

C2

Poor Prognosis in Spontaneous Gastrinoma When Additional Hormones and Peptides are Secreted

Joy Ardill¹; Brian Johnston¹; David McCance¹

¹NET Group, Royal Victoria Hospital Belfast and Queen's University Belfast, Northern Ireland, UK

Background: Gastrinoma (ZES) occurs in 0.5-3/million/year. ZES may be familial (MEN1) or spontaneous, benign or malignant. Tumors in patients with MEN1 or those that are spontaneous may produce other peptides and hormones in addition to gastrin.

Methods: We studied patients with ZES who attended our clinic. Clinical notes and laboratory records were examined.

Results: Fifty seven patients had ZES, 13 with MEN1 and 44 with spontaneous tumors. Of the spontaneous group 7 were benign and 37 malignant. Thirteen of those with malignant tumors later secreted additional hormones.

MEN1 patients presented with circulating gastrin 900 (125-3,200) ng/l, eight had tumours secreting other peptides and hormones including PTH, prolactin, insulin, glucagon, PP, somatostatin, VIP, calcitonin or NKA. Several patients had multi-hormone secretion. Median survival was 17 (3-34) years.

Patients with benign tumors presented with circulating gastrin 260 (130-1,050) ng/l all were cured by surgery. Survival in each individual was >17years.

Twenty four patients with metastatic ZES presented with circulating gastrin 500 (145-15,800) ng/l, and median survival was 8.2 (0.5-26) years. None of these patients secreted additional peptides with the exception of PP <2,500ng/l. (PP is secreted by many pancreatic NETs. PP secreted by PPoma is usually >10,000ng/l)

Thirteen patients with metastatic ZES presented with gastrin 750 (255-70,000) ng/l. All in this group later secreted additional hormones, PTH, prolactin, PP, insulin, glucagon, VIP, NKA, somatostatin or calcitonin. Several patients secreted 2 or 3 additional hormones. Median survival was 1.9 (0.1-9.9) years. No patient survived more than 2 years post multi-hormone secretion. Those producing ACTH, PP (>20,000ng/l) or insulin had poor survival.

Conclusions: A third of patients with spontaneous metastatic ZES later secrete additional hormones resulting in worse prognosis. ZES patients should therefore be screened regularly, using relevant biomarkers, making possible, earliest appropriate treatment intervention.