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Oesophageal Stenosis Dilatation Through Retrograde Trans-gastrostomal Approach in a Patient with Systemic Scleroderma

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Rezumat

Dilatația stenozei esofagiene prin abord retrograd trans-gastrostomă la un pacient cu sclerodermie sistemică

Scop: Lucrarea prezintă soluționarea chirurgicală a unei stenoze esofagiene folosind un montaj de dilatație pneumatică prin abord transgastrostomie, la o pacientă pluritarată, cu handicapuri invalidante, malnutriție secundară severă, cunoscută cu sclerodermie.
Materiale și metoda: La internare, pacienta prezenta casexie (37 kg, 170 cm), facies bizantin caracteristic, microstomie, rezorția falangelor distale la ambele mâini și disfagie completă, cu limitarea deschiderii gurii. Examenul baritat arată stenoză esofagiană distală, cu dilatare importantă a esofagului supraiacent.
Rezultate: S-a poziționat o gastrostomie de alimentare, procedeu Gavriliu, sub anestezie generală cu intubație transtraheostoma. După 3 ani, pacienta revine în clinica noastră, cu ameliorarea semnificativă a statusului metabolic (59 kg) și solicită o soluție terapeutică pentru a reveni la alimentația pe cale naturală. Dificultățile tehnice întâlnite în acest caz au fost determinate de ocluzia limitată a gurii, care nu permite dilația cu balon sau bujienajul pe cale oro-faringiană, și nici intubația oro-traheală. Valorificând prezența gastrostomei, cunoscând procedeul von Hacker de dilatație mecanică și utilizarea kitului metalic Key Med cu bile a creat cadrul tacticii și strategiei ghidării unui fir-ghid (mandren) radio-opac introdus prin gastrostoma, transstenotic și exteriozat la nivel oral. O sondă Foley modificată (procedeu personal) a fost atașată mandrenului ghid. Prin tracțiune retrogradă transstenotică, balonasul Foley a fost factorul pneumatic dilatator ce a permis ulterior dilatația facilă cu dilatatorul metalic Key Med, până la diametrul maxim. Evoluția postoperatorie a fost favorabilă, pacienta revenind la nutriția pe cale naturală.
Concluzii: Dispozitivul de dilatație pneumatică esofagiană permite prin tehnica chirurgicală prezentată o plastie blândă efectuată sub control radiologic și reduce riscul de accidente. Dilatația realizată a permis ulterior utilizarea kitului Key Med. Noutatea constă în adaptarea unei tehnici cunoscute la un montaj nou, brevetat, de dilatație pneumatică prin abord bidirecțional sub control radiologic pentru a soluționa un caz atipic.

Cuvinte cheie: sclerodermie, stenoză esofagiană, abord transgastrostomă

Abstract

Aim: The paper presents the surgical solving of an oesophageal stenosis, using a device of pneumatic dilatation with trans-gastrostomal approach, in a patient with multiple disabling handicaps, secondary severe malnutrition and previously diagnosed with scleroderma.
Materials and method: The patient was admitted with severe cachexia (37 kg, 170 cm), characteristic byzantine face with microstomy, distal phalanges resorption in both superior limbs and complete dysphagia, with limitation of mouth opening. The Barium swallow test revealed distal oesophageal stenosis, with an important dilation of the oesophagus above.

