



A 70 year old female patient is admitted to the surgery department of a tertiary care hospital for protracted diarrhea. As the intern on duty you take the history of the patient which reveals episodes of flushing of the face with difficulty in breathing, which is often exaggerated when she gets stressed. Your surgery professor attends to the patient and immediately points to the patient's neck (shown in the picture) and clinches the diagnosis. After which your professor instructs you to order a very specific urine test.

1. What is your provisional diagnosis?

Answer- Carcinoid Syndrome or Carcinoid heart disease.

Carcinoid tumors arise from neuroendocrine organs(eg-Pancreas) or neuroendocrine-differentiated gastrointestinal tract, and more than 40% occur in the small intestine.

The carcinoid tumor when localized to the small intestine releases vasoactive substances such as serotonin , bradykinin , kallikrein, histamines, prostaglandins and tachykinins. However these mediators are carried to the liver via the portal vein where they are metabolized via the "first pass effect" similar to oral drugs.Now the issue occurs when the carcinoid tumor metastasizes to the liver.Now the vasoactive substances released by the tumor can no longer be metabolized by the liver leading to systemic manifestations once these substances enter the hepatic vein.

The vasoactive substances cause the following clinical manifestations such as cutaneous flushing,sweating,brochospasm (difficulty breathing) and diarrhea.Symptoms are exaggerated on consumption of alcohol and emotional stress.

Now these vasoactive substances on entering systemic circulation finally enter the right side of the heart leading to valvular fibrosis resulting in tricuspid insufficiency.The clinical sign given in the image is engorged neck veins due to tricuspid insufficiency.The left side of the heart is not affected as enzymes present in the lung metabolizes the substances before it reaches the left side of the heart.

2.What specific urine test did the professor order?

Answer- 24 hour urinary collection for 5-hydroxy-indoleacetic acid which is a breakdown product of serotonin.

3.How will you treat this patient?

Answer-For symptomatic management of flushing and diarrhea octreotide can be used.Imaging studies such as octreotide scan and CT scan can be used to elaborate the extent of the disease.This is followed by surgical resection of the tumor.

