Sustained Response to Alpha Interferon in a Patient with an Advanced Metastatic Serotonin Secreting Endocrine Tumour - Case Report

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Case Report: This 52-year-old lady presented in 2001 at a GI clinic complaining of occasional abdominal cramps sometimes severe and prolonged. Symptoms were not associated with diarrhoea or constipation. Weight loss of 4.5Kg over 4 months was noted. Coeliac disease was excluded and a diagnosis of irritable bowel was made.

In January 2002 she represented with further weight loss (total 8Kg), cyclical symptoms of diarrhoea lasting 3-5 days and occasional flushing. Neuroendocrine tumour markers were measured. Urinary 5HIAA was grossly elevated at 637 (Reference Range RR10-47), 5HT 12.05 (RR 0.30-1.30), pancreastatin (PST) >1,000ng/L (RR 0-50) and neurokinin A (NKA), 350ng/L (RR 0-20). CT and Octreotide scintigraphy showed extensive hepatic metastases with para-aortic and iliac lymphadenopathy. No primary tumour was identified. The surgical team considered hepatic disease, inoperable.

Treatment with somatostatin analogues was commenced. Symptoms continued, urinary 5HIAA remained grossly elevated and PST and NKA rose dramatically (9300ng/L and 4500ng/L respectively). Somatostatin analogue dose was increased twice without improvement. Alpha interferon concomitant with somatostatin analogues, was commenced 1.5 MU 3 times weekly increasing to 9MU 3 times weekly. Within 2 months symptoms eased and this regimen was continued. By 6 months symptoms had abated. Urinary 5HIAA settled around the upper limit of normal and 5HT returned within RR. Circulating NKA was secured below 100ng/L within a year and was maintained thereafter at 40-80ng/L. Both PST and Chromogranin A remained >10 times RR. Radiology showed stable/reduced disease.

Due to symptoms of migraine and fatigue, interferon dose was reduced occasionally and the drug was withdrawn for short periods. This resulted in an immediate rise in NKA. In the summer of 2010 the patient decided to discontinue interferon and received three cycled of Yttrium 90 PRRT. She declined gradually and died in June 2011. Survival, post diagnosis was 10 years 5 months.