Clinical Cases

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Cervico-mediastinal Esophageal Duplication Cyst - Case Presentation

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Rezumat

Chist de duplicație esofagiană cervico-mediastinal - prezentare de caz

Introducere: chisturile de duplicație esofagiană pot fi împărțite în două categorii: chisturi simple epiteliale si chisturi de duplicație. Acestea din urmă reprezintă o duplicație embriologică a musculare si mucoasei esofagiene, fără duplicație epitelială.
Prezentare de caz: prezentăm cazul unui bărbat în vârstă de 48 de ani, fără antecedente personale patologice semnificative, tranferat în serviciul nostru prezentând greață, vărsături, anorexie și pierdere în greutate. Evaluarea imagistica CT a evidențiat prezența unei formațiuni pseudotumorală, bine delimitată, cu dimensiuni maxime de 6.5 cm, situată la nivelul mediastinului superior și a regiunii cervicale inferioare. Abordul ales în scopul eliminării complete a acestei formațiuni a fost printr-o cervico-sternotomie în „Y", permițându-ne astfel accesul atât în mediastinul superior, cât și la nivelul regiunii cervicale anterioare.
Discuții: rezecția completă a tuturor malformațiilor tubului digestiv superior este recomandată. Având în vedere localizarea chistului de duplicație esofagiană în cazul prezentat, considerăm abordul prin cervico-sternotomie ca fiind cel mai adecvat, oferindu-ne o expunere bună a mediastinului superior și a regiunii cervicale antero-inferioară. Această expunere ne-a permis disecția și îndepărtarea completă a chistului de duplicație esofagiană.
Concluzii: Toate chisturile esofagiene trebuiesc evaluate și, eventual, rezecate. Cei mai mulți pacienți cu astfel de chisturi devin simptomatici la un moment-dat, motiv pentru care recomandăm rezecția acestor chisturi în momentul în care ele sunt identificate.

Cuvinte cheie: esofag, chist de duplicație, cervico-sternotomie

Abstract

Introduction: esophageal cysts can be divided into 2 categories: simple epithelial-lined cysts and esophageal duplication, which is an embryologic duplication of a portion of the muscle and submucosa of the esophagus without epithelial duplication.
Case presentation: 48 year-old male, with no relevant history, was transferred in our department with vomiting, dysphagia, anorexia, and weight loss. The CT scan highlights a tumor mass, well defined, with maximum size of 6.5 cm, developed in the left upper mediastinum and lower left cervical region. We decided to perform surgery in order to remove the mass completely. The approach was through a "Y" cervical incision, allowing access to the upper mediastinum, as well as for the anterior cervical region.
Discussion: Complete surgical excision is recommended for all foregut cyst malformations. Considering the localization of the esophageal cyst in our patient, we decided cervico-sternotomy will be the best approach, giving a wide exposure of the mediastinum and of the lower cervical region if necessary. The lesion was carefully dissected from the surrounding structures and completely removed.
Conclusion: All esophageal cysts should be evaluated and,

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eventually, resected. Most patients with esophageal cysts eventually become symptomatic; therefore, cysts should be resected when they are diagnosed.

Key words: esophagus, duplication cyst, cervical approach

Introduction

The primitive foregut gives rise to the pharynx, respiratory tract, and upper portion of the gastrointestinal tract from the esophagus to the duodenum at the ampulla of Vater. Foregut malformations originate from the region of the laryngeotracheal groove, which develops from the primitive foregut in the third to fourth week of gestation. The stage of embryonic development and separation during which the error occurs seems to play a role in the location and type of malformation, though the exact embryogenesis is yet to be fully elucidated.

In 1711, Blasius initially described esophageal cysts as duplications. In 1881, Roth also described these cysts, which can be divided into 2 categories: simple epithelial-lined cysts and esophageal duplication, which is an embryologic duplication of a portion of the muscle and submucosa of the esophagus without epithelial duplication. The term esophageal cysts applies to both categories.

Case report

48 year-old male, with no relevant history, was transferred in our department from internal medicine department with vomiting, dysphagia, anorexia, weight loss, and with no signs of upper gastrointestinal bleeding.

