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Myotomy of cricopharyngeal muscle in the therapy of dyskinesia of upper esophageal sphincter

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Dyskinesia of the esophagus includes the entire esophagus from the upper esophageal sphincter to the lower esophageal sphincter together with a wide spectrum of physiopathological and clinical variations. Related to functional diseases such as Dyskinesia of UES (Upper Esophageal Sphincter), myotomy is an excellent procedure for patients with cervical dysphagia. In our experience, the surgical treatment is the only procedure to treat UES dyskinesia. For a successful intervention is mandatory: a pharyngeal pump efficiency, hypertension of UES and conservation of esophageal peristalsis in the absence of GERD (Gatroesophageal Reflux Disease). From January 2006 to December 2011, the Authors have treated 11 patients, 9 of which suffered from idiopathic Dyskinesia of UES, 2 had a previous brain ictus; 4 of them had an associated Zenker diverticulum; in 1 case there was an associated goiter. In all these cases we have performed an extramucosal myotomy of cricopharyngeal muscle through a right laterocervical approach. All the patients had benefit from the operation immediately, with resolution of dysphagia, one patient with previous brain ictus, has enjoyed only a partial improvement of symptoms, in one case we performed contemporary a thyroidectomy. The authors believe in the surgical treatment as the only procedure, after accurate selection of patients, to the cure of the patients.

Key words: **Esophageal sphincter, upper - Movement disorders - Surgical procedures, operative.**

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Dyskinesia of the esophagus includes the entire esophagus from the upper esophageal sphincter to the lower esophageal sphincter together with a wide spectrum of physiopathological and clinical variations. Related to functional diseases such as Dyskinesia of UES (Upper Esophageal Sphincter), myotomy is an excellent procedure for patients with cervical dysphagia. In our experience, the surgical treatment is the only procedure to treat UES dyskinesia. For a successful intervention is mandatory: a pharyngeal pump efficiency, hypertension of UES and conservation of esophageal peristalsis in the absence of GERD (Gatroesophageal Reflux Disease).

Case series

In our case-histories, from January 2006 to December 2011, we have treated 11 patients, 9 of which suffering from Idiopathic Dyskinesia of UES; in 2 cases it was possible to recognize a previous ictus as probable cause of the oesophageal dyskinesia (Wallenberg Syndrome);[1-4] in 4 cases there was a Zenker diverticulum (Figure 1), and in 1 case a big goiter was associated to the oesophageal dyskinesia (thyroidectomy was performed before the myotomy of cricopharyngeal muscle). The mean age of patients was 65.

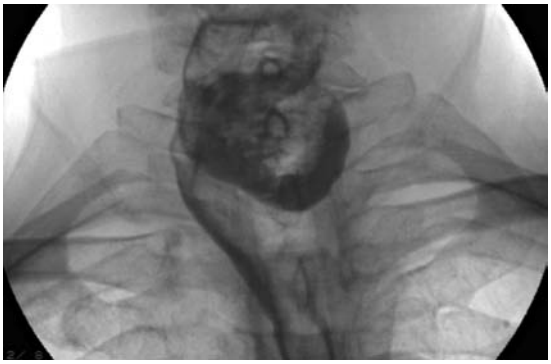

Figure 1.—Cine-esophagram control showed Dyskinesia of UES causing Zenker diverticulum.

Figure 2.—Small aspiration of the Barium during barium swallow X-ray.

Before entering in the surgical protocol it is mandatory for the patients: adequate force of the lingual muscle, pharyngeal pump efficiency and integrity mechanisms governing the beginning of the voluntary swallowing, demonstration of obstacle (by videofluorography) to transit-bolus, hypertension of UES by manometry, conservation of esophageal peristalsis in the absence of GERD and last but not the least, neurological favourable prognosis.

Our patients, after surgical treatment, were already able to start a liquid nourishment in the first post-operative day, and a normal diet during the following day. The mean stay in the hospital was 4 days. Nine of the 11 patients had benefit from this procedure immediately, with resolution of dysphagia and quick recovery of weight after having left the hospital. One patient, with previous brain ictus, has enjoyed only a partial improvement of symptoms, maintaining a light dysphagia for solid food because of a deficiency of the pharyngeal pump, and occasional respiratory symptoms.

In one case we performed contemporary a total thyroidectomy, but the patient had a respiratory crisis immediately after the operation caused by an edema of the arytenoid cartilages. The patient was discharged in the tenth post-operative day after normalization of respiratory symptoms. The follow-up during the following weeks allowed to verify the resolution of dysphagia even in this case.

All patients underwent a cine-esophagogram after 4 weeks to verify the functional results.

Case 1.—In 2007 we admitted a woman 66 aged year-old, with about 6 months of cervical dysphagia, with immediate regurgitation food, solid and liquid, through the nasopharynx, and significant weight loss (25 kg).

The first diagnostic test for patient was cine-esophagogram, positive for esophageal dyskinesia and small aspiration of the Barium (Figure 2), negative for presence of an associated oesophageal diverticulum and gastro-esophageal reflux.

