Laparoscopic Seromyotomy with and without Endoscopic guidance in Cardiac Achalasia

Thesis

Submitted in Full Fulfillment of the Requirement for the Master Degree in General Surgery

Presented By

Gamal El-Deen M.Atia

(M.B.B.CH.)

Supervised by

Prof.Dr. Safwat Abdel-Kader Salem

Professor of General Surgery
Faculty of Medicine
Cairo University

Prof. Dr. Ahmed Shehata Kawashti

Professor of General Surgery
Faculty of Medicine
Al-Azhar University

Assisted in Supervision

Dr. Tamer Nabil

Lecturer of General Surgery
Faculty of Medicine
Cairo University

Faculty of Medicine
Cairo University
2008

Abstract

Achalasia affects both sexes equally, typically presenting between the ages of 20 and 50, though it can occur at all ages. Ineffective Relaxation of the lower esophageal sphincter (LES) combined with loss of esophageal peristalsis leads to impaired emptying and gradual esophageal dilatation. Dysphagia is the cardinal feature of achalasia, accompanied by varying degrees of aspiration, weight loss, and pain. The anatomic defect appears to be a decrease or loss of inhibitory nonadrenergic, noncholinergic ganglion cells in the esophageal myenteric plexus. Histological analysis of esophagi resected from patients who had end – stage achalasia demonstrates myenteric inflammation, progressive depletion of ganglion cells and subsequent neural fibrosis. There is a significant reduction in the synthesis of nitric oxide and Vasoactive Intestinal polypeptide (VIP), the most important mediators of relaxation in the lower esophageal sphincter.

Key Words :

Cardiac Achalasia - Endoscopic guidance

ACKNOWLEDGEMENT

First of all, I am deeply thankful to God by the grace of whom this work was possible

I wish to thank ***Prof. Dr. Safwat Abdel-Kader Salem, Professor of General Surgery, Faculty of Medicine, Cairo University***. I have been most fortunate in having his continued guidance in supervising every section in this thesis . His constant encouragement and valuable advice were indispensable. I am definitely indebted to him more than I can express.

The credit of bringing this work to light goes to ***Prof. Dr. Ahmed Shehata Kawashti , Professor of General Surgery, Faculty of Medicine, Al-Azhar University***. His continued help and guidance made it possible to bring this work to its final shape. No words can express my feelings towards him.

My deepest gratitude, appreciation and thanks to ***Dr. Tamer Nabil, Lecturer of General Surgery, Faculty of Medicine, Cairo University***, for his sincere supervision, cooperation, continuous support throughout this work.

My deepest gratitude, appreciation and thanks to ***Dr. Sherif Emad El.Dine, Lecturer of General Surgery, Faculty of Medicine, Cairo University***, for his sincere supervision, cooperation, continuous support throughout this work.

Finally, I wish to thank everyone who helped me to complete this work.

Gamal El-Dean M. Atia
2008

To

My father

My mother

My Wife

And all my family

Thank you deeply

From my heart

CONTENTS

INTRODUCTION

INTRODUCTION

Laparoscopic Seromyotomy with and without Endoscopic guidance in cardiac Achalasia

Achalasia is a primary esophageal motor disorder of unknown etiology.

Achalasia affects both sexes equally, typically presenting between the ages of 20 and 50, though it can occur at all ages. Ineffective Relaxation of the lower esophageal sphincter (LES) combined with loss of esophageal peristalsis leads to impaired emptying and gradual esophageal dilatation. Dysphagia is the cardinal feature of achalasia, accompanied by varying degrees of aspiration, weight loss, and pain. The anatomic defect appears to be a decrease or loss of inhibitory nonadrenergic, noncholinergic ganglion cells in the esophageal myenteric plexus. Histological analysis of esophagi resected from patients who had end – stage achalasia demonstrates myenteric inflammation, progressive depletion of ganglion cells and subsequent neural fibrosis. There is a significant reduction in the synthesis of nitric oxide and Vasoactive Intestinal polypeptide (VIP), the most important mediators of relaxation in the lower esophageal sphincter.

