Atypical Location and Treatment for a Rare Neuroendocrine Tumor

H. El-Gayar, D. Morganstein & K. Meeran
Department of Investigated Medicine, Imperial college, Hammersmith hospital, London, UK

Somatostatinomas are rare neuroendocrine tumors of D cell origin usually arising from the pancreas and duodenum. Symptoms include hyperglycemia, cholelithiasis, diarrhea and steatorrhoea. Treatment with somatostatin analogue may appear paradoxical, but can lower somatostatin levels and improve symptoms.

Case: A 60yr old gentleman presented with diarrhoea and steatorrhoea, opening his bowels between 12-13 times a day causing him to stop working and become depressed. Repeated fasting gut hormone profiles showed an isolated elevated somatostatin level. CT abdomen did not show any pancreatic lesions and colonoscopy was normal. Octreotide scan revealed a well defined lesion in the base of the right lung, confirmed on a CT thorax. Visceral angiography with calcium stimulation and sampling from the femoral artery revealed elevated somatostatin in all samples (> 400pmol/l). Somatostatin levels taken from the right atrium and femoral artery, showed a higher level in the arteries (venous 152pmol/l, arterial 185pmol/l), suggesting a pulmonary source. However, the surgical risks of resection were thought to be to high. He had a trial of Octreotide therapy without benefit. Therefore he was treated with a combination of Lanreotide 30mg every 14days, and Octreotide 50mcg TDS, resulting in a dramatic improvement in symptoms. He now opens his bowels once or twice a day. His symptoms worsened when Octreotide was withdrawn. He also noticed two days prior to the lanreotide injection be opens his bowels more frequently.

Discussion: There are five subtypes of somatostatin receptors (SSTR). All five SSTRs bind to the natural somatostatin. Two different somatostatin analogues are used clinically, octreotide and lanreotide. These analogues bind principally to the receptor subtype 2 and 5. Although most studies have shown no major difference between octreotide and lanreotide in terms of receptor affinity or biological activity, this case illustrates that combination therapy may have clinical benefits.