Progressive GI Symptoms: Could it be Scleroderma?

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Overview

Functional gastrointestinal (GI) disorders are extremely common. However, on rare occasions, patients thought to have a functional GI disorder can develop progressive GI symptoms from another underlying condition. This is *truly rare*, but when symptoms do not respond as expected, or if symptoms get worse and worse despite a clinician's best efforts, it may be worth considering other things. This article is about scleroderma, which is one of many different conditions that can affect the GI tract. Most people with progressive GI symptoms do *not* have scleroderma, but some patients with early scleroderma can present with typical IBS symptoms, like bloating, constipation, diarrhea, and others. The sections that follow discuss what we know about scleroderma, and how scleroderma can affect the GI tract. As you read it, keep in mind that scleroderma is very rare, so chances are that you or a loved one does *not* have this condition. Nonetheless, we wish to raise awareness about scleroderma, discuss how it can affect the digestive tract, and provide pointers for how to recognize whether or not your GI symptoms might be from scleroderma.

Epidemiology and Causes of Scleroderma

Scleroderma literally means "thick skin." Patients with scleroderma can develop thickening of their skin, on the outside of their body, but also in the tissues within their body. The condition is uncommon, affecting approximately 300,000 people in the population. The cause of scleroderma remains unclear, but it is thought to be a problem with the immune system in which collagen is deposited in unusually high amounts throughout the body. This process can affect the blood vessels, in particular, but can also disrupt the gastrointestinal tract, among other bodily systems. Although scleroderma is usually managed by rheumatologists, it sometimes comes to the attention of gastroenterologists due to its strong impact on the bowels, as described further, below. The management and prognosis of scleroderma is beyond the scope of this short review, but treatment can be complex and difficult, and typically involves using potent therapies to inhibit the immune system, such as steroids, methotrexate, and several other drugs.

General Types of Scleroderma

Scleroderma is a progressive condition that presents in many different ways; it is not a "one-size fits all" disorder. Rheumatologists divide scleroderma into different types, including localized disease (affecting certain areas of the body only) and systematic sclerosis (SSc). SSc is a form of scleroderma that has skin and internal organ involvement, including the GI tract. Localized scleroderma does not affect internal organs. However, GI involvement occurs in approximately 90–95% of patients with SSc and is the leading cause of morbidity and the third most common cause of mortality in SSc. From here on in this article, scleroderma refers to SSc.

How Scleroderma Affects the GI Tract

After the skin, the digestive system is the most commonly affected organ system in people with scleroderma. Most people with gut involvement experience symptoms that interfere with their day-to-day activities and quality of life. The function of the gut is to push the food and liquid down from the mouth to the large intestine (or colon), extract and absorb nutrients, and excrete the waste in the form of stool. It does so by well-orchestrated and rhythmic motions of the gut muscles (also known as peristalsis). The primary events that cause trouble in the scleroderma gut are due to a decrease in the blood supply to the nerves, which are needed to stimulate the bowel. With decreased stimulation, there is progressive weakening of muscle strength and tone and resultant slowing and dyscoordinated motion of the gut. Virtually every gut symptom in scleroderma is the result of weakening of the gut muscle. The weakening starts in the esophagus (food pipe) and stomach, and works its way down to the small and large intestine. The sections that follow explain the reasons for the GI symptoms that scleroderma patients may experience, starting from the top down, so to speak.

Mouth

Approximately 20 percent of people with scleroderma develop secondary Sjögren's syndrome, a condition associated with very dry eyes and dry mouth. If you have progressive GI symptoms and find yourself with increasingly dry eyes and lacking saliva, then you should ask your doctor about Sjögren's syndrome and the possibility of underlying scleroderma. When dry mouth occurs, it can cause difficulty swallowing, gum

disease, or cavities. If people have symptoms of dry mouth, they should see a dentist at least every six months. In addition, use sugar-free lozenges to keep your mouth moist and use over-the-counter dry mouth products such as Biotene mouthwash and oral gel. Prescription medications such as Evoxac® and Salagen® may be helpful.

Esophagus (Food Pipe)

The esophagus transports food to the stomach by coordinated contractions of its muscular lining. This process is automatic and people are usually not aware of it, except when they swallow something too large, try to eat too quickly, or drink very hot or very cold liquids. They then feel the movement of the food or drink down the esophagus into the stomach, which may be an uncomfortable sensation.

The esophagus is the area where most scleroderma patients have problems, and fortunately, it has been the area where the most advances have been made in therapy. The most common symptom is heartburn (sensation of burning behind the breast bone), which is due to irritation of the esophagus by acid regurgitating (backing up or refluxing) from the stomach. Repeated or persistent heartburn is a symptom of gastroesophageal reflux disease, or GERD. Other symptoms may include dry cough at night, recurrent chest pain behind the breast bone, persistent hoarse voice, asthma (wheezing and shortness of breath), difficulty swallowing, mouth ulcers, and acid taste in the mouth.

It is important to keep in mind that GERD symptoms are remarkably common, but scleroderma is not. If you have GERD symptoms, it is very unlikely that you also have scleroderma. However, if you have GERD symptoms, progressive difficulty with swallowing, and other GI symptoms, then you may have something more than a typical functional GI disorder or normal variety GERD. Be sure to talk to your doctor if these things happen.

