Focus on Carcinoid Tumors

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Definition
Carcinoid is a collective term for tumors with similar pathologic characteristics. They are neuroendocrine tumors that secrete various hormones. Although they are not as aggressive as adenocarcinoma, lesions can vary from benign to metastatic. Tumors are found primarily in the respiratory tract and gastrointestinal system. Other possible carcinoid tumor sites include the gynecologic system, biliary tract, and head and neck region.

Incidence
A. Overall incidence has been rising since 1973.
B. Analysis of the National Cancer Institute Surveillance, Epidemiology, and End Results program found an incidence of 38.5 per million.
C. Average age: 60.9 years
D. Risk factors and etiologies
   1. Gender: Both are almost equally affected, females slightly more than males.
   2. Age: generally older than 50 years
   3. Race: No characteristic feature exists, although African American patients have a higher incidence and poorer survival rate.

Normal Physiology
A. Found in the enterochromaffin cells, which are located along the gastrointestinal mucosa
B. Enterochromaffin or neuroendocrine cells are part of the amine precursor uptake and decarboxylation (also known as APUD) system. These cells are a part of a group of cells that are able to secrete hormones.

Pathophysiology
A. A lesion has a yellow to grayish-white appearance, may be ulcerated, and can occur in clusters.
B. Neurosecretory granules are a part of the cell structure that secretes hormones.
C. Hormones secreted by the tumor include serotonin, gastrin, histamine, tachykinin, bradykinin, and adrenocorticotropic hormone.
   1. Serotonin produces diarrhea, flushing, bronchospasm, and cardiac lesions.
   2. Tachykinins are vasodilators, which increase cardiac output. Examples of tachykinins include substance P, neurokinin A, neurokinin B, and neuropeptide K.
   3. Bradykinin is part of the inflammatory process and causes flushing.

Signs and Symptoms
A. Abdominal pain is the most frequent complaint.
B. Nausea and vomiting
C. Bowel obstruction: pain and distention
D. Carcinoid syndrome occurs in less than 10% of those with carcinoid tumors.
   1. Symptoms of carcinoid syndrome include flushing, diarrhea, wheezing, dyspnea, and hypotension.

History and Physical Examination
A. Findings are classified by site of origin.
   1. Respiratory tract: flushing, pulmonary edema, Cushing syndrome, or hemoptysis
   2. Stomach: peptic ulcer disease, abdominal pain, bleeding, epigastric pain, early satiety, bloating, and dysphagia
   3. Ileum: abdominal pain and, rarely, pellagra, which is a skin condition caused by niacin deficiency. Carcinoid syndrome is associated with this site of origin when hepatic metastases are present.
   4. Colon and rectum: pain, weight loss, and anorexia
B. Diagnostic tests involve laboratory and radiology. Blood and urine samples are used to establish diagnosis and monitor tumor activity. Frequency of testing depends on patient status.
   1. 5-HIAA (5-hydroxyindoleacetic acid): urine test used to diagnose and follow tumors. Levels greater than 30 mg in a 24-hour urine sample indicate carcinoid syndrome.
   2. 5-HTP (5-hydroxytryptophan): a serum measure of serotonin level used to monitor carcinoid syndrome

Assessment
A. The majority of patients are asymptomatic, with tumors found incidentally or on autopsy.
   1. Examine lung sounds if patient reports recurrent cough, hemoptysis, or chest pain.
   2. Examine abdomen for chronic weight loss, anorexia, abdominal pain, and anemia.

Key Words: malignant carcinoid syndrome, somatostatin

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