Abdominal ultrasound did not show any pathological alterations. Cervical ultrasound revealed a normal right thyroid lobe, but the left lobe was pushed towards the right side by a well defined parenchymal structure, relatively homogenous, 7-8 cm wide (Figs. 1, 2). The thyroid isthmus was within normal limits.

The CT scan highlights a solid mass, well defined, with maximum size of 6.5 cm, developed in the left upper mediastinum and lower left cervical region, with a starting point apparently from the left thyroid lobe. This tumor mass produced compression and displacement of the trachea and esophagus. The CT scan also revealed multiple adenopathies: left mediastinal, left tracheal, sub-carinal and left para-esophageal (Figs. 3 – 6).

Thyroid scintigraphy: normal positioned thyroid gland, its dimensions within normal limits; low capture of the radioisotope with a relatively homogenous distribution in both thyroid lobes. Lower thyroid pole is medial displaced due to a lacunar area situated lateral and caudal (Fig. 7).

Upper GI tract echo-endoscopy: at the aortic arch level a solid, heterogeneous mass with a width of 52 mm and a length of 12 cm. This mass is well defined and it does not invade the adjacent structures. Other findings: "A" grade esophagitis and eritemateous gastropathy with no compression of the esophagus (Figs. 8, 9).

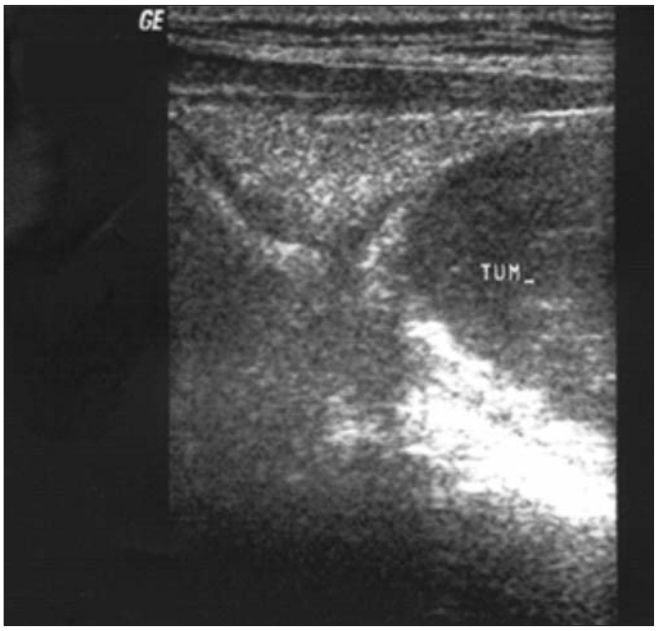

Figure 1. *Ultrasound image - left thyroid lobe*

Figure 2 *Ultrasound image - tumor located near the left thyroid lobe*

Fiber bronchoscopy: left tracheal wall recurrence on the upper third, normal carina, extrinsic compression right main bronchus and intermediate bronchus.

Cytological exam (fine needle aspiration): yellow-grey viscous, gelatinous fluid with rare epithelial grouped cells, with typical aspect, small nuclei, heterogeneous base material with cholesterol crystals.

We decided to perform surgery in order to remove the cyst completely. The approach was through a "Y" cervical

