A Case Report: To be or not to be—Carcinoid Syndrome?

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Introduction:
Carcinoid syndrome occurs as a result of a carcinoid tumor secreting a variety of peptides namely serotonin, bradykinins, histamine and substance P draining directly into the systemic circulation. These carcinoid tumors can metastasize to the liver or can be commonly seen in lungs as bronchial carcinoids. These tumors could originate anywhere from the foregut (bronchus, stomach, pancreas), midgut (small intestine and ascending colon) or hindgut (distal colon and rectum). The most common symptoms associated with carcinoid syndrome are flushing and diarrhea, 95% and 75%, respectively. The most useful initial diagnostic test to date to diagnose a carcinoid tumor is a 24-hour urinary excretion of 5-hydroxyindoleacetic acid (5-HIAA) followed by tumor localization via Octreotide Scan.

Case Presentation
A 47 year old male patient presents to the clinic for flushing, fatigue and intermittent night sweats over the past year. The initial symptoms were exacerbated by eating, not resolved by a gluten-free diet. He had mild resolution of his symptoms on low-carbohydrate foods but continued to have spontaneous flushing in the face and neck. Associated symptoms included abdominal distention, early satiety, 30 pound weight loss, dizziness, insomnia, arthralgias, congestion, palpitations, elevated blood pressure, generalized fatigue and intermittent night sweats over the past year. The initial treatment modalities such as arthroplasty or hepatic resection were sought; since the risks outweighed the benefits of therapy or tumor recurrence after excision. The only diagnosis has been confirmed; mostly to follow the response to therapy or tumor recurrence after excision. The next difficult interpretation was the lack of elevation in 5-HIAA, which is thought to be the highest in the setting of primary tumors of mid-gut (jejunoileal, appendiceal, ascending colon), which was absent in this patient.

The real question was whether this case was an early pre-clinical presentation, before the onset of the disease. As the patient presented with no serious clinical symptoms, no treatment modalities such as Somatostatin analogs or hepatic resection were sought; since the risks outweighed the benefits and the episodes of flushing were not particularly debilitating for the patient.

The remainder of the workup, which included a Celiac disease panel, plasma free metanephrines and normetanephrines, Vasoactive Intestinal Polypeptide, and somatostatin were negative.

Discussion
Although the most reliable biochemical diagnostic tool to use to diagnose Carcinoid disease are a 24-hour urinary excretion of 5-HIAA, a serum serotonin level and chromogranin A levels, although it has its caveats. Chromogranin A is more appropriately used to assess disease progression once a diagnosis has been confirmed; mostly to follow the response to therapy or tumor recurrence after excision. The only problem is that chromogranin A may be falsely elevated in patients on PPI therapy, which this patient was on. The next difficult interpretation was the lack of elevation in 5-HIAA, which is thought to be the highest in the setting of primary tumors of mid-gut, jejunoileal, appendiceal, ascending colon, which was absent in this patient.

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References

Figure 1. Whole body scan 24 hrs after administration of Indium-111 Pentreotide.

Figure 2. Hepatic lesion in posterior right hepatic lobe with increased uptake at 24 hours.

Figure 3. Two hepatic lesions in right hepatic dome with increased uptake at 24 hours.