Carcinoid Syndrome

Carcinoid tumors are slow-growing tumors derived from neuroendocrine cells, typically found in the GI tract. These tumors secrete vasoactive substances like serotonin and kallikrein, and are the most common malignancy of the appendix. The most common originating sites include the ileum, rectum and stomach. These tumors can be identified via detection of chromogranin A peptides and neuron enolase stain, and visualization of dense core bodies on electron microscope. While these tumors are in the GI tract, they are mainly asymptomatic due to hepatic degradation of vasoactive substances. Carcinoid syndrome occurs in approximately 10% of carcinoid tumors and occurs when vasoactive substances produced by the carcinoid tumor enter systemic circulation. This typically occurs when carcinoid tumors metastasize to the liver as they bypass hepatic degradation. Symptoms of carcinoid syndrome include cutaneous flushing, wheezing, right sided heart murmurs, and diarrhea.

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Pathophysiology

Neuroendocrine Cells
Neuron-indy-car
Carcinoid tumors are slow-growing tumors derived from neuroendocrine cells, typically found in the GI tract. These neuroendocrine cells receive neural input and can release hormones into systemic circulation as a response.

Serotonin
Silver-tonic
Carcinoid tumors typically produce serotonin (5-HT), which is a monoamine neurotransmitter. In the intestines, serotonin is produced by enterochromaffin cells of the gut, where it plays a role in the regulation of intestinal peristalsis, among other activities.

Appendix
Appendix-pen
While carcinoid tumors remain quite rare, they are the most common cancer of the appendix, accounting for over half of all appendiceal malignancies. However, the most common originating site is the small bowel, specifically the ileum. Both the rectum and stomach are also sites of carcinoid tumor genesis. Since the introduction of widespread colonoscopy screening protocols around the year 2000, rectal carcinoid tumors have been diagnosed at the highest frequency of the locations listed.

Diagnosis

Chromogranin A
Chrome-grandma (A) Apple
Chromogranin A is a protein structure related to neuroendocrine secretory proteins. It is measured as a status indicator for carcinoid tumors and associated carcinoid syndrome. It is also elevated in pheochromocytomas and pancreatic cancer.

Neuron Specific Enolase
Neuron Emo-lace
Neuron-specific enolase, or NSE, is a protein present in high concentrations in neurons and neuroendocrine cells. Because carcinoid tumors derive from neuroendocrine cells, serum concentrations of neuron-specific enolase are used as a means of tracking tumor progression or digression, response to treatment, and recurrence.

Dense Core Bodies on EM
Apple Core Body
Neuroendocrine cells contain dense core bodies, which are neurosecretory granules. Visualization of dense core bodies on an electron microscope is characteristic of carcinoid tumors.

Signs and Symptoms

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Symptoms When Metastasize to Liver

Giant Liver

Carcinoid syndrome occurs in approximately 10% of carcinoid tumors and presents when vasoactive substances, primarily serotonin and kallikrein produced by the carcinoid tumor, enter systemic circulation. This presentation typically takes place when carcinoid tumors metastasize to the liver because the liver usually degrades these substances from the portal circulation, preventing introduction to systemic circulation.

Flushing

Flashlight

Flushing of the skin, most often of the head and upper thorax, is the most important clinical finding of carcinoid syndrome. Flushing is caused by the secretion of kallikrein, which plays a role in the conversion of kininogen to bradykinin, a powerful vasodilator. This vasodilation of the superficial blood vessels results in flushing.

Wheezing

Weasel

Wheezing caused by bronchoconstriction is a common sign of carcinoid syndrome. The exact pathogenesis of bronchoconstriction is unknown but is thought to be caused by excess serotonin.

Right Heart Murmur

Writing-on-heart Merman

Carcinoid syndrome is associated with fibrotic lesions on the cardiac valves and endocardium. These typically affect the right side of the heart as the right heart receives blood from systemic circulation. This is because serotonin is broken down by monoamine oxidase, which is found in the lungs (along with the liver), with metabolism occurring before active serotonin arrives at the left heart.

Diarrhea

Toilet

Carcinoid syndrome typically causes secretory diarrhea with abdominal cramping. This symptom can quickly become severe and cause dangerous electrolyte imbalances coupled with dehydration.

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