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Results: A gastrostoma was placed to allow nutrition (Gavriliu procedure), under general anaesthesia with trans-tracheostomal intubation. After 3 years, with her metabolic status improved (59 kg), the patient returned to our clinic asking for a solution for natural feeding. The technical difficulties in solving this case were determined by the limited mouth opening, which made anterograde oro-oesophageal balloon dilatation or bougienage impossible, as well as oro-tracheal intubation. Making use of the presence of the gastrostomal orifice, knowing von Hacker's mechanical dilation procedure and using the metallic Key Med kit with balls offered the possibility of the tactics and strategy of guiding a metallic guidewire introduced via the gastrostoma, then trans-stenotic and pulled out through the oral orifice. A modified Foley catheter (personal procedure) was attached to the initial catheter. The trans-stenotic retrograde traction of the Foley balloon was the pneumatic dilator factor that later allowed easy dilatation with the metallic dilator of the Key Med, to the maximum size. The follow-up showed good results, the patient returned to natural nutrition.
Conclusions: The device of oesophageal pneumatic dilatation allows, using the presented surgical technique, a gentle plasty done under radiological supervision and lowers the frequency of accidents. The dilation permits the following use of Key Med kit. The novelty consists in adapting a well-known technique to a new patented device of pneumatic dilation with bidirectional approach under radiological control, for solving this atypical case.

Key words: scleroderma, oesophageal stenosis, trans-gastrostomal approach

Introduction

Surgical solutions designed for solving cases with low incidence in clinical practice create the starting point for developing new branded procedures in the future. The intersection of a collagenosis such as systemic sclerosis (scleroderma) with digestive surgical pathology is extremely rare. A review of the literature describes a possible oesophageal involvement in scleroderma with several clinical aspects: progressive dysphagia, with oesophageal dismotility and hipomotility, and characteristic image of "stiff glass tube oesophagus", as shown by barium swallow. A frequently encountered pathological aspect in this collagenosis is gastro-oesophageal reflux disease, with peptic esophagitis, ulcer and even adenocarcinoma arising in Barrett's oesophagus (occasionally) (1,2).

The tissular alterations in systemic sclerosis proved to be an important risk factor in performing digestive anastomosis, implying technical intraoperative difficulties and also increased rate of postoperative complications. Intestinal malabsorption is also an associated deficiency that must be taken into account in the planning of a therapeutic strategy in case of an oesophageal stenosis in a patient with scleroderma.

Over time, our department has had the experience of solving several atypical cases, for which the specific therapeutic approach wasn't previously described in the clinical practice guidelines.

Case report

This article presents the case of a 34-year-old female, diagnosed with systemic sclerosis since 1990 and treated for the disease according to the protocols in use in our country (corticotherapy). The patient was admitted in the Superior Digestive Surgery Department with a poor medical condition due to severe cachexia (37 kg, 170 cm). The clinical exam revealed the following findings: byzantine face with microstomy, distal phalanges resorption in both superior limbs and complete dysphagia, with limitation of mouth opening (Fig. 1).

The Barium swallow revealed distal oesophageal stenosis, with an important dilation of the oesophagus above (Fig. 2).

Figure 1. *Clinical exam: byzantine face, microstomia*

Figure 2. *Barium swallow (initial exam): oesophageal stenosis, with an important dilation of the oesophagus above.*

The superior digestive endoscopy, performed with 0.9 diameter bronchoscope, revealed: oesophageal circular stenosis (5-6 mm diameter) at 35 cm from the superior dental archway, with white-coloured mucosa in the upper part and red for the segment under the stenosis. Dilatation of the oesophagus from above the stenosis was significant. The maxilla-facial exam showed severe limitation in opening the mouth, due to bilateral degeneration of the temporomandibular joints. 10 ultrasound physiotherapy sessions and mecanotherapy were prescribed, with limited improvement of the mobility.

Due to her poor physical condition and cachexia, a minimal surgical intervention for feeding was decided and an alimentary gastrostoma with peritoneal collar was performed for the complete dysphagia due to inferior oesophageal stenosis. A temporary tracheostomy was needed for the intervention, as orotracheal intubation was not possible due to the limited temporomandibular joint mobility. Intraoperatively, there was no evidence of morphological changes of the digestive tract typical for systemic sclerosis. (Fig. 3)

The postoperative evolution was satisfactory. The patient came for control in our clinic after 3 years and 2 months of nutrition through gastrostoma, with a weight of 69 kg, asking for a surgical re-evaluation and hoping to return to natural feeding.

