
1 (0.3%) small cell lung carcinoma
1 (0.3%) large cell lung carcinoma
1 (0.3%) atypical thymical NEN

GI-NENs Grading:
139 (47.8%) Ki-67≤2% (grade 1)
111 (31.3%) Ki-67:3-20% (grade 2)
24 (8.2%) Ki-67>20% (grade 3)

Line of treatment

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Molecular targeted therapy: everolimus, bevacizumab, sunitinib.
Chemoembolization: Transarterial Chemoembolization (TACE) or Transarterial Embolisation (TAE).

Patients with long term treatment, multiple therapeutic schemes and multiple therapeutic combinations

• In the last follow-up of the present analysis 21 patients died from their disease: 5 had grade1 neoplasm, 10 grade2, 2 grade3, 1 atypical thymic and another atypical lung NEN but 2 patients did not have an available Ki-67.

Conclusion

The present registry imply that the majority of the patients with NENs have slow progression and long survival despite the presence of disseminated disease, confirming the necessity of the recent introduced therapeutic and diagnostic options following the guidelines as well as their management from referral centers under multidisciplinary teams.

References