An esophageal manometry was performed, to verify the presence of hypertonia of UES and the existence of an efficient pharyngeal pump that is considered necessary to perform the myotomy of cricopharyngeal muscle.

An esophageal-gastroduodenal endoscopy (EGDS) completed the diagnosis of suspected, giving us the possibility to exclude organic diseases able to cause dysphagia such as tumours, peptic lesions, etc.

The patient was able to swallow liquids in first day post-surgery, normal diet the next day. The stay was 3 days.

Case 2.—A 43-year-old man was referred to our surgery department with complaints of dysphagia for both solids and liquids.[5] The symptom was non-progressive. He also had recurrent immediate vomiting and regurgitated food. He had considerable weight loss. The physical examination was unremarkable. Ultrasound scan of the abdomen showed normal findings. Upper gastrointestinal endoscopy showed a normal esophageal body and normal peristalsis was seen. The upper esophageal sphincter was tightly closed. With gentle pressure, the endoscopist was able to negotiate the endoscope into the esophagus.. Retroflexion of the endoscope revealed no mass lesion in the esophagogastric segment.

All of these features pointed towards the diagnosis of primary (idiopathic) dyskinesia of UES.

Barium swallow in this patient not showed dilated esophagus ("bird beak" appearance of the distal esophagus was not present) and an esophageal air fluid level.

Since the clinical history, examination, upper gastrointestinal endoscopy and barium swallow X-ray were suggestive of Dyskinesia of UES and pneumatic balloon dilatation of the upper esophageal sphincter was planned.[6, 7]

Under sedation, pneumatic balloon dilatation of the upper esophageal was carried out. The procedure was performed under endoscopic vision. The balloon was placed across the upper esophageal sphincter and inflated. The balloon was kept in the

Figure 3.—Cine-esophagram control showed not complete expansion of UES causing Dyskinesia.

Figure 4.—Cine-esophagram control after 4 weeks, showed expansion of UES.

inflated position for 2 minutes. With the help of the endoscope, active bleeding was seen, and also the successful dilatation of the upper esophageal sphincter. There were no procedure-related complications.

The patient was able to swallow liquids in first day post-endoscopy, then resume normal diet in next days. The stay was 1 day.

Unfortunately, the cine-esophagogram control after 4 weeks, showed not complete expansion of UES (Figure 3), and shortly afterwards the patient showed the reappearance of signs and symptoms of dysphagia. For this reason the decision was agreed, in accordance with the patient, to perform a surgical treatment, hesitate to make good results in the short and long term, as shown by cine-esophagogram control after 4 weeks, showed expansion of UES (Figure 4). The Authors strongly believe in surgery for a cure of Dyskinesia of UES, and in this case the patient, after the unfortunately endoscopy-pneumatic balloon therapy, showed adequate force of the lingual muscle, pharyngeal pump efficiency, integrity mechanisms governing the beginning of the voluntary swallowing, hypertension of UES, conservation of esophageal peristalsis in the absence of GERD and last but not least, neurological favourable prognosis, all parameters that could make surgery first therapeutic act.

Discussion

Cervical dysphagia, or the sensation of difficult cervical transit of food, is the main symptom that characterized all the motor disorders of UES.

These functional diseases are known to be often complex, and the most frequent of them is the Dyskinesia of cricopharyngeal muscle, that doesn't relax or that relaxes incompletely, similarly to what happens in Achalasia of LES; beside the motor disorder may consist of pharyngo-oesophageal incoordination. This causes a functional obstacle at the UES able to determine dysphagia for solid and/or liquid intake, and sometime inhalation of food through upper airways and the possibility of ab-ingestis pneumonia. The pressure increasement upstream the obstacle can progressively cause the herniation of pharyngeal mucosa through

a less resistant area known as Killian's triangle, and the forming of an oesophageal diverticulum.

Several pathological conditions can lead to a Dyskinesia of UES.

We use distinguish Primary forms (Idiopathic Dyskinesia of UES) and Secondary forms due to gastro-oesophageal reflux (the irritation caused by the acid reflux can lead to spasm of UES), neurological causes like ictus cerebri (oropharyngeal akinesia, sphincteric Dyskinesia), stroke, head trauma, brain tumours, Central Nervous System inflammatory processes, dementia, etc, and chronic neuromiogenic diseases like miastenia gravis, bulbar poliomyelitis, oropharyngeal muscle dystrophy.

Dyskinesia of UES, associated or less to the presence of an esophageal diverticulum, can cause important consequences on the nourishment of the patient who often comes to the attention of the surgeon in poor nutritional conditions. Beside it is an insidious disease, because often misunderstood and underestimated, and some patient is frequently considered suffering from psychiatric pathology, with obvious diagnosis delay.

The first diagnostic test for patients, who present symptoms suggesting an esophageal dyskinesia, is cine-esophagogram. It is an atraumatic diagnostic test able to reveal the spasm of UES, and the eventual presence of an associated oesophageal diverticulum. This test, extended to the rest of the esophagus, allows to discover the eventual presence of a gastro-esophageal reflux, that is actually considered a contraindication to the myotomy of UES.

