

SURGICAL TREATMENT ANALYSIS OF IDIOPATHIC ESOPHAGEAL ACHALASIA

Análise da terapêutica cirúrgica da acalásia idiopática do esôfago

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ABSTRACT - Background: Idiopathic esophageal achalasia is an inflammatory disease of unknown origin, characterized by aperistalsis of the esophageal body and failure of the lower esophageal sphincter in response to swallowing, with consequent dysphagia. **Aim:** To demonstrate the results of surgical therapy in these patients, evaluating the occurred local and systemic complications. **Methods:** Were studied retrospectively 32 patients, 22 of whom presented non-advanced stage of the disease (Stage I/II) and 10 with advanced disease (Stage III/IV). All of them had the clinical conditions to be submitted to surgery. The diagnoses were done by clinical, endoscopic, cardiologic, radiologic and esophageal manometry analysis. Pre-surgical evaluation was done with a questionnaire based on the most predisposing factors in the development of the disease and the surgical indication was based on the stage of the disease. **Results:** The patients with non-advanced stages were submitted to cardiomyotomy with fundoplication, wherein in the post-surgical early assessment, only one (4,4%) presented pulmonary infection, but had a good outcome. In patients with advanced disease, seven were submitted to esophageal mucosectomy preserving the muscular layer, wherein one patient (14,2%) presented dehiscence of gastric cervical esophagus anastomosis as well as pulmonary infection; all of these complications were resolved with proper specific treatment; the other three patients with advanced stage were submitted to transmediastinal esophagectomy; two of them presented hydropneumothorax with good evolution, and one of them also presented fistula of the cervical esophagogastric anastomosis, but with spontaneous healing after conservative treatment and nutritional support. The two patients with fistula of the cervical anastomosis progressed to stenosis, with good results after endoscopic dilations. In the medium and long term assessment done in 23 patients, all of them reported improvement in life quality, with return to swallowing. **Conclusion:** The strategy proposed for the surgical treatment of idiopathic esophageal achalasia according to the stages of the disease was of great value, due to post-surgical low morbidity complications and proper recovery of swallowing.

HEADINGS - Megaesophagus.
Esophagectomy. Idiopathic achalasia.

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RESUMO - Racional: A acalásia idiopática do esôfago é doença inflamatória de causa desconhecida, caracterizada por aperistalse do corpo do esôfago e falha do relaxamento do esfíncter esofágico inferior em resposta às deglutições, com consequente disfagia. **Objetivo:** Demonstrar os resultados da terapêutica cirúrgica desses pacientes, avaliando suas complicações locais e sistêmicas. **Métodos-** Foram estudados retrospectivamente 32 pacientes portadores de acalásia idiopática do esôfago, sendo 22 com doença não avançada (Grau I/II) e 10 com doença avançada (Grau III/IV); todos tinham condições clínicas de serem submetidos à terapêutica cirúrgica. O diagnóstico foi realizado por meio de análise clínica, endoscópica, cardiológica, radiológica e manométrica. Foi realizada avaliação pré-operatória com questionário baseado nos fatores mais predisponentes ao desenvolvimento da doença, e a indicação da técnica cirúrgica foi baseada no grau da lesão. **Resultados:** Os pacientes com doença não avançada foram submetidos à cardiomiectomia com funduplicatura, sendo que na avaliação precoce do pós-operatório apenas um deles (4,4%) apresentou infecção pulmonar, mas com boa evolução. Os pacientes com doença avançada em sete foi realizada a mucosectomia esofágica com conservação da túnica muscular, sendo que um paciente (14,2%) apresentou deiscência da anastomose esofagogástrica cervical e também infecção pulmonar, tendo ambas complicações sido resolvidas com tratamento específico; os outros três com doença avançada foram submetidos à esofagectomia transmediastinal, sendo que dois apresentaram hidropneumotórax, com boa evolução; um destes pacientes também apresentou fistula da anastomose esofagogástrica cervical, mas com fechamento espontâneo após tratamento conservador e suporte nutricional. Os dois pacientes que apresentaram fistula da anastomose cervical, evoluíram com estenose, mas com boa evolução após dilatações endoscópicas. Na avaliação a médio e longo prazos realizada em 23 pacientes, todos relataram acentuada melhora na qualidade de vida com retorno da deglutição. **Conclusão:** O tratamento cirúrgico proposto da acalásia idiopática do esôfago de acordo com grau da doença foi de grande valia, devido às complicações pós-operatórias presentes serem de baixa morbidade, além de proporcionar retorno adequado da deglutição.