Even with her metabolic status improved, the patient's condition did not allow an oesophagoplasty - due to the degeneracy of the temporomandibular joint with restriction of the mouth opening. On the other hand, the tissular alterations represent a risk factor for prolonged bleeding, increased time for wound healing and stenosis of the digestive anastomosis.

The decision to close the gastrostoma and create a solution for natural nutrition was taken for the following reasons:

1. *stable radiological aspect of the oesophageal stenosis (the same diameter in size) for 3 years (barium swallow);*
2. *the patient's infirmity in using her hands, which made her totally dependent to a third person for feeding through the gastrostoma;*
3. *the patient's expressed will;*
4. *the chance of improving the patient's biological status, with her multiple handicaps.*

The technical difficulties in solving this case were related to the impossibility of a comfortable opening of the temporomandibular joint, which caused:

1. *the impossibility of anterograde oro-oesophageal balloon dilation or bougienage;*
2. *the impossibility of oro-tracheal intubation;*
3. *in case of a new general anaesthesia needed for surgery, a new tracheostomy had to be performed on the old scar;*

The therapeutic strategy was the following: Valuating the presence of the gastrostomal orifice and using a technical variant of von Hacker's mechanical dilation procedure (3), but using the metallic Key Med with balls, an unclassified procedure was performed: oesophageal stenosis dilatation through retrograde trans-gastrostomal approach. (Fig. 4, 5)

We tried to introduce the dilator by the anterograde oro-pharyngeal way, but we could not pass it through the stenosis. Using a gastro-oesophageal retrograde endoscopic approach through the gastrostoma, we managed to introduce a radio-opaque guide-wire through the gastrostoma and, under radiological guidance, we succeeded to pass it through the stenotic zone of the inferior oesophagus.

This step of visualization and catheterization of the oesophagus with a metallic catheter (Fig. 6) ended by pulling out the guidewire through the oral orifice (Fig. 7, 8). From this moment, a bidirectional approach of the stenosis was possible. A Foley modified catheter (personal procedure) was attached to the initial guidewire and the contrast radiopaque substance was injected into the balloon (Fig. 9, 10). This way, the whole route until the level of the stenosis was visualized under radiological control.

Under radiological guidance, the balloon was placed at

Figure 3. *Postoperative aspect, with tracheostoma, limited mouth opening*

Figure 4. *X-ray exam with barium injected through gastrostoma*

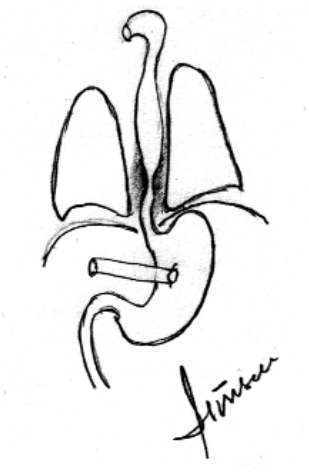

Figure 5. *The graphic scheme of the digestive system before removal of gastrostoma status*

Figure 6. *Gastro-oesophageal retrograde endoscopic approach through gastrostoma, with a guide-catheter introduced through gastrostoma and passed through the oesophageal stenosis*

Figure 7. *The guide-catheter is exteriorized through the mouth*

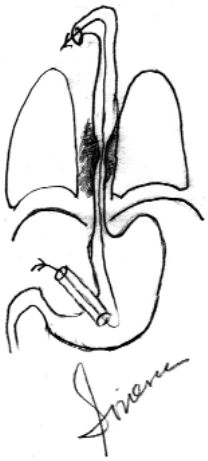

Figure 8. *The graphic scheme of the guide-catheter trans-gastrostoma exteriorized through the mouth*

Figure 9. *Using the modified Foley catheter*

Figure 10. *The Foley catheter is passed through the stenosis until the oral orifice*

Figure 11. *Radiological confirmation of the gentle pneumatic dilation with modified Foley catheter*

the level of the stenosis and progressive pneumatic gentle dilation was done, which permitted a true local plasty of the distal oesophageal stenosis (Fig. 11, 12).