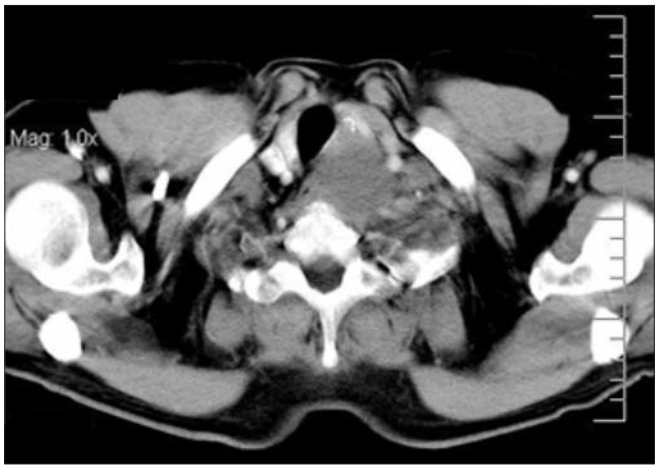

Figure 3. *CT scan - left cervical mass*

Figure 4. *CT scan - intrathoracic mass*

Figure 5. *CT scan – enlarged mediastinal lymph node*

Figure 6. *CT scan – enlarged sub-carinal lymph node*

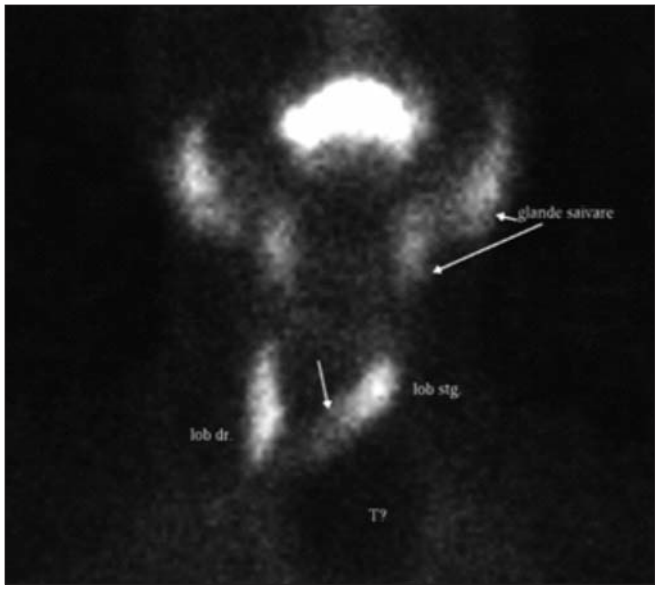

Figure 7. *Scintigraphy – left thyroid lobe displaced by a tumor mass*

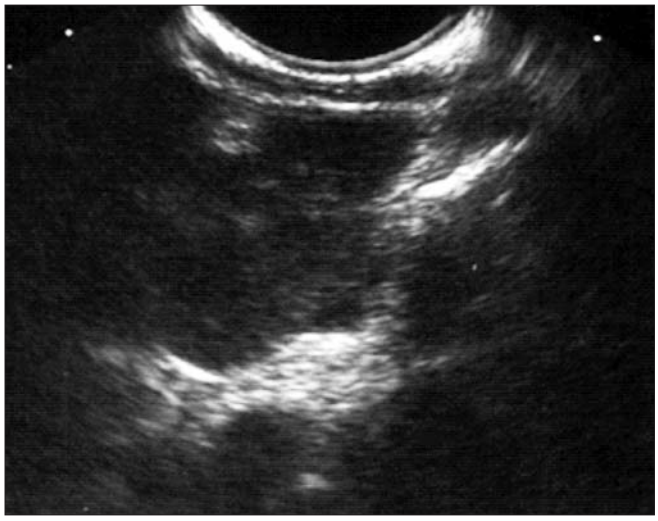

Figure 8. *Echo-endoscopy - heterogeneous mass at aortic arch level*

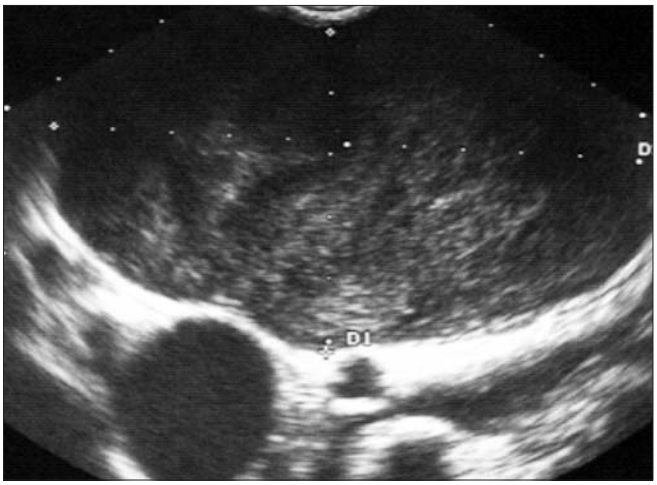