DESCRIPTORIOS - Megaesôfago.
Esophagectomia. Acalásia idiopática.

INTRODUCTION

Idiopathic esophageal achalasia (IEA) is an inflammatory disease of unknown origin, characterized by aperistalsis of the esophageal body and failure of the lower esophageal sphincter in response to swallowing, with consequent dysphagia; so, the patient may progress to malnutrition^{16,21,22,29}.

Amongst the main causes that may predispose this disease are lesions by toxic agent due to prolonged exposure to potent chemicals - herbicides being the most reported^{2,16,22}-, autoimmune disease, such as collagen-related diseases^{10,11,14}, previous viral infections -especially those of childhood such as varicella, measles, mumps and type 1 herpes virus^{13,22}-, family history with this disease^{17,30}, emotional factor with continuous use of psychiatric drugs¹¹⁹, prolonged use of tobacco and alcoholism^{2,10,19}.

Some studies have demonstrated comparatively that there are similarities and differences between idiopathic and chagasic achalasia. As for the destruction of the esophageal intramural nerve plexus, it is considered the same in both diseases, although this is not a well-accepted consensus^{10,16,21}. On the other hand, the lower esophageal sphincter is usually more hypertensive in the idiopathic achalasia than in the chagasic disease, and the esophagus dilatation is more intense in the latter^{10,16}.

The diagnosis of IEA is done after the exclusion of the possibility of Chagas disease origin, that is, negative serology for *Trypanosoma cruzi*, the non-association with megacolon, the exclusion of heart disease and absence of epidemiologic antecedent for Chagas disease^{2,10,14,16,19,21}.

IEA is found practically in every country, being able to attack any race, with variable incidence of 1 to 3 cases out of 100.000 inhabitants per year^{2,16,19,21}. This disease is little known in Brazil, because of the existence of the esophagopathy secondary to Chagas disease, endemic in many states, presenting signs and symptoms similar to those of IEA, which makes it difficult to estimate the exact incidence of this disease.

Thus, in the national scenario, because few authors referred experience with surgical treatment of IEA, the objective of this study is to demonstrate the indication and results of surgical treatment on it, regarding mainly local and systemic complications.

METHOD

Patients

From January 1993 to December 2013, 32 patients with IEA were studied at the Department of Thoracic Surgery of Celso Pierro Hospital, Pontifical Catholic University of Campinas, Brazil. There was a male predominance in 21 patients (65,6%) with ages ranging from 29 to 63 years old, average of 42,3 years old. All of them had indication for surgical treatment.

Preoperative evaluation

IEA diagnosis was done through:

a) Clinical evaluation - All patients presented progressive dysphagia for solid and pasty food for three to 11 years, with average of 5,7 years. Twelve also reported to have had frequent regurgitation in the past 24 months; all of them reported weight loss ranging from 5 to 18 kg. No patient reported to have progressive constipation or previous history of fecaloma or volvulus of sigmoid colon. Twenty four patients were from Campinas region, São Paulo state, refereeing no contact with endemic regions of Chagas disease. The other patients were from countryside regions of the north/northeast of Brazil, but reported that they were not contaminated with the triatomid transmissor. Thirteen patients reported to have contact with herbicides for having worked in coffee and cotton plantations for seven to 20 years; nine of the same patients reported varicella and/or mumps. Two other patients reported that besides varicella, one

had hepatitis A and another hepatitis B. Three reported that uncles or grandparents presented achalasia; two of these also used benzodiazepine medicines for emotional causes. Twenty three patients reported smoking (average of 20 cigarettes per day) from 12 to 26 years, 14 of whom reported to have quit in the past 8-14 years and the remaining maintain the habit. Eighteen patients reported alcoholism, drinking from one to two doses of alcoholic distillate per day with time ranging from 13 to 22 years, nine of whom reported to maintain this habit. In all immunofluorescence for Chagas disease with three samples was negative.