Macroscopically, there may often be thickening of the circular layer of the distal esophagus. Achalasia also carries a slightly increased risk of squamous cell carcinoma in particular. (Todd et al., 2005)

Patient presentation

Most patients who have achalasia present with progressive dysphagia to solids and liquids, though symptoms may be subtle and nonspecific early in its course. The mean duration of symptoms before presentation is 2 years, and the diagnosis often takes much longer because the symptoms are often attributed to gastroesopahageal reflux disease (GERD) or other disorders.

Initially, the patient may complain of the sensation of retro-sternal "sticking" of foodstuff . Stress or cold liquids may exacerbate dysphagia.

Patients may regurgitate undigested food, especially after meals or when lying supine, Patients may stand after eating or raise their arms over their head to enlist gravity and to increase the intra-thoracic pressure in an attempt to aid esophageal emptying. If unable to force food into the stomach by the ingestion of liquids or other means, spontaneous or forced regurgitation are often employed to evacuate the esophagus. Occasionally, physicians even confuse the disease with an eating disorder . The regurgitation aspiration of esophageal contents may lead to pulmonary disease . In fact up to 10% of patients who have achalasia experience significant bronchopulmonary complications (Howard et al., 2005).

Many patients express a sensation of heartburn explaining why many are initially diagnosed as gastro-esophageal reflux disease . Though patients who have achalasia may experience gastro-esophageal reflux (Shoenut et al., 1988) more often heartburn is secondary to fermentation of retained undigested food in the esophagus.

Chest pain, clearly distinguishable from heartburn, occurs in 30% to 50% of patients. The etiology of this pain is unclear, and is unpredictably relieved by esophageal myotomy and other treatments. Weight loss is variable and tends to be insidious. The magnitude of weight loss tends to correlate with the severity of the underlying disease. Rapid onset of symptoms (<6 months), advanced age (>50 years), or significant weight loss should raise suspicion for pseudoachalasia, usually secondary to malignancy or extra-luminal obstruction. In these cases, a thorough work-up with a CT scan or endoscopic ultrasound must be performed before further therapy is considered. (Todd et al., 2005)

Evaluation

A barium eosophago-gram is typically the first imaging study used in the evaluation of dysphagia. The scout film may demonstrate an air-fluid level in the esophagus with a paucity of gastric air. The classical appearance of achalasia on a barium study is the "bird's beak" tapering of the distal esophagus, with a column of contrast in the esophageal lumen. Variable esophageal

dilation is seen ranging from mild in the early stages to the massive sigmoid-shaped esophagus of end-stage achalasia. Fluoroscopic evaluation may also reveal non-propulsive, tertiary contractions of the esophageal body, with failure to clear the barium bolus from the esophagus.

Manometry is the gold standard for confirming the diagnosis of achalasia.

The resting lower esophageal sphincter pressure may be elevated, but is usually normal, and fails to relax completely with swallowing .Complete absence of peristalsis is the key of achalasia. The waveforms are usually simultaneous and of low amplitude. (Todd etal 2005) When present, they are simultaneous and non-propulsive in nature.

Subsets of patients who have "vigorous" achalasia are found to have high-amplitude contractions. In patients who have a dilated and tortuous esophagus, the LES may be difficult to in-tubate, requiring fluoroscopic guidance for manometry catheter placement. In the patient who will not tolerate esophageal manometry, nuclear scintigraphy can be used to evaluate esophageal transit. (Todd et al., 2005)

Twenty four–hour PH monitoring is neither required nor usually helpful.