Stomach

Stomach symptoms in scleroderma are due to slow emptying of the food into the small intestine. The retention of food in the stomach leads to a sensation of nausea, vomiting, fullness, or bloating (also known as gastroparesis). In some people with scleroderma, the stomach can also have dilated blood vessels (telangiectasia) lining the walls of the stomach. This is also known as “watermelon stomach” due to its appearance on endoscopy. Slow and intermittent or rapid bleeding from these dilated blood vessels can cause low red blood cell count (anemia). The person may or may not have stomach symptoms and may only feel very tired and fatigued. If your doctor finds that you have anemia, then the diagnosis of a functional GI disorder should always be questioned until the anemia is otherwise explained, especially if you have something called “iron deficiency anemia.” Scleroderma is one of many different conditions that can cause iron deficiency anemia. Other, more common, conditions include erosions from GERD, peptic ulcer disease in the stomach, and celiac sprue, among many others.

Small Bowel

The small bowel is the place where we gain most of the nutrition from our food. The food is pushed from the stomach into the small bowel by the rhythmic motions of the gut. Once in the small bowel, the nutrients and vitamins are extracted from the food and the waste is propelled into the large bowel and excreted as stool. The symptoms, because of bowel involvement, are due to lack of muscle tone, leading to stagnation of food in the small and large bowel.

When scleroderma affects the small bowel, the symptoms are similar to gastroparesis (bloating, nausea, and vomiting), but abdominal pain may also occur. In addition, due to stagnation of food, the bacteria, which normally reside in small quantities in the small bowel, can multiply in the food causing small intestinal bacterial overgrowth, also called “SIBO.” These bacteria compete with nutrients and vitamins causing vitamin deficiencies, weight loss, and inability to gain weight. There may also be symptoms of diarrhea with foul smelling stools, which may be oily or hard to flush. Some people describe their stools like “oil and water,” in which beads of oil float on the surface of the water; this is *not* typical of functional GI disorders.

Another small bowel symptom that occurs in scleroderma is pseudo- (false) obstruction. In this condition, the bowel is not physically blocked as in true bowel obstruction but has just “had enough” and stops working. This is caused by weakening of the gut muscle. The patient experiences belly pain, distention, vomiting, and inability to “pass gas”.

Large Intestine (Bowel/Colon)

The main function of the large bowel is to reabsorb water and salts that have been secreted by the rest of the gut. This helps the formation of stools and helps to move the waste along. In scleroderma, there is weakening of the gut muscles and impaired motility. This can lead to constipation. Constipation means different things to different people. For many people, it simply means infrequent stools. Medically speaking, constipation usually is defined as fewer than three bowel movements per week. Severe constipation is defined as less than one bowel movement per week. This is usually associated with pain and feeling of incomplete emptying of the bowel. People may also describe severe straining with defecation and a sense of fullness even after trying to move their bowels. In scleroderma, some people develop so-called “wide mouthed diverticula,” or large pouches, along the lining of their colon. These do not ordinarily cause any symptoms. However, on rare occasions, the stool can get impacted in these pouches, which can lead to inflammation of the diverticula, called diverticulitis (this is similar to, but not exactly the same as, the common diverticulosis that you often hear about). Diverticulitis usually presents with belly pain and fever. People can also get diarrhea due to impacted stool.

Rectum

Stool incontinence (symptom of accidentally soiling the underwear before being able to get to a bathroom) occurs in up to a third of people with scleroderma. This is due to the weakening of the rectal muscle and poor control over the rectal sphincter. Again, keep in mind that incontinence can occur for a wide variety of reasons – by no means does it indicate scleroderma alone. But if there is progressive or increasingly common incontinence, coupled with worsening abdominal complaints, dry eyes, dry mouth, or trouble

swallowing, then you should ask your doctor about whether the incontinence could be a sign of scleroderma.
When this occurs from scleroderma, biofeedback therapy may be helpful by improving voluntary squeezing of the rectal muscle. Major scleroderma centers offer classes to teach people this technique. Surgery by an experienced surgeon might also be helpful. Preliminary studies have shown promise of sacral nerve stimulation (nerves that control rectal tone) in decreasing episodes of rectal incontinence, both in patients with scleroderma, and even incontinence patients without scleroderma.

Summary

In summary, scleroderma is a very rare disorder that can frequently affect the gut. When early in its course, some people with scleroderma can be misdiagnosed as having a functional GI disorder, such as IBS, functional constipation, or functional bloating, among other conditions. Although it is very rare for patients to be misdiagnosed like this, it is worth keeping scleroderma in mind if GI symptoms are progressive, are associated with unusually dry eyes, dry mouth, or trouble swallowing, and have not responded to usual therapies by your doctor. Other symptoms including painful and swollen joints, skin tightening, and Raynaud's phenomenon (reduced blood flow to fingers, toes, and rarely to the nose, ears, and lips) are common in scleroderma. The symptoms of scleroderma can be distressing and can cause impairment of a person's quality of life as well as other complications. An experienced rheumatologist and laboratory tests can make a definitive diagnosis of SSc. Appropriate treatments are often effective.

[The Scleroderma Foundation provides support and information about the management of scleroderma. They may be contacted at: 300 Rosewood Drive, Suite 105, Danvers, MA 01923. Phone 978-463-5843 or 800-722-4673. www.scleroderma.org]

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