b) Heart evaluation - This evaluation excluded all the patients with chagasic heart disease.

c) Radiographic evaluation - Esophagogram showed achalasia with dilatation stages I and II in 22 patients (68,7%) and in the remaining 10 (31,3%) with stages III and IV, according to the classification of the megaesophagus proposed by Rezende et al.²⁵. Barium enema was done in 24 patients, who did not present megacolon.

d) Endoscopic evaluation - Showed the esophagus with an increased diameter without presenting associated lesions in 10 patients and in the other 22 did not show any abnormality.

e) Manometric evaluation - Was done in 17 patients, four presented achalasia with large esophagus where there was lack of incomplete relaxation of the lower esophageal sphincter. In the other 13 patients, all with non-advanced stages of the disease, this exam showed lack of incomplete relaxation of the lower esophageal sphincter with a little hindrance in the esophageal body contractility.

In all patients, clinical and nutritional evaluations were performed, and four patients needed nutritional support with enteral feeding 15 to 20 days prior to the proposed day of surgery, because these patients presented greater weight loss of 10% in relation to the regular weight.

Surgical technique

The indication for surgery was selected according to the stage of achalasia. Thus, for the less advanced stages (stage I and II - 22 patients), the recommended surgical technique was cardiomyotomy with anterior fundoplication. For seven patients with megaesophagus stages III/IV, the recommended surgical technique was esophageal mucosectomy preserving the muscular layer¹⁴; three were submitted to transmediastinal esophagectomy¹⁵.

Postoperative evaluation

This evaluation was in relation to:

Systemic complications, notably cardiovascular, respiratory and infectious ones. The diagnoses were based on daily clinical progress of the patients and on results from laboratory and imaging exams when necessary.

Local complications, related mainly with the dehiscence and stenosis of the gastroesophageal suture. Regarding the dehiscence of the gastroesophageal suture with consequent fistula, the diagnosis was of clinical origin, through the observation of salivary secretion output around the cervical region until the 7th postoperative day. As for the stenosis of the esophagogastric anastomosis, the diagnosis was also of clinical origin, oriented by dysphagia, mainly from the 30th postoperative day and corroborated with radiological contrast exams of the pharynx and endoscopy.

c) Life quality, in this item, the postoperative day was considered, in which the patients started with normal swallowing and in case of dysphagia, its level was evaluated if it was mild (solid food), moderate (pasty food) and intense (liquids).

RESULTS

In the early assessment until the 30th postoperative day, the patients submitted to cardiomyotomy did not present any local complication, having initiated oral diet initially progressing from liquid to solid after the 2nd postoperative day with good acceptance; one of them (4,0%) presented pulmonary infection, but with good progress after specific clinical treatment.

Of the three patients submitted to transmediastinal esophagectomy, two progressed to hydropneumothorax, but with good results after thorax drainage; one presented fistula of the cervical esophagogastric anastomosis in the 5th postoperative day, but with good outcome after conservative treatment and nutritional support with diet through jejunostomy until the 21st postoperative day. In this day, lacking clinical evidence of digestive fistula at the cervical level, contrasted X-ray was performed and with no extravasation at anastomosis, oral progressive diet was offered related to patient's acceptance.

Of the seven patients submitted to esophageal mucosectomy, one patient presented two complications, being one fistula with secretion output by the drain in the cervical region from the 2nd postoperative day, consequent to dehiscence of the cervical esophagogastric anastomosis; due it was on early postoperative period, a reoperation was indicated with partial resection of the anastomosis and new suture being performed, with good evolution. In the 7th day of this reoperation, a contrasted X-ray was performed and not showing any anastomotic extravasation was introduced oral progressive diet, according to the patient's acceptance. Another complication that the same patient presented was pulmonary infection after the 5th postoperative day of the 2nd surgical intervention, with good clinical evolution.