Although patients who have achalasia may experience some element of gastro-esophageal reflux, it is not clinically relevant. Further, acidification of esophageal contents secondary to fermentation of retained food may lead to a false positive study. (Smart etal 1987)

Endoscopic evaluation is used to rule out other processes that may mimic achalasia. The characteristic appearance is in a tonic, dilated esophagus with a tightly closed LES that does not open with insufflation. With gentle pressure, the scope is admitted through the LES with a "pop" in contrast with a peptic stricture or a malignancy, which do not yield. The GE junction, including a retroflexed view of the gastric cardia, should be carefully

inspected. Biopsies of any mucosal abnormality should be obtained. If pseudoachalasia can not rule out, endoscopic ultrasound or a CT scan may prove informative. (Todd et al., 2005) .

Any treatment of achalasia is directed at the palliation of symptoms and can not change the underlying pathology. The neuromuscular defected is not corrected. The goal of all therapeutic options is to relieve the functional obstructed of the distal esophagus, thus improving esophageal emptying.

Treatment options

A- Treatment involves the following options:

The use of drugs to treat achalasia would seem to be attractive because of its noninvasive nature. Several agents relax smooth muscle, and thus theoretically decrease LES pressure. Unpredictable and incomplete absorption of oral formulations secondary to poor esophageal emptying is one limitation; thus sublingual administration is the most effective route. Nitrates and calcium –channel blockers are the most commonly used medications. They are more effective in patients who have mild symptoms without severe esophageal dilatation. Side effects such as headaches and peripheral edema are common, limiting their adoption. Relief from these agents is inconsistent and generally short-lived most patients showing continued progression of their disease (Traube etal 1989). Their usefulness is limited to temporizing symptoms until more effective therapy is attempted, or in those patients deemed to frail to undergo invasive treatment.

B- Botulinum toxin:

Botulinum toxin A (Botox, Allergan, Irvine, California) is a neurotoxin produced by Clostridium Botulinum that binds to cholinergic nerves and irreversibly inhibits acetylcholine release. This effect is eventually overcome by regeneration of new synapses. Botox is injected into the LES through the working port of a flexible endoscope, with minimal incidence of immediate complications. Early enthusiasm has waned because results have

not proven to be durable Botox injection is initially effective in 60% to 85% of patients but 50% develop recurrent symptoms within 6 months (Vaezi & Richter 1998) . Repeat administration is a possible, but efficacy is diminished with subsequent injections. Because Botox is a relatively expensive therapy, the need for repeat treatment limits both its conveniences and cost-effectiveness. Another problem with this therapy is that Botox injection can cause an intense inflammatory reaction of the GE junction, with subsequent fibrosis. This may impact future surgical therapy , because most patients have continued or progressive symptoms , the author's experience during esophageal myotomy is that there is more difficulty in finding the sub-mucosal plane in patients who have had prior Botox therapy than in untreated patients (53% versus 7%) . Although there does not appear to be a difference in the ultimate relief of dysphagia, they did experience an increased rate of perforation (7% versus 2%) in patients who have had prior Botox therapy (Horgn etal 1999). Although this is not well understood, Botox appears to be more effective in older patient (more than 55 years) and in those who have vigorous achalasia. Botox should be reserved for patients unwilling or deemed unfit to undergo an invasive procedure. Also, in patients who have atypical presentation, Botox my be considered as a diagnostic procedure. By simulating the effect of esophageal myotomy, it can identify patients who are likely to have relief with an operation (Todd et al, 2005).

C -Pneumatic dilatation:

The oldest treatment of achalasia is forceful dilatation of the LES, originally accomplished by the passage of a piece of whole bone with a sponge fixed to the end. (Wiuis 1679) This therapy has become more effective with the development of graded polyethylene balloons. Under fluoroscopic guidance, balloons (at least 30 mm in diameter) are passed through the LES and inflated, disrupting the fibers of the LES. The balloons are keeping inflated from 1 to 3 minutes and then deflated. This is followed by an esophagram with water –soluble contrast to evaluate the LES diameter and to evaluate for perforation. If no extravasation is seen, the patients is observed for 6 hours and then discharged to home. The "graded" approach refers to the use of serially larger

