

Introduction

- Carcinoid tumor is a well-differentiated neuroendocrine tumor (NET) that arises from enterochromaffin cells.
- NETs produce serotonin which is metabolized to the inactive 5-hydroxyindoleacetic acid (5-HIAA) by the liver and the lungs.
- Carcinoid syndrome is caused by secretion of serotonin into the systemic circulation and is usually caused by metastatic NET that originate from the midgut.
- Clinical features of carcinoid syndrome include episodic flushing, diarrhea, and in some cases right-sided valvular heart disease
- The primary screening method for carcinoid syndrome is by 24 hour urinary 5-HIAA (U-5HIAA) which has a 90 percent sensitivity and specificity in diagnosing carcinoid syndrome.

Case Presentation

- A 29-year-old female presented to our clinic with abdominal pain, bloating, diarrhea and weight loss associated with post prandial facial flushing.
- She reports around 9 lbs of weight loss
- Her medications at time included dextroamphetamine and fremanezumab.
- Based on her symptoms, U-5HIAA was ordered and was elevated at 148 mg (normal <6)
- Carcinoid syndrome was suspected based on symptoms and markedly elevated U-5HIAA. Imaging studies ordered to identify primary or metastatic disease
- CT abdomen/pelvis was unremarkable and showed no abdominal mass or adenopathy
- Ga68 Dotatate (NETSPOT) PET was ordered and revealed no tracer avid malignancy.
- Upper endoscopy and Colonoscopy were also negative.

Urine-5HIAA Levels

Initial Labs	3 days after stopping dextroamphetamine
148 mg	10 mg

Figure 1. Illustrates the down trend of U-5HIAA levels after cessation of dextroamphetamine

Management

- At this time, carcinoid syndrome was less likely given negative imaging and colonoscopy despite elevated U-5HIAA
- Additional etiologies for elevated U-5HIAA were considered.
- The patient was asked to discontinue her dextroamphetamine and recheck her U-5HIAA
- The patient expressed difficulty in stopping dextroamphetamine for the urine collection but due to the negative evaluation for NET she finally agreed to hold it for 3 days.
- Repeat U-5HIAA was 10.4 mg (down from 148 mg)
- Thus, her symptoms were then believed to be functional bowel disease and the elevated U-5HIAA a result of her dextroamphetamine use.
- Her symptoms improved with Mirtazapine and Dicyclomine and she regained her weight

Discussion

- This is a unique case with a misleading U-5HIAA. While mildly elevated levels of U-5HIAA can be seen with tryptophan rich foods and certain drugs including amphetamines, marked elevation like in our case is more specific and usually seen with the carcinoid syndrome.
- Our patient had symptoms suspicious of carcinoid syndrome and a U-5HIAA of 148 mg which is 25 times the upper normal. However, there was no evidence for NET on extensive work up and the levels rapidly decreased with stopping dextroamphetamine
- To our knowledge, this is the highest falsely positive U-5HIAA reported.
- This case underscores the importance of a complete drug history including over the counter medications and that when the 5-HIAA is elevated the test need to be repeated after discontinuing all potentially interfering medications, although may be difficult with addictive medications like in our case