Figure 9. *Echo-endoscopy – well defined tumor mass located by the aorta*

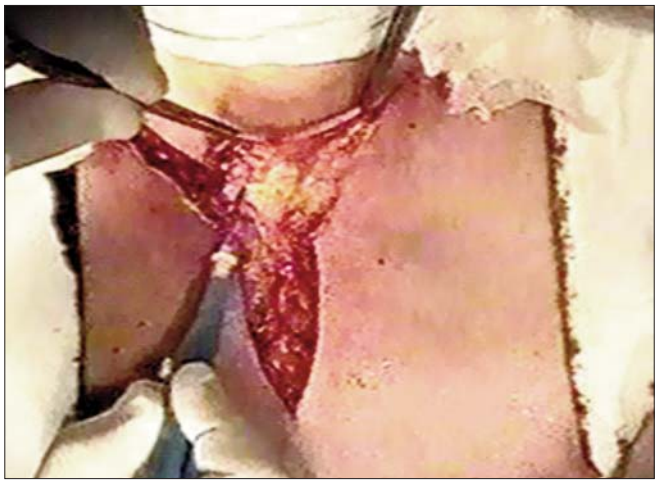

Figure 10. *"Y"-shaped cervical incision*

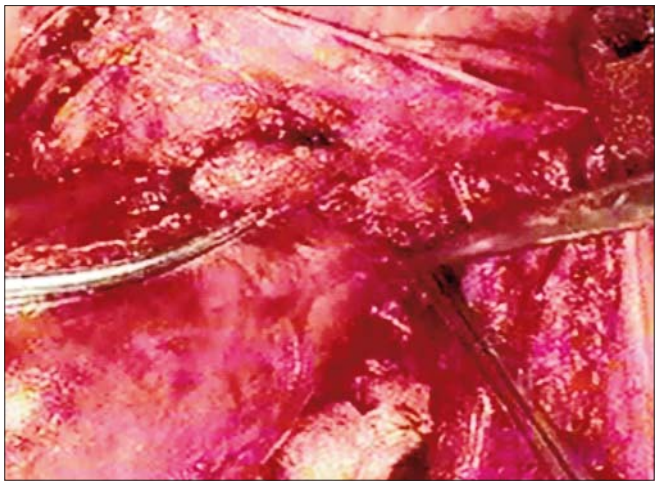

Figure 11. *Dissection of the upper pole of the tumor*

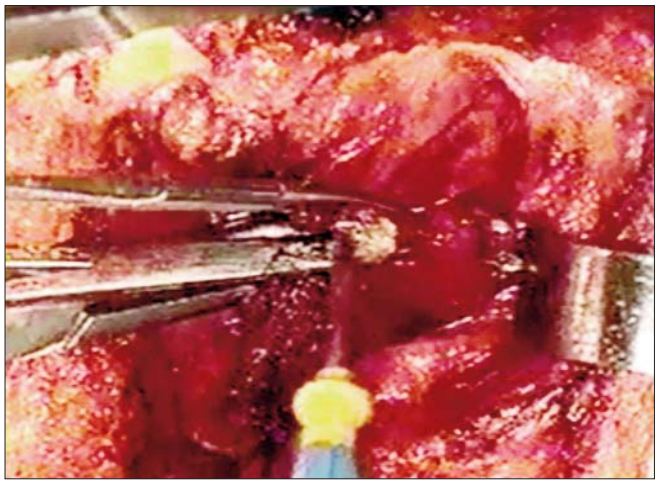

Figure 12. *Sectioning of the sternocleidomastoid and omohyoid muscles*

incision (Fig. 10), followed by the subcutaneous plan dissection and the upper pole of the tumor (Fig. 11).

