MANAGEMENT OF CARDIAC ACHALASIA

ESSAY
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BY

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List Of Abbreviations

BOTOX..…….. Botulinum Toxin

CCK-OP…….…CholecystoKinin-OctaPeptide

CNS……Central Nervous System

CT……...Computed Tomography Scan

DM……..……Diabetes Mellitus

DMN……..……Dorsal Motor Nucleus

DES…….……Diffuse Esophageal Spasm

GE……..…….Gastroesophageal junction

GERD……. Gastroesophageal Reflux Disease

GIT……..……Gastrointestinal Tract

HLA……...…..Human Leukocytic Antigen

IgG……..….Immunoglobulin G

LES…….….Lower EsophagealSphincter

NADPH……Nicotinamide-Adenine Dinucleotide Phosphate

NO……...Nitric Oxide

NPO…….……Nothing Per Orum

POEM……..…Per-Oral Endoscopic Mytomy

SEMS……...…Self Expanding Metal Stents

UES…….……Upper Esophageal Sphincter

VIP……..….Vasoactive Intestinal Peptide

Summary

The cause of achalasia is unknown. There is loss of nerve cells in the Auerbach's plexus between the two muscle layers of the esophageal wall and in the lower esophageal sphincter. This defect results in loss of peristalsis, and failure of the lower esophageal sphincter to relax. These changes result in failure of esophageal emptying and retention of solid and liquid foods in the esophagus.

Dysphagia, regurgitation, heart burn and chest pain are the most annoying symptoms in patients with achalasia. Regurgitation, especially at night when the patient is recumbent, can result in aspiration of fluid into the lungs causing bronchitis and pneumonia.

Once the diagnosis is established, a decision regarding therapy is in order. Some patients with early achalasia and mild symptoms may elect to delay treatment or try medical (drug) treatment. Such treatment with calcium channel blocker drugs may help by transiently relaxing the closed sphincter but this response is usually short-lived and not reliable for long-term use.

The standard medical treatment option is dilation of the tight sphincter with balloon dilators. These instruments are intended to over-stretch the sphincter to the point that the muscle fibers lose

their ability to contract tightly and thereby allow the esophagus to empty more efficiently. This form of therapy provides good to excellent improvement in 70 to 90% of patients. The main risk of this treatment is perforation or full-thickness tear of the esophagus that occurs in 2 to 5% of patients. If this occurs a surgical operation usually is needed to close the tear.

Another treatment for achalasia used for the past 10 years is the injection of Botulinum toxin (Botox) into the esophageal sphincter. It prevents the release of a chemical (acetylcholine) from the nerves in the lower esophageal sphincter resulting in relaxation and reduction of the high sphincter pressure. Improvement of the patient's swallowing and regurgitation in about 80% of cases. Its only significant drawback is a limited duration of relief.

In the past years, when balloon dilations had failed on two occasions, an operation called a Heller myotomy was recommended. This surgical procedure gave good to excellent results and can be done either through transthoracic or transabdominal approach. Currently laparoscopic myotomy can be performed, As more experience has been gained this procedure has become the preferred technique. Current long-term improvement is being confirmed in over 80% of cases. If the sphincter muscle is rendered too weak by myotomy, there is a risk

for acid in the stomach to reflux into the esophagus and cause esophagitis or a stricture. Surgeons may attempt to reduce this risk by performing an antireflux operation at the time of the myotomy. Even if this problem occurs the acid reflux can be adequately treated by acid suppressing drugs.

It can be concluded that laparoscopic myotomy is considered the best option for treating patients with achalasia among all therapeutic method because it shows the best long-term results, particularly in young patient, and this laparoscopic procedure is considered to be minimally invasive. Pneumatic dilatation should be seen as the second-line therapeutic option. This method also shows good long-term results, but has a higher complication rate. Botulinum toxin injection should only be considered for selected patients as in elderly patients, patients with severe comorbidity.

- **Conclusion**

- The exact cause of achalasia is unknown.
- Dysphagia, regurgitation, heart burn and chest pain are the most annoying symptoms.
- Plain X-ray chest, barium swallow, esophageal manometry and endoscopy are the most important investigation.
- Laparoscopic myotomy is considered the best option for treating patients with achalasia.

Introduction

Achalasia is a primary esophageal motor disorder of unknown etiology, which is characterized by aperistalsis of the esophageal body and incomplete lower esophageal sphincter (LES) relaxation. Abnormalities in both the muscle and nerve components can be detected in patients with achalasia. The destruction and loss of the inhibitory myenteric ganglion cells is thought to be a primary mechanism for the disease; however, severe myopathy of the smooth muscle cells has also been suggested to be the primary lesion (***Pohl and Tutuian, 2007).***

The etiopathogenesis of achalasia is mostly unknown . Histopathological examination revealed irreversible degenerative changes of myenteric plexus primarily involving inhibitory nonadrenergic and noncholinergic neurons, chronic inflammation of myenteric plexus as a result of neuroadrenerative , infectious or autoimmune primary disorder is the most frequently mentioned pathological mechanism ***(Obuchi et al., 2006).***

The common symptoms of achalasia are dysphagia, regurgitation and chest pain varying in their

frequency and severity with cardiac origin of atypical chest pain had been reasonably ruled out on the basis of electrocardiogram results and normal cardiac enzymes . Achalasia related chest pain can be defined as chest pain with chest oppression ***(Omura et al., 2006).***

Methods of diagnosis of Achalasia include barium esophagogram which is performed to evaluate the gastroesophageal junction, the morphology of the distal esophagus, an upper gastrointestinal endoscopy is performed to rule out the presence of peptic ulcer or an esophageal cancer, esophageal manometry and 24-hour pH manometry are done for preoperative assessment ***(Arain et al. , 2004).***

Achalasia is not curable and current treatment options are limited to reducing the pressure gradient across the LES, thus facilitating gravity–aided esophageal emptying. The most effective and current therapeutic options include pneumatic dilatation, laparoscopic or surgical myotomy & use of self expanding metal stents (SEMS).

Pharmacological agents, such as Botulinum toxin, calcium channel blockers, nitrates are also used though they are less effective and temporary; despite controversy

regarding the optimal choice of initial management for achalasia, pneumatic dilation and cardiomyotomy remain the most common therapy for esophageal achalasia in many institutions ***(Yi et al., 2008).***

Aim of the work

This essay was made for better understanding of the etiology and pathogenesis of cardiac achalasia; as well as diagnostic modalities & the controversies regarding the different treatment options, including recent trends in the diagnosis and treatment.