Eosinophilic Esophagitis

Overview

What is Eosinophilic Esophagitis?

Eosinophilic esophagitis (also known as EoE) is a disease characterized by the presence of a large number of a special type of white blood cell, the eosinophil, that can cause inflammation in the esophagus. This inflammation can lead to stiffening or narrowing of the esophagus, which can lead to difficulty swallowing (dysphagia) or food getting stuck in the esophagus. Reflux of stomach acid contents into the esophagus can also cause eosinophils as well as inflammation in the esophagus. In EoE, the eosinophils are present even after acid reflux has been treated. Although eosinophils may be found in the rest of the gastrointestinal tract in a healthy person, when present in the esophagus, this usually suggests an abnormal condition. While other illnesses such as gastroesophageal reflux disease (GERD), parasitic diseases or inflammatory bowel disease may cause eosinophils in the esophagus, EoE is the most common cause of large numbers of eosinophils in the esophagus.

How common is EoE in adults?

While EoE was previously thought to be a rare disease, it has recently been recognized as one of the most common causes of difficulty swallowing and food impaction in young adults. This has become a global trend with increased cases of EoE being reported from five continents. The cause for this rise is likely a combination of increasing occurrences of EoE and a growing awareness of the condition among gastroenterologists, allergists and pathologists. It is thought that the disease may be increasing similar to the increases seen in other allergic disorders such as asthma and allergic rhinitis. Estimated occurrences of this condition in adults may be as high as 1-3 per 10,000 people, based on information from patients in Australia and Switzerland. Although some studies suggest that the disease is more common in the Caucasian population, cases have been seen in patients of African American, Asian and Hispanic descent. EoE affects males three times more often than females.

Symptoms

What are common symptoms of EoE in adults?

The most common presenting symptoms in adults are difficulty swallowing solid food and food impactions in which food gets lodged in the esophagus and is unable to pass into the stomach. If patients develop a food impaction, an endoscopy is often needed to help relieve this obstruction. Most adults with symptoms are between the ages of 20 to 40, although there have been cases of adults presenting at much later ages. Other less common symptoms include heartburn and chest pain.

Diagnosis

How is EoE diagnosed?

Currently, the only way to diagnose this condition is by performing an upper endoscopy with biopsy (taking tiny pieces of tissue) of the esophagus. During endoscopy, a thin, flexible tube with a camera, which allows the doctor to see the inside of your esophagus, is inserted into the esophagus while the patient is sedated (sleep caused by medication). Biopsies taken from the lining of the esophagus (mucosa) are later inspected under a microscope by a pathologist for characteristic changes of EoE. These changes include large numbers of eosinophils in the superficial portion of the tissue biopsy and signs of inflammation in the tissue. Sometimes scaring or fibrosis can be seen in the deeper portions of the tissue. In EoE, the eosinophils are limited to the esophagus and are not present in the stomach or duodenum.

Usually, there are characteristic features that the gastroenterologist can see in the esophagus of patients with EoE. These include linear furrows or creases in the esophagus and concentric rings of the superficial layer of the esophagus. Other features, including narrow esophagus, white spots on the esophageal tissue and short, very narrow segments of the esophagus called strictures, may also be seen. While these changes are suggestive of EoE, their presence alone does not diagnose the condition. The esophagus can also appear normal in adult patients with EoE.

Symptoms of gastroesophageal reflux disease (GERD) such as heartburn or regurgitation can overlap with symptoms of EoE. Since gastroesophageal reflux disease is much more common than EoE in the adult population and can also be a cause of eosinophils in the esophagus, it is important to distinguish the two. Therefore, if eosinophils are found on a tissue biopsy of the esophagus, it is suggested that the patient start treatment with acid reducers to see if the eosinophils go away once the reflux is treated. This requires another endoscopy. If the eosinophils are still in the tissue after reflux has been treated, then the patient most likely has EoE. Another method to identify if acid in the esophagus is contributing to the eosinophils is to complete a test called an esophageal pH test. In this test, a very thin tube is placed through the nose into the esophagus and stomach, or a temporary sensor is placed in the esophagus via endoscopy. Both allow levels of acid in the esophagus to be monitored for a period of time, usually 24 - 72 hours. If this test shows high levels of acid in the esophagus, it suggests that GERD may be the cause of the eosinophils in the esophagus. In more complicated situations,

some patients have both GERD and EoE and therefore will need to have treatment for both conditions.