The two patients that presented cervical fistula caused by dehiscence of the cervical esophagogastric anastomosis, progressed to stenosis between the 29th to 47th postoperative day, confirmed by contrasted X-ray and endoscopy. These patients were submitted to four to seven sessions of endoscopic dilations with good outcomes.

The other seven patients with achalasia stage III/IV, six submitted to esophageal mucosectomy and one to transmediastinal esophagectomy, did not present any systemic or local complications. These patients started oral progressive diet from the 7th postoperative day, after the contrasted X-ray did not show any cervical contrast extravasation.

In the medium and long term postoperative evaluations between six months to 18 years (average of 7,6 years) done with 15 patients with non-advanced stage of the disease submitted to cardiomyotomy, reported to have a good life quality for having restored proper swallowing for solid food.

In the same period, eight patients with the advanced stage of the disease were also evaluated. They were submitted to esophagectomy and/or esophageal mucosectomy and also reported proper swallowing and return to their daily activities; three of them, however, reported intermittent regurgitation, without compromising their life quality.

DISCUSSION

Although many factors related to IEA developing have been suggested, the basic alteration is a muscular defect; however, its place of origin and its failure mechanism are still very debatable. Alterations in both intrinsic and the extrinsic nervous system of the esophagus have been demonstrated, besides the abnormalities in the number of neurons^{10,11,14,29}.

Histological studies have demonstrated that there are lesions present in the intramural plexus of the esophagus, becoming more severe as the disease progresses. These alterations are more prevalent in the body of the esophagus, occurring from mononuclear inflammatory infiltrate of the Auerbach's plexus to the total substitution of the intramural ganglion cells by the fibrous tissue^{10,13,27,29}.

More recently, it has been demonstrated in the physiopathology of IEA that a previous inflammation of the myenteric nerve plexus of the esophagus must occur, consequent to several factors and that it could occur as a later autoimmune chronic response in people with greater genetic predisposition and it could in sequence destroy the esophageal innervation^{14,22}. This

might explain the diagnostic potential of IEA in the patients in this study, because several of them reported in their antecedents one or more factors that could predispose a previous inflammation of the intramural nerve plexus of the esophagus, such as prolonged contact with chemicals, use of psychiatric drugs, previous viral infections, long-term smoking and alcohol consumption. Such factors have already been studied by other authors^{2,10,11,14,16,17,19,22,30}.

The similarity between idiopathic esophageal achalasia and chagasic achalasia from the point of view of symptoms, radiological and histopathological studies is very large, making the two diseases practically the same in Brazil, thus making the diagnosis of IEA difficult. In other continents such as North America, Europe and Asia, in which IEA is found rather frequently, Chagas disease does not exist^{2,16,19}.

The radiological findings of IEA are similar to those of chagasic megaesophagus, but more advanced forms have been found in greater prevalence in the latter^{2,9,16,19}. Although there is no statistical significance because the studied sample was not so relevant, the data served to confirm, because there was the predominance of the non-advanced forms present in 68,7% of the patients in the present series.

Because the non-advanced form of the disease is more present in IEA, they have a shorter course of dysphagia in comparison to Chagas disease, as Oliveira et al. have well demonstrated that IEA patients presented an average of 4,4 years with this symptom, whereas Chagas disease 10,7 years¹⁹. They also correlated the age and demonstrated that IEA patients were younger, with average of 37,6 years old in contrast to 49,4 years old for chagasic patients.

These factors were also similar in this study, because the average of dysphagia in the patients was of 5,7 years with average age of 42,3 years old.

The manometric alterations of the IEA are very similar to those found in chagasic esophagopathy, with esophageal hypomotility, characterized by low amplitude contraction waves and with lower esophageal sphincter presenting absence or decrease in the relaxation in swallowing^{2,16,19}. These alterations were also demonstrated in the patients of this study in whom this exam of performed.