It is strictly necessary to perform also an esophageal manometry, to verify the presence of hypertonia of UES and the existence of an efficient pharyngeal pump that is considered necessary to perform the myotomy of cricopharyngeal muscle.

In the forms of Dyskinesia of UES that are consequence of neurological diseases, it is mandatory to verify the efficiency of the mechanism that allows the beginning of voluntary swallowing, and maintenance of an appropriate propulsive strength of the tongue.

An EGDS completes the diagnosis, giving us the possibility to exclude organic diseases able to cause dysphagia such as tumours, peptic lesions, etc.

The Authors, for a cure surgery, take into account an accurate selection of patients; the same must be have pharyngeal pump efficiency, demonstration of obstacle to transit-bolus, hypertension of UES by manometry and conservation of esophageal peristalsis in the absence of GERD.

According to our actual guide-lines the gold-standard treatment of many kinds of Dyskinesia of UES is surgical, and consist of myotomy of cricopharyngeal muscle, associated or less to a diverticulectomy; however the performing of this kind of procedure requires a rigid selection of patients.

The surgical technique we use consist of an oblique incision along the anterior border of the right sternomastoid muscle, and after divarication of sternocleidomastoid muscle and of jugular and carotid vessels, and after isolation and division of omohyoid muscle and inferior thyroid artery, we perform the myotomy of cricopharyngeal muscle (Figure 5) that should be about 5 cm long.

In case of associated esophageal diverticulum, we use to perform diverticulectomy indifferently either using a stapler device (TA 30), or sewing the muscular breach with separate stitches.

Myotomy is an excellent procedure for patients with hypertensive and incoordination of UES.

Figure 5.—Intraoperative image showing myotomy of cricopharyngeal muscle (arrows).

Other techniques that we use, depending on the case reports, are: Endoscopic Approach (often in case of associated Zenker diverticulum, especially in patients with high risk), Pneumatic dilatation of the UES (in the case of fibrosis cricopharyngeal) or Approach with Toxin Botulinum (infiltration transcervical cricopharyngeal or by endoscopic).

Conclusions

The Authors emphasize the importance of a careful selection of patients undergoing surgical treatment (myotomy of the cricopharyngeal muscle), being decisive, in personal experience, in all patients treated, without abandoning alternative operative techniques, also in agreement with the patient[8].

Doing an accurate clinical and instrumental examination, is vital in some cases such as secondary Dyskinesia of UES to neurological causes, in which case surgical therapy not only has the potential to cure the patient, but also to save it (importance of communication Surgeon-Neurologist).

The follow-up (on-going), up to now, showed no complications or cases of reoperation, making sure the authors of the efficacy of the surgical treatment.

Riassunto

La Discinesia esofagea è un disturbo del movimento che può manifestarsi lungo l'intero esofago, dallo sfintere esofageo superiore allo sfintere esofageo inferiore. Nella nostra esperienza, l'atto chirurgico prevedente una miotomia, è l'unica procedura per il trattamento di malattie funzionali come la Discinesia dell'UES (sfintere esofageo superiore). L'efficienza della pompa faringea, la presenza di UES iperteso e la conservazione della peristalsi esofagea, in assenza di GERD (Malattia da Reflusso Gastro-Esofageo), sono condizioni imprescindibili per considerare un paziente eleggibile chirurgicamente. Gli Autori sottolineano l›importanza di una attenta selezione dei pazienti sottoposti al trattamento chirurgico (miotomia del muscolo cricofaringeo), essendo decisivo, nella esperienza personale, in tutti i pazienti trattati, senza abbandonare tecniche operative alternative, anche in accordo con il paziente. Dal gennaio 2006 al dicembre 2011, gli Autori hanno trattato 11 pazienti, di cui 9 affetti da Discinesia idiopatica dell'UES, 2 affetti da discinesia dell'UES secondaria a ictus cerebri; quattro di loro presentavano un diverticolo di Zenker associato, in 1 caso era presente un gozzo associato; di questi, l'autore riporta la clinica di due pazienti, clinicamente e strumentalmente più rappresentativi. In tutti questi casi abbiamo effettuato una miotomia extramucosa del muscolo cricofaringeo attraverso un approccio laterocervicale destro. Tutti i pazienti da noi trattati, hanno avuto benefici precoci, con risoluzione della disfagia; uno dei pazienti con ictus cerebri, ha goduto di miglioramento solo parziale dei sintomi; in un caso abbiamo eseguito una tiroidectomia contemporanea. Gli autori ritengono che il trattamento chirurgico sia l'unica procedura, dopo accurata selezione dei pazienti, capace di portare i pazienti verso una cura. Il follow-up (in corso), fino ad ora, non ha evidenziato complicazioni o casi di reintervento, rafforzando l'idea degli autori, riguardo l›efficacia del trattamento chirurgico.

Parole chiave: Sfintere esfageo superiore - Disturbi del movimento - Trattamento chirurgico.

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