Then, the metallic dilator of the Key Med kit was introduced without difficulties, until the maximum value in the kit (Fig. 13, 14).

The post-operative evolution was good, and the results were satisfactory at the 6 month follow-up. The radiological aspects are presented below (Fig. 15, 16 A,B).

Knowing the patient's will and the radiological findings, the surgical team decided to close the gastrostoma and to recommend natural feeding.

Discussions

Progressive systemic sclerosis (PSS) causes smooth muscle atrophy and fibrosis of the distal two-thirds of the oesophagus. Motility studies show reduced-amplitude or absent peristaltic contractions in this region and normal or decreased lower

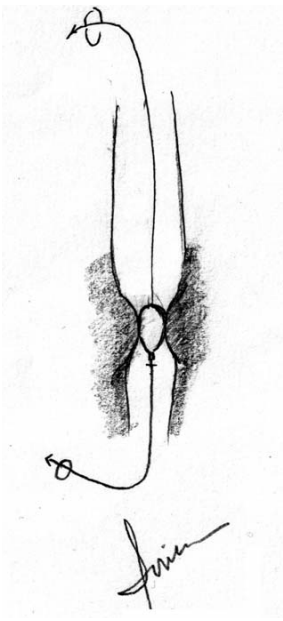

Figure 12. *The graphic scheme of the gentle pneumatic dilation*

Figure 13. *Metallic retrograde dilatation with Key Med kit – exteriorized at the gastrostoma level*

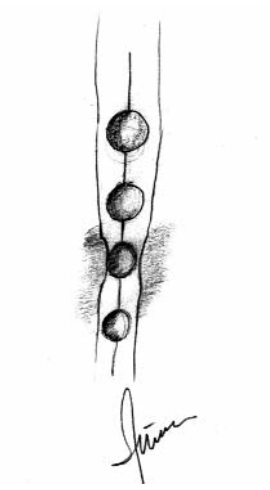

Figure 14. *Graphic scheme of the retrograde Key Med dilatation – done without any difficulties*

