Carcinoid Syndrome: a Rare and Complex Disease

by PAULA CLARK

Understanding Carcinoid Syndrome

It’s common knowledge that cancer tumors can occur in different shapes and sizes. What’s less known is how they can travel at different speeds.

Carcinoid tumors have been called "cancers in slow motion". They fall somewhere between a benign, non-threatening growth and a metastatic, life-threatening growth.

While the small, slow-growing neuroendocrine tumors that occur throughout the body don’t always cause symptoms, they can cause a rare condition called carcinoid syndrome.

What Is Carcinoid Syndrome?

Carcinoid syndrome is caused by the hormones secreted by carcinoid tumors.

Most carcinoid tumors are found in the gastrointestinal tract including the small intestine, appendix, and rectum. They can also spread to the liver, lungs, bones, skin and other organs. Only 10% of carcinoid tumors start in the lungs. They’re called bronchial or airway tumors.

There’s a benefit to having carcinoid tumors in the gastrointestinal tract. The hormones get broken down, so they don’t cause any problems.

When the tumors spread to organs outside the intestine (usually the liver, and occasionally an ovary) the hormones are no longer broken down. That’s when carcinoid syndrome can occur.

The good news is having carcinoid tumors doesn’t mean you’ll automatically develop carcinoid syndrome. But since the tumors can be present without symptoms, they can be difficult to find and treat before carcinoid syndrome develops.

So, what causes carcinoid syndrome? No one knows for sure what the underlying cause of carcinoid tumors, or how carcinoid syndrome is triggered.

Some studies point to risk factors like smoking and diet, but more research is needed to fully understand it.

Who Is Affected?

Carcinoid tumors occur rarely and only 10% of people with the tumors will develop carcinoid syndrome.

Males and females are affected equally as are most races, save for a slight increase in numbers among black African males. All ages are affected, but they occur most often among the middle-aged.

Warning Signs and Symptoms

Carcinoid tumors can be present for years with no symptoms, which are also easy to mistake for other conditions like irritable bowel syndrome.

The most common symptoms of carcinoid syndrome are facial flushing (having a hot flushed face), facial skin lesions, diarrhea, and wheezing.

Complications

As carcinoid syndrome becomes more serious, heart problems can arise as a rapid heartbeat, low blood pressure, heart murmur, shortness of breath with activity, and swelling in the legs and feet. Carcinoid heart disease occurs in more than 50% of patients with carcinoid syndrome.

Since the hormones released by carcinoid tumors can disrupt different parts of the endocrine system, carcinoid syndrome may lead to other conditions:

- The hormones can signal the adrenal glands to produce too much cortisol, causing Cushing's Syndrome.
- Pellagra happens when the tumors use tryptophan to make serotonin instead of niacin.
- Zollinger-Ellison syndrome occurs when the tumors release a hormone that signals the stomach to make too much acid and may lead to stomach ulcers.

Finally, a carcinoid crisis can happen when carcinoid tumors release an overwhelming amount of hormones. This can be triggered by anesthesia during surgery. According to WebMD "irregular and life-threatening heart rhythms, severe increases or drops in blood pressure, extreme difficulty in breathing, and delirium can happen during these episodes".

Diagnosis

Carcinoid syndrome is a challenge to prevent because finding the tumors in the early stages is difficult.

Many people live with the tumors their entire life with no symptoms or complications, and once carcinoid syndrome has developed, its symptoms mimic other disorders which makes diagnosis a challenge.

That’s why practitioners need to approach diagnosis from several angles. This includes taking a patient’s health history, tissue biopsies, lab tests, and nuclear imaging.

Treatment Options

Fortunately, there are ways to treat and manage carcinoid syndrome.

Surgery can be performed to remove the tumors and reduce the spread. The type of surgery required depends on where the tumors are located.

New approaches to reducing metastases in the liver include cryoablation (using cycles of freezing and thawing to kill cancer cells) and radiofrequency ablation (using electrical energy and heat to kill cancer cells).

Hepatic artery catheterization (which means blocking the artery feeding liver tumors with inert particles) is also an option. The particles can be used on their own or mixed with chemotherapy drugs.

Using chemotherapy systemically is another option and benefits about one third of patients.

Once the tumors have abated ongoing management is needed to control symptoms.

The drug Octreotide (Sandostatin) is used most often and can be administered multiple ways:

- Subcutaneous injection four times per day
- One intramuscular injection every three to four weeks
- By continuous infusion with a subcutaneous pump

To control diarrhea Xermelo (telotriastat ethyl) tablets were recently approved by the FDA to be used in combination with somatostatin analog (SSA) therapy when SSA therapy isn't managing the symptom well enough.

Reducing alcohol and foods high in tyramine (such as aged meat and fermented foods) is also recommended because they can make symptoms worse.

Finally, to help patients navigate the new normal that comes with carcinoid syndrome, the Carcinoid Cancer Foundation has created a smartphone app patients can customize to track their symptoms and food reactions, get medication reminders, and document their moods and emotions.

Prognosis

As with most cancers, catching and treating carcinoid tumors early improves your prognosis.

Finding tumors when they're small and haven't spread gives patients the chance for a cure by removing them surgically.

While only 25% of deaths are caused by tumor growth and spread, once carcinoid syndrome sets in the prognosis is worse. Fortunately, there have been significant improvements in treatment over the past ten years.

A decade ago, the average survival after the first appearance of carcinoid syndrome flushing face was three years. With treatment advances using multiple approaches, that number has increased to 12 years.

Taking Action

While carcinoid syndrome became more widely known among physicians in the 90s after the development of Octreotide (Sandostatin) and subsequent education, it's still considered a rare disease.

Make an appointment with your family doctor and take some information from a reputable source like The Carcinoid Cancer Foundation or the National Organization for Rare Disorders to make sure you're communicating your concerns clearly and effectively.