Cause

What is the cause of EoE?

Currently the cause of EoE in adults has not been clearly identified. Some studies have suggested an allergic reaction to environmental and food allergens. There may also be a genetic cause that may lead to EoE in some patients. A recent study has identified an increase in a gene coding for a protein called eotaxin-3 in patients with EoE. Further support that there may be a genetic link is that some adults have a family history of allergic disorders and a family history of EoE.

A history of allergic conditions such as allergic rhinitis, asthma, eczema or food allergy has been seen in up to 70% of adults with EoE either by history or positive allergy testing. In one recent study, adults with EoE treated with dietary elimination improved their EoE, but recurrence of EoE happened when certain foods were added back to the diet. This suggests that food allergens play a role in some adults with EoE.

Treatments

What are the most common treatments for EoE in adults?

Currently, there is no one accepted therapy for all patients with EoE. Although dietary therapy is the most common treatment of pediatric EoE, this has not been widely accepted among gastroenterologists who treat adult patients. Many adult patients are initially treated with acid-blocking medications to rule out GERD. If this does not improve symptoms or tissue changes of the eosinophils, then steroids taken using an asthma inhaler, but swallowed rather than inhaled by the patient, have been tried with good, although limited results. This treatment tends to be well tolerated; side-effects of a fungal infection called thrush or candida of the esophagus are relatively rare.

Dietary treatment may consist of an elemental diet, a "six-food-elimination diet" or a targeted-elimination diet, usually for six weeks. After this point, if the disease improves, foods are reintroduced one at a time to help identify the food trigger. An elemental diet is another potential treatment. It is an amino-acid based formula, taken usually for six weeks. Elemental formula can have a poor taste and can be costly; therefore, a six-food-elimination diet is preferred by most patients. A six-food-elimination diet is a diet that contains no milk, soy, egg, wheat, nuts or seafood. These foods are the most common food allergens found in patients with EoE. A targeted-elimination diet is a diet that is based on eliminating foods found to be positive on allergy testing.

Dietary therapy has been shown to be helpful in some adults with EoE and may be tried with motivated (willing to follow through with food avoidance) patients under the care of an experienced provider or dietician.

If patients do not respond to medical therapy or diet exclusion, an esophageal dilation or "stretching" is sometimes performed if there is narrowing of the esophagus. Although dilation may be helpful in the short term, repeated dilations may be needed to control symptoms. Because dilation alone does not affect the underlying inflammation in the esophagus, this procedure is usually performed in patients who are also being treated with medical or dietary therapy. The risks of esophageal dilation include chest pain after the procedure and in rare cases a perforation or tear of the esophagus. While dilation can be performed safely, it must be done with caution and is almost always performed after a trial of medical or dietary therapy has failed.

What is the long term consequence of having EoE?

There is limited information about the natural history of this disease in adults. Current studies suggest that it is a chronic, reoccurring condition. Patients have continued symptoms, although esophageal eosinophil levels may change over time. Complications, including esophageal strictures and food impactions, may occur. In very rare cases, forceful vomiting, prolonged food impactions and esophageal dilations may result in a perforation or tear of the esophagus which needs immediate medical attention. Currently treatment of EoE is directed at controlling symptoms, reducing eosinophil levels in the tissue, and preventing complications of the disease, such as food impactions.

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Patient LInks

- American Partnership for Eosinophilic Disorders (http://www.apfed.org/)
- Cured Foundation (http://www.curedfoundation.org)
- The International Gastrointestinal Eosinophil Researchers (TIGER) (http://www.tiger-egid.cdhnf.org)

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