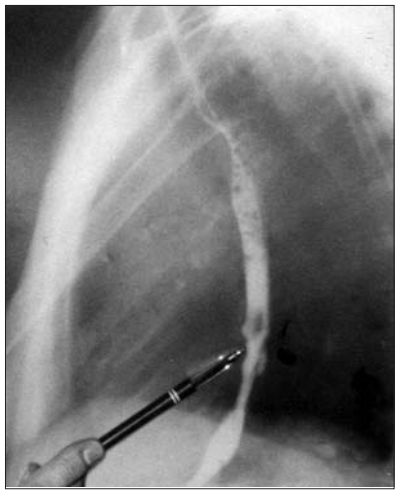

Figure 15. *Radiological postoperative status*

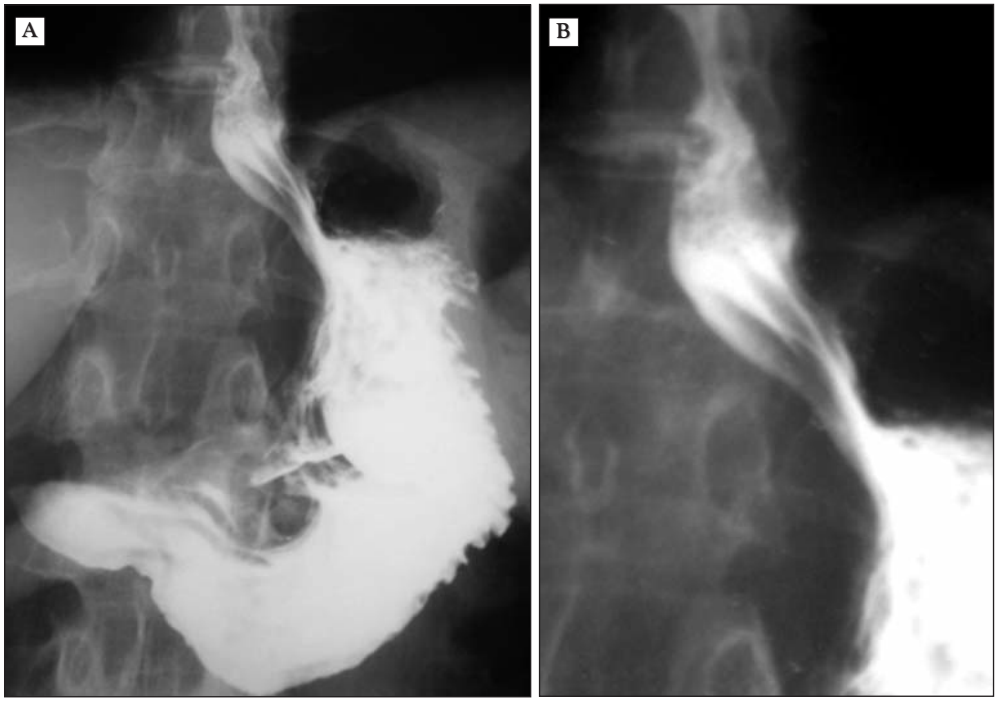

Figure 16. *(**A, B**) Barium swallow - late (6 month) postoperative aspect after closing of the gastrostoma*

oesophageal sphincter pressure (1). Scintigraphy is also useful for the study of the relationship between gastro-oesophageal reflux (GER) symptoms and oesophageal dysmotility in cases with endoscopically confirmed gastro-oesophageal reflux disease. (3) Long-term complications include strictures found in 17% to 29% of patients and Barrett oesophagus in 0% to 37%. (1)

In the presented case, there is a discrepancy between the lesions evidenced at the examination of the viscero-cranium, with bilateral temporo-mandibular ankylosis and the retractile collagen degeneration of the fingers (that caused total functional invalidity, the patient being 100% dependent of another person who takes care of her daily necessities), and on the other hand, the absence of the visceral sclerodermic alterations of the connective tissue. The typical "stiff glass tube oesophagus" is not present, on the contrary, we noticed its suprastenotic dilatation. We consider that the patient had an oesophageal ulcer, consecutive to gastro-oesophageal reflux – a frequently encountered feature associated to the oesophageal motility disorders in scleroderma. This ulcerative lesion evolved typically to distal oesophageal stenosis.

Performing a gastrostoma is a minimally invasive procedure which can be a salutary solution for feeding and improving the metabolic status in patients with systemic sclerosis and severe

malnutrition or cachexia due to dysphagia (5,6).

Regarding the therapeutic approach in distaloesophageal stenosis, endoscopic balloon dilatation and bougienage are considered the first choice. In young patients, the prognostic is reserved, as the strictures are difficult to dilate, requiring often several sessions and recurrences appear more frequently. In refractory cases, or in severe oesophageal dysplasia or suspicion of malignancy, distal oesophagectomy must be considered. The reconstruction of the digestive tube may be done using gastric material, jejunum or colon. One thing to take into account is that scleroderma may also affect the myenteric plexus of the latter two. (7,8,9)

In the presented case, there were no significant alterations of the oesophageal wall, but the temporo-mandibular bilateral ankylosis made conventional endoscopy technically impossible. This was the main reason to choose a non-standardized approach of the stricture. In a review of the literature, trans-gastrostomy interventional endoscopy is a new developing approach, used for different upper digestive tract pathologies (10), but there are very few evidences for treating distal oesophageal stenosis, using a double antegrade-retrograde rendez-vous technique (11).

Conclusions

Gentle pneumatic dilatation done under radiological supervision lowers the possibility of accidents; this first expansion also facilitates the later use of the Key Med kit. This retrograde trans-gastrostomal technique can be used for modelling the oesophageal stenosis, in cases in which the stenosis diameter allows catheterization of the oesophagus with a metallic tube.