Although the endoscopy performed in patients in this study has not demonstrated lesions in the esophageal lumen, the performance of this exam is fundamental, because as in the chagasic megaesophagus, can also occur chronic ulcerative esophagitis secondary to chronic stasis of food, causing long-term leucoplasia, epithelial dysplasia or even progressing to carcinoma with variable incidences from 0,3 to 20%^{2,8}.

Regarding therapy, in countries where IEA is prevalent, surgery is not always the established treatment. The preference for many authors initially was forced dilatation of the cardia, because they thought that the dilations would benefit the patients, recommending surgical treatment only for those who did not present improvement or that had recurrence of symptoms^{1,23,28}.

Because of the improvements of the surgical technique, as well as today's better pre and postoperative surgical care, many authors recommend surgical treatment in most patients with IEA as well⁷.

Many conservative surgical techniques or resections have already been recommended to treat IEA, esophagocardiomyotomy described by Heller¹² in 1913 being the most utilized. Today, just as in the chagasic megaesophagus, this procedure has a more selective indication recommended in non-advanced stages of the disease (I and II) and associated with the anti-reflux valve with good outcomes in the medium and long-term evaluations and minimum rates of morbidity and mortality^{2,15,26}. More recently, with the advent of laparoscopic surgery demonstrating advantages of minimally invasive surgery, there was also evidence that this access offers good results in cardiomyotomy in patients with non-advanced IEA¹⁸.

The data serves to confirm positive results in the patients of this series with non-advanced stages of the disease, because in the early assessment in which the cardiomyotomy

was performed, no patient presented local complications and in the medium and long term evaluations in 15 patients, all of them reported to be satisfied with the surgery, because besides having returned to their usual activities, they also presented normal swallowing, improving their life quality.

As for the advanced stages of the disease both stage III and mainly stage IV, as well as in case of chagasic megaesophagus, esophagectomy has been the preferred choice because it acts directly in the physiopathology of the disease, being the transmediastinal technique recommended by Pinotti²⁴ the most widely used technique. However, in a more critical analysis, it has been demonstrated that this technique is not free from complications. Among them, it can be emphasized pleuromediastinal complications translated to hemomediastinum and hemothorax, which can contribute to great morbidity in the postoperative time^{5,20}. This can happen, as the advanced megaesophagus presents periesophagitis, making this organ adhere to important structures of the mediastinum, which may predispose lesions during dissection.

This difficulty induced Aquino⁶ to propose the esophageal mucosectomy preserving the muscular layer and transposition of the stomach inside the muscular layer of the esophagus for the reconstruction of the digestive transit and anastomosis of the stomach to the cervical esophageal stump. Therefore, both in the early and late assessment, it has been demonstrated in patients with advanced chagasic megaesophagus a minor index of complications related to the transmediastinal esophagectomy without thoracotomy, for not transgressing the mediastinum during dissection of the esophagus^{3,4}.

Due to the good results obtained with the esophageal mucosectomy in patients with advanced chagasic megaesophagus, made these authors to recommend this procedure to seven patients that presented advanced IEA. Although the results are not very significant, due to its small sample, the esophageal mucosectomy seems to be more proper, because no patient presented pleuromediastinal complications, although one of them progressed to fistula to the esophagogastric anastomosis and also pulmonary infection, but presenting good results with early and proper treatment; as for the other patients, two of the three submitted to transmediastinal esophagectomy presented pleural complications.

Hence, as IEA is confused with chagasic megaesophagus, it is difficult to identify its carriers most of the time. Some antecedents present in the previous history of patients in this study and criteria used to identify already cited IEA cases seem to be logical to exclude esophagopathy of chagasic origin, as other authors have also been demonstrated^{2,19}.

The introduction of new serological techniques with high sensitivity for Chagas disease diagnosis will permit the exclusion of chagasic patients and facilitate the identification of IEA carriers in the national scenario^{11,14,29}.

CONCLUSION

The strategy proposed for the surgical treatment of idiopathic esophageal achalasia according to the stages of the disease was of great value, due to post-surgical low morbidity complications and proper recovery of swallowing.

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