Patients with multiple endocrine neoplasia type 1 (MEN1) have late progression and long survival despite the presence of disseminated disease: the experience of a referral center in Greece

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BACKGROUND

Multiple endocrine neoplasia type 1 (MEN1) is a genetic disorder involving mainly parathyroid tumors, pancreatic neuroendocrine neoplasms (pNENs) and pituitary tumors

AIMS

Registration of demographic, clinical, imaging, pathological features, therapeutic options, response to treatment, overall survival of patients with MEN1

METHODS

35 patients with MEN1 from our data-base registered during the period 2004-2014 have been retrospectively studied

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

24 (68.6%) patients were the index cases.

19 (54.3%) had a positive gene mutation while 5 (14.3%) did not have the mutated gene.

11 (32.4%) patients had a functional syndrome.

Metastatic liver deposits were found in 6 (17.6%) patients.

Age 39 (15-64) years

Gender Males 20
Females 15

Syndrome components Primary hyperparathyroidism 32
Pituitary Adenoma NFPA 20
PRLoma 8
GH 1
Pancreatic Neuroendocrine Neoplasm 32
Adrenal Adenoma 14
Acarcinoma 1
Fibroma 3

Line of treatment

Surgery 14
Chemotherapy 6
Somatostatin receptor analogs 8
Peptide receptor radionuclide therapy 1
Chemoembolization 1
Follow up only 21

Molecular targeted therapy: everolimus, sunitinib.

Chemoembolization: Transarterial Chemoembolization (TACE) or Transarterial Embolisation (TAE).

27 patients are alive with mean follow-up time 94,64 months (range 10-316.33) since diagnosis

3 (9.67%) patients died of their disease out of the 31 under current follow-up with a mean survival time 121 months (range 62.3-190) since diagnosis:2 with grade 1 pNEN and 1 grade 2 pNEN-1 patients died during surgery (pulmonary embolism)

The present registry implies that the majority of the patients with MEN1 have late progression and long survival despite the presence of disseminated disease, confirming the necessity of specific therapeutic and diagnostic options following the guidelines as well as their management from referral centers under multidisciplinary teams.

References: Thakker et al. J Clin Endocrinol Metab. 2012 Sep;97(9):2990-3011

Conclusion

The present registry implies that the majority of the patients with MEN1 have late progression and long survival despite the presence of disseminated disease, confirming the necessity of specific therapeutic and diagnostic options following the guidelines as well as their management from referral centers under multidisciplinary teams.

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