

*G Chir Vol. 30 - n. 1/2 - pp. 17-20
Gennaio-Febbraio 2009*

casistica clinica

Acute onset of esophageal duplication cyst in adult. Case report

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SUMMARY: Acute onset of esophageal duplication cyst in adult. Case report.

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Introduction. Esophageal duplication (ED) cyst is unusual congenital disorder of the foregut, accounting for 10% to 15% of duplications of all foregut cysts. We report a case of esophageal duplication with acute clinical presentation, treated successfully with surgical resection.

Case report. 46-year-old man with acute dysphagia and gastroesophageal reflux of 2 weeks duration, showed submucosal bulging mass in the posterior wall of the middle third of oesophagus, fluid-filled cystic structure, dyshomogeneous, low-attenuation mass with smooth borders compatible with an hemorrhagic esophageal duplication cyst. With the suspect of acute presentation of a complicated esophageal cyst, the patient underwent surgery by right posterolateral thoracotomy. A gastrografin esophagogram was performed on third postoperative day and showed no leaks.

Discussion. Up to 80% of the esophageal duplication cysts are diagnosed in childhood and the majority of young patients develop symptoms, while symptomatic cyst into adult life is very rare (<7%). Acute onset is generally due to complications, i.e. intracystic hemorrhage, perforation, and infection, especially of the cyst with esophageal communication. Complete surgical excision by thoracotomy or thoracoscopy is the therapy of choice even if the patient is asymptomatic because of the risk of complications such as aspiration and bleeding.

Conclusions. The diagnosis and treatment of esophageal cysts is still evolving. The pathological diagnosis of oesophageal duplication requires the presence of the Ladd and Gross criteria. The infrequent acute onset doesn't modify the management and the surgical resection is the procedure of choice. The future of the treatment of esophageal cysts lies in the advancement of minimally invasive operative techniques.

RIASSUNTO: Esordio acuto di cisti da duplicazione esofagea nell'adulto. Caso clinico.

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Introduzione. La cisti da duplicazione esofagea è una rara malformazione congenita del tratto intestinale superiore (10-15% di tutte le duplicazioni intestinali). Riportiamo di seguito un caso di duplicazione esofagea con esordio clinico acuto in un adulto, trattata chirurgicamente con successo.

Caso clinico. Un uomo di 46 anni, con disfagia acuta e reflusso gastroesofageo da 2 settimane, risultava affetto all'esofagoduodenoscopia da una formazione della parete posteriore dell'esofago, a contenuto cistico, disomogenea, con quadro radiologico compatibile con una cisti da duplicazione esofagea a contenuto emorragico. Con il sospetto di una cisti esofagea complicata, il paziente è stato sottoposto a toracotomia posterolaterale destra al fine di enucleare per via smussa la cisti integra, mantenendo la tonaca muscolare. Un transito esofageo con mezzo iodato idrosolubile in III giornata postoperatoria non ha mostrato leakage.

Discussione. Più dell'80% delle cisti da duplicazione esofagea sono sintomatiche e diagnosticate nell'infanzia, mentre è raro (<7%) l'esordio clinico in età adulta. La presentazione acuta può essere dovuta ad emorragia intracistica, perforazione, infezione, specie nel caso di continuità con il lume. La completa escissione chirurgica toracotomica o toracoscopica è il trattamento di scelta, anche se la cisti è asintomatica, per il rischio di polmoniti da aspirazione di contenuto gastrico e di emorragia.

Conclusioni. La diagnosi e il trattamento delle duplicazioni esofagee sono in evoluzione. La ecografia endoscopica è il metodo di scelta per la diagnosi, da porre con certezza in accordo con i criteri di Ladd e Gross. La presentazione clinica acuta non modifica il management e la resezione chirurgica è il trattamento di scelta. Trattamenti mini-invasivi possono essere indicati in soggetti accuratamente selezionati.

**KEY WORDS: Oesophageal duplication - Acute onset - Adult - Surgery.
Duplicazione esofagea - Esordio acuto - Adulto - Chirurgia.**

Introduction

Oesophageal duplication (ED) cyst is unusual congenital disorder of the foregut, often asymptomatic. Some patients may present with dyspnoea, chest pain, or dysphagia. A diagnosis may be made with a combination of imaging technique. The standard management is surgical resection even for asymptomatic patients, considering possible complications during the natural course of the disease and because definitive diagnosis can be established only on surgical specimen.

We report a case of ED in adult with acute onset treated successfully with surgical resection.

Case report

A 46-year-old man presented with acute dysphagia and symptoms of gastroesophageal reflux of short duration (two weeks). Physical examination was normal.

Upper endoscopy revealed a submucosal bulging mass in the posterior wall of the middle oesophagus, synchronous with respiratory acts, lined by intact mucosa, narrowing the esophageal lumen. CT scan revealed a fluid-filled cystic structure, dyshomogeneous, low-attenuation mass with smooth borders in the esophageal wall (Fig. 1A) close to left bronchus and left atrium. Endoctrasonography (EUS) was compatible with an hemorrhagic oesophageal duplication cyst (Fig. 1B). With the suspect of complicated esophageal cyst, the patient was operated.