balloons (up to 40 mm) with subsequent dilatations for initial nonresponders. Only a single dilatation is performed per session. Response rates of 60 % to 90% can be achieved, with approximately 70% of patients obtaining substantial relief of dysphagia after 1 year (Csendef etal 1989). Repeat dilation is often used 'but its efficacy is diminished after two sessions. Patients who have a poor result after initial dilatation or early return of their symptoms are predictably lees likely to respond with subsequent dilation .most patients are able to tolerate pneumatic dilatation. Interestingly, younger patients do not respond as well as older patients (Clouse etal 1991). This is thought to be due to their tissues being more compliant, and simply stretching during dilatation rather that tearing. The presence of a hiatal hernia, significantly dilated esophagus (>7 cm), or an epiphrenic diverticulum increases the risk of perforation and these are relative contraindications. Although the likelihood of improving dysphagia increases with increasing balloon diameter, so does the likelihood of perforation. Overall, the incidence of perforation is about 2%per dilation attempt Reynolds and park man 1989. Whether to treat achalasia initially with pneumatic dilatation depends on physician and surgeon expertise, patient age and comorbidities, and patient preference. If dilation fails, a myotomy can usually be performed without additional difficulties.

D-surgical therapy

Ernest Heller first described cardiomyotomy for achalasia in 1914. His original description was of two myotomies, one anterior and one posterior, along the GE junction .This has been modified, and now only an anterior myotomy is performed. Excellent results with cardiomyotomy can be achieved in 90% to 95%of patients. Extra mucosal cardiomyotomy provides more reliable relief of dysphagia than pneumatic dilation, because it allows accurate division of LES muscle fibers rather than blind disruption (Abdominal approach). Each approach is associated with an obvious incision and postoperative stays of 7to 10 days. For this reason, despite superior long-term results from surgical myotomy (Csendes et al., 1989), most patients were treated by less invasive therapies, such as pneumatic dilatation. Recent

developments in minimally invasive techniques now allow performance of cardiomyotomy by either a thoracoscopic (Pellerini et al., 1992) or a laparoscopic approach. (Cuschieri et al. 1991) .

Reduction in postoperative pain and morbidity, as well as improved cosmoses, has made the surgical option to a myotomy because of poor relief of dysphagia. The authors feel that with laparoscopy that there is little to lose by attempting a myotomy and reserving an esophagectomy for failures. Using this approach, the majorities of patients obtain relief and avoid subsequent esophagectomy.

The first approach using minimally invasive techniques was thoracoscopy (Pellerini et al., 1992) .Through a left thoracoscopic approach, a long myotomy could be performed, extending approximately 0.5cm across the GE junction (similar to the open approach). The thought was that this would provide relief of dysphagia without rendering the cardia completely incompetent and resulting in significant reflux. Initial were promising, with 89% of patients experiencing relief of dysphagia (Pellerini et al 1993); however, over time several patients (9/35) in the authors' series required myotomy extension or postmyotomy dilation to relieve dysphagia (pahi. atal.1999). Furthermore, over 60% had abnormal reflux when 24-hour pH monitoring was performed. Thus it became clear that successful relief of dysphagia often depends on extending the myotomy well onto the stomach, which can only be done from an abdominal approach. Also, even a limited gastric myotomy and hiatal dissection, such as is performed with a thoracoscopic myotomy, result in a high incidence of reflux.

For these reasons, Todd et al and most esophageal surgeons perform a Heller myotomy via a laparoscopic approach. The advantages include excellent visualization of the distal esophagus and the stomach, so that an extended gastric myotomy and an anti-reflux procedure may be performed. Moreover, it a voids the anesthetic complexity of single –lung ventilation required for thoracoscopy. Todd et al have substantial experience with both approaches , and have found that laparoscopy is more effective in relieving dysphagia (93% versus 85%) with a shorter hospital stay

(46 versus 72 hour) and less postoperative reflux (17% versus 60%) (Patti et al 1999).