This microinvasive surgery in the complex upper digestive segment seems to gain real importance - the hospitalization of those patients being really short and the main accidents described being almost zero.

The moment and the chosen strategy depend on the experience in the field. In this case, the experience of our department's staff is the one who permitted us to use not only standard surgical techniques, but also to adapt the surgical tactics and strategies in this complex oesophagus pathology, and to register a patent of a new pneumatic dilation device. The novelty with practical utility consists in adapting a well-known technique for a pneumatic dilation with bidirectional approach under radiological control, for solving this specific case.

In any surgical treatment performed in the oesophagus region, it is much better to have, right from the start, the opinion of a specialist with high experience in this field. As Prof. Dan Gavriliu always said: «The doctor who begins a repair treatment acting on the oesophagus must perfectly know all potential therapeutic alternatives and is bound to continue this surgical treatment until the problem is completely solved.»

References

1. Ebert EC. Esophageal disease in scleroderma. J Clin Gastroenterol. 2006;40(9):769-75.
2. Ntoumazios SK, Voulgari PV, Potsis K, Koutis E, Tsifetaki N, Assimakopoulos DA. Esophageal involvement in scleroderma: gastroesophageal reflux, the common problem. Semin Arthritis Rheum. 2006 Dec;36(3):173-81. Epub 2006 Oct 11.
3. Nakajima K, Inaki A, Hiramatsu T, Hasegawa M, Fujimoto M, Takehara K, et al. Esophageal transit scintigraphy and structured questionnaire in patients with systemic sclerosis with endoscopically proven reflux esophagitis. Ann Nucl Med. 2009;23(9):771-6. Epub 2009 Oct 24.
4. Earlam R, Cunha-Melo JR. Benign eosophageal strictures: historical and technical aspects of dilatation. Br J Surg. 1981; 68(12):829-36.
5. Das L, Bowden A, Cooper RG, Mitchell W, O'Sullivan M, Herrick AL. Percutaneous endoscopic gastrostomy feeding—a life-saving intervention in SSc–myositis overlap with pharyngeal dysfunction. Rheumatology (Oxford). 2012;51(8):1518-20. Epub 2012 Mar 29.
6. Fynne L, Kruse A, Borre M, Søndergaard K, Krogh K. Percutaneous endoscopic gastrostomy in patients with systemic sclerosis. Scand J Rheumatol. 2010;39(3):266-8.
7. Yekeler E, Gürsan N. Esophagectomy in scleroderma: report of a case. 2008;2(3):499-504. Epub 2008 Dec 5.
8. Jeyasingham KJ, Shields TW, Lo CJ, Ponn RB. Benign strictures of the esophagus; General Thoracic Surgery. Philadelphia: Lippincott Williams & Wilkins; 2000. p. 1865–1880.
9. Stummvoll GH. Current treatment options in systemic sclerosis (scleroderma). Acta Med Austriaca. 2002;29(1):14-9.
10. Nishiwaki S, Hatakeyama H, Iwashita M, Asaki H. Transgastrostomal observation and management using a an ultra-thin endoscope after percutaneous endoscopic gastrostomy, Therapeutic Gastrointestinal Endoscopy, Prof. Oliviu Pascu (Ed.), ISBN: 978-953-307-743-7, InTech, Available from: http://www.intechopen.com/books/therapeutic-gastrointestinal-endoscopy/transgastrostomalobservation-and-management-using-an-ultra-thin-endoscope-after-percutaneous-endosco.
11. Maple JT1, Petersen BT, Baron TH, Kasperbauer JL, Wong Kee Song LM, et al. Endoscopic management of radiation-induced complete upper esophageal obstruction with an antegrade-retrograde rendezvous technique. Gastrointest Endosc. 2006; 64(5):822-8.