Through a right posterolateral thoracotomy (Fig. 1C), blunt dissection was used to enucleate the cyst. We carefully excised it preserving the muscle layer. Both vagal nerves were identified and preserved. Mucosal integrity was checked intraoperatively by air insufflation through the nasogastric tube previously inserted. The muscle layers of the oesophagus were approximated using 2-0 polypropylene interrupted stitches thus preventing pseudodiverticula formation in an area of disorganized muscular anatomy. A Gastrografin esophagogram was performed on third postoperative day and showed no leaks (Fig. 1D).

The postoperative course was uneventful: the patient swallowing was normal and he took liquids within third postoperative day.

Pathological examination showed a 6x8,5 cm cystic lesion with dual muscular layer and no cartilage or bronchial glands. The inner layer was composed of squamous epithelium with intramural huge area of hemorrhage.

Discussion

Duplication cysts are rare congenital anomalies of the foregut that were first described in 1711 by Blasius. In 1881, Roth described these cysts, which were divided into two categories: *a*) simple epithelial-lined cysts, and *b*) oesophageal duplication, both not usually communicating with the lumen of the oesophagus (1, 2). The ED account for 10% to 15% of duplications of all foregut cysts after the ileum with a male predominance. ED constitutes only 0.5-2.5% of all

oesophageal tumors, usually grouped with other benign lesions of the oesophagus. Although rare, malignant degeneration can occur (1).

In 60% of patients the duplication cysts are located in the lower oesophagus. The rest are distributed equally between the upper and middle third of the oesophagus (2). The embryologic basis of this disorder is poorly understood even if it is thought ED developing from the primitive foregut, around the fourth to eighth week of gestation, because failure of vacuolization of the primitive oesophagus during embryologic development. Of all oesophageal duplications, the cystic form is far more common than the tubular form (2).

ED may be attached to the oesophagus in a paraoesophageal location or may be intramural. Attached cysts often protruded into the posterior mediastinum. They are lined by non-keratinizing squamous or ciliated columnar epithelium: a double layer of smooth muscle in their walls and absence of cartilage are necessary findings to exclude a diagnosis of bronchial cyst.

Up to 80% of cysts are diagnosed in childhood and the majority of young patients develop symptoms, while it is extremely rare for a patient to remain asymptomatic into adult life (<7%) (2, 3). When symptoms occur, they are caused by compression of surrounding structures. ED can result in compression of the trachea with dyspnea or of the esophagus causing dysphagia. In the lower third of the oesophagus difficulty swallowing from compression is the most common symptom; in the upper third of the oesophagus respiratory difficulty from compression of the tracheobronchial tree is the most common symptom; and in the middle third of the oesophagus retrosternal chest pain and difficulty swallowing are the most common symptoms. Posterior cysts in the lower third of the oesophagus can cause cardiac arrhythmias (3). Acute onset is due to complication by intracystic hemorrhage, perforation, and infection, especially in those with oesophageal communication.

The suspected diagnosis is often on chest X-ray (mediastinum enlargement). CT and endoscopy usually are insufficient by themselves to obtain a definitive diagnosis. EUS plays an important role in evaluating these lesions: the ED cysts commonly appear as thin-walled cystic structures. MRI may also play a role when the differential diagnosis with other mediastinal "tumors" is difficult (4, 5).

Ladd and Gross in 1941 suggested three criteria for the diagnosis of ED: a lesion adherent or attached to some segment of the foregut, the presence of two layers of smooth muscle in the wall, and an internal lining of gastrointestinal mucosa; moreover, the differential point with bronchogenic cyst is that the wall of the lesion may be thicker (absence of cartilage) (2, 4).