There are two main important points surrounding Heller myotomy. One is the extent of the esophageal myotomy, the other is whether an anti-reflux procedure should be performed, and if so, which one. Although there is agreement that the proximal extent of the myotomy should reach 6 to 7 cm above the GE junction, this distal extent of the myotomy is to controversial. Some consider that the goal in performing a myotomy is to adequately relieve dysphagia without unnecessary disrupting the anti-reflux barrier. Todd et al have found that there is no length of esophageal myotomy that maximally relieves dysphagia and minimizes the occurrence of reflux. This is emphasized by the thoracoscopic approach that , despite a limited distal extension of the myotomy , produced GERD in most patients .We recently compared a more traditional approach (a 1.5- 2 cm gastric myotomy) with an extended gastric myotomy (at least 3 cm) . They found that the longer myotomy resulted in less dysphagia (1.2 versus 2.1 on a 5 point frequency scale) and fewer interventions for recurrent dysphagia (3% versus 17%) (Oelsch – Lager etal 2003). Because we advocate an extended gastric myotomy, we feel that an anti-reflux procedure is prudent in most cases. Those who advocate not performed an anti-reflux procedure with good clinical result and low incidence of heartburn (Sharp etal 2002) however, few groups perform postmyotomy 24-hour PH monitoring to evaluate for the true incidence of GE reflux without a fundoplication. Moreover, this practice results intervention rates for dysphagia as high as 14% (Richards etal 1999) which we think is likely the result of a limited gastric myotomy.

Most surgeons find that performing an anti-reflux procedure in conjunction with laparoscopic myotomy does not add significant time or morbidity to the operation, and is not associated with increased postoperative dysphagia. Certainly , a partial fundoplication (eg , Nissen fundoplication) may cause a functional obstruction for a nonpropulsive esophagus , resulting in a high incidence of dysphagia (Wius and Hunt 2001) An anterior (Dor) fundoplication requires less posterior dissection , and thus is easier and theoretically preserves more of the antireflux barrier .

Also, because the warp is brought anterior the myotomy, it potentially covers any undetected mucosal injuries .
A posterior (Toupet) fundoplication is the preferred partial fundoplication when indicated for GERD; however, its superiority in preventing reflux after myotomy has not been demonstrated.
Because it holds the edges of the myotomy open, a Toupet may provide protection against recurrent dysphagia. For these reasons, it is the procedure of choice for the author's group. Although each of these antireflux techniques has its own theoretical benefits and champions, there is no strong evidence supporting one over the other.

E- Laparoscopic Heller myotomy – operative technique:

The setup is the same as that for a Nissen fundoplication. The patient is placed in a modified lithotomy position, with a standard five –port placement. In the authors' group, Todd et al routinely mobilizes the gastric fundus and short gastric vessels to minimize tension on the subsequent fundoplication. An extensive anterior and lateral hiatal and mediastinal esophageal dissection is performed to maximize the length of the myotomy. It is important to identify the left (anterior) vagus and separate it from the esophagus to enable the performance of a continuous esophagogastric myotomy.

A lighted 52 Fr bougie is past into the body of the stomach. The Bougie illuminates the esophagus, which helps with identification and stability when performing the myotomy. The fat pad overlying the cardioesophageal junction is excised, a step critical to accurately identifying the GE junction. A Babcock clamp opened over the bougie just distal to the GE junction provides exposure of the distal esophagus. Todd et al perform the myotomy with an L-shaped hook electrocautery device, although minimal energy is used to as to minimize mucosal injury. The appropriate plane may be more difficult to identify in patient who have previously undergone treatment with Botox or pneumatic dilatation. The longitudinal muscle fibers are divided first, exposing the inner circular muscle, which is then separated from the mucosa. For this reason most submucosal bleeding should be controlled with pressure and time. The myotomy should be carried