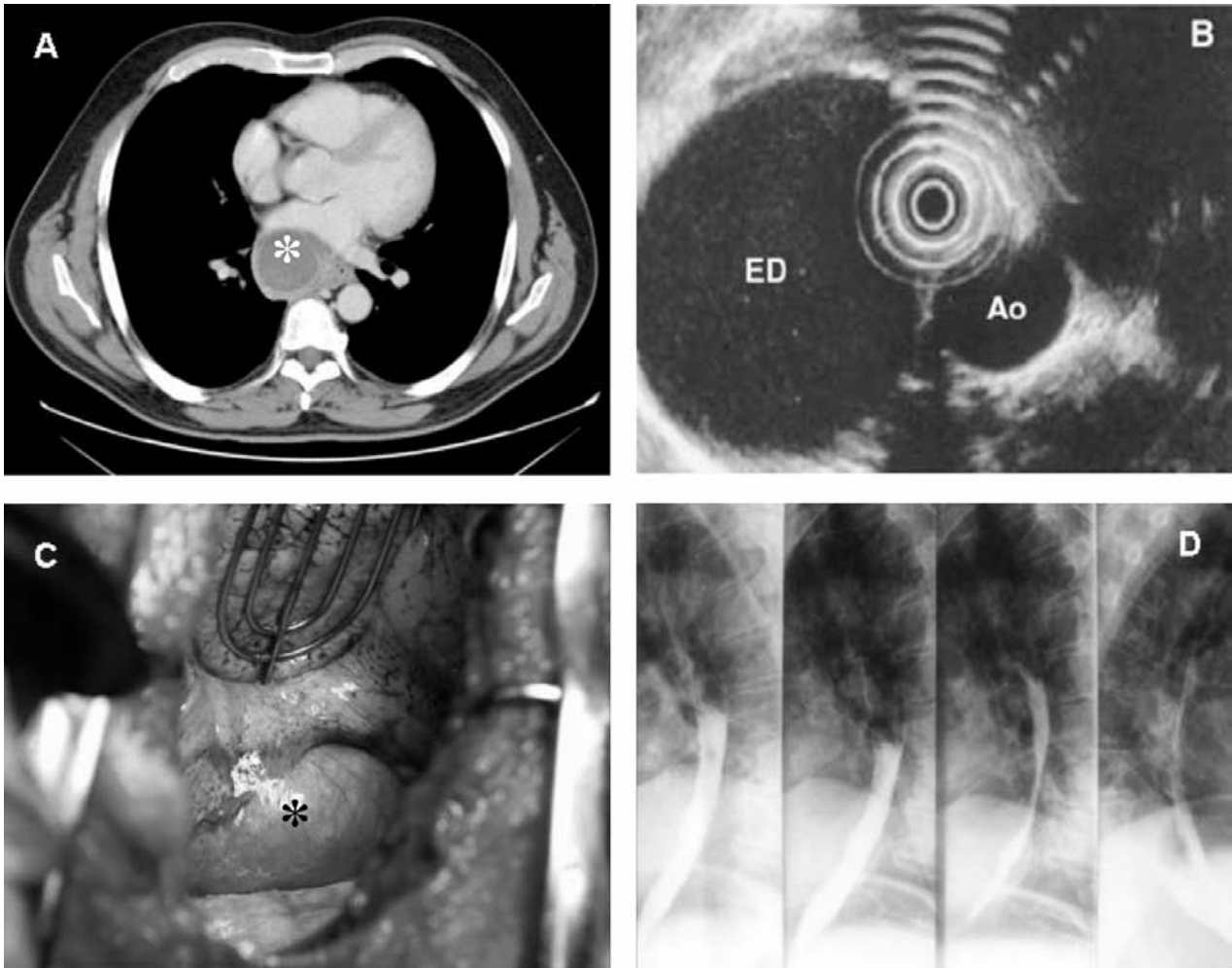


Fig. 1 - A) CT scan. B) EUS (Ao, aorta). C) Intraoperative view (cyst is *). D) Postoperative barium esophagogram.

Complete surgical excision by thoracotomy or thoracoscopy (Video-Assisted ThoracoScopy, VATS) is the therapy of choice even in asymptomatic patients because of the risk of complications such as aspiration and bleeding (4). There is a growing experience with the thoracoscopic treatment of mediastinal masses and primary oesophageal motility disorders (5). Some Authors reported a new mini-invasive approach with endoscopic resection in carefully selected patients, which essentially creates a lumen from the cyst into the esophageal lumen (1), and with robotic-assisted surgery (6).

Patients require close follow-up care. Pseudodiverticulum can develop if the muscle layer is not completely approximated and complications of vagal injury develop if these nerves are not preserved. Recurrence is rare, especially if the entire cyst was excised, and should be considered as a technical error. The morbidity rate is low. The overall complication rate is

very low and they often are inherent to the thoracotomy or VATS. Complications include pneumonia, persistent air leak, deep venous thrombosis, oesophageal leak or pseudodiverticulum, vagus nerve paralysis, and wound infection (3).

Conclusions

Dysphagia is the most frequent symptom of ED. The diagnosis and treatment of oesophageal cysts is still evolving. EUS is considered today the most sensitive method of diagnosis. The pathological diagnosis of oesophageal duplication requires the presence of the Ladd and Gross criteria.

The infrequent acute onset did not modify the management: resection of the ED is the procedure of choice. Treatment is currently shifting from thoracotomy to less-invasive procedures, including VATS and

endoscopic treatment. Even though the cysts are often asymptomatic at the time of diagnosis, the surgical excision is advised because definitive diagnosis is better done on the surgical specimen and most patients with untreated ED will experience dysphagia or develop complications such as bleeding or aspiration. Malignant degeneration of oesophageal duplication is very rare event.

Traditionally, the resection is accomplished via a thoracotomy; however, there are obvious advantages associated with the minimally invasive approach. The

surgical technique must emphasize evaluation of the integrity of the oesophageal mucosa, approximation of the muscle edges over the area where the cyst was present, in order to avoid a pseudodiverticulum, and identification of vagal nerves. Partial resection of the cyst can be done in difficult cases.

The future of the treatment of esophageal cysts lies in the advancement of minimally invasive operative techniques, which will lessen morbidity and mortality rates. Endoscopic treatment has been reported as a feasible and safe alternative in carefully selected patient.

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