Sectioning the sternocleidomastoid and omohyoid muscles was necessary because otherwise we could not have achieved an appropriate approach over the esophageal cyst (Fig. 12). We continued the dissection until the cyst was freed from trachea, thyroid gland, esophagus and the left vascular pedicle of the neck (Fig. 13).

When the cyst was completely freed from the surrounding tissue, we punctured it and partially evacuated its contents (Figs, 14, 15) in order to dislocate it from the mediastinum. Now the vascular pedicle has been individualized and it was ligated and sectioned (Fig. 19). The esophageal cyst was at this moment completely removed.

Histological exam (after surgical removal of the cyst): cystic walls with ciliated cylindrical epithelium of respiratory type and stratified epidermoid tissue, smooth and striated muscle tissue, connective tissue, fat tissue containing

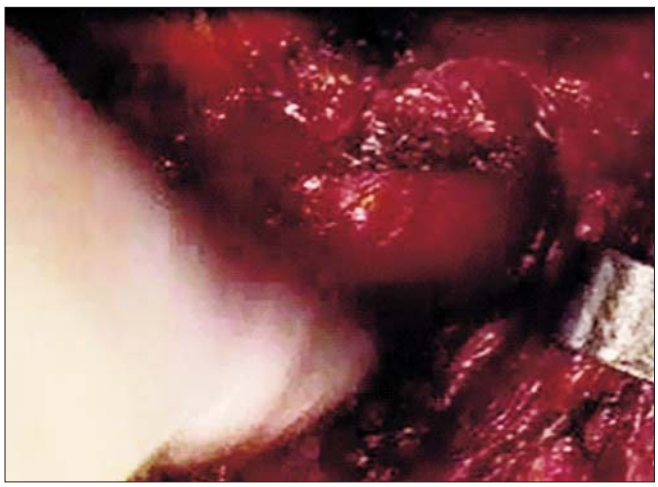

Figure 13. *The cyst is freed from trachea, thyroid gland, esophagus and the left vascular pedicle of the neck*

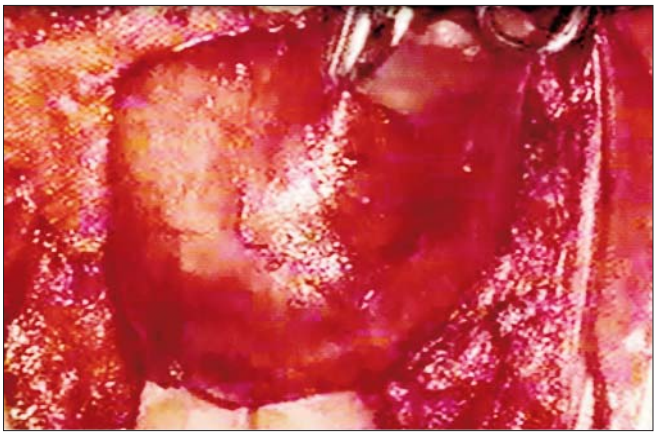

Figure 14. *The mass is completely identified*

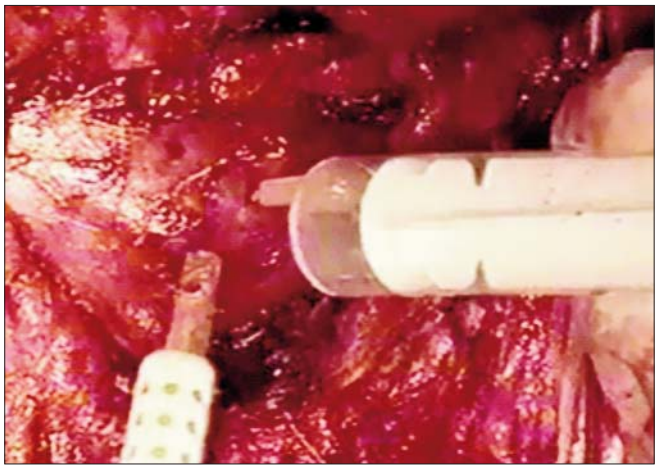

Figure 15. *Puncturing the mass in order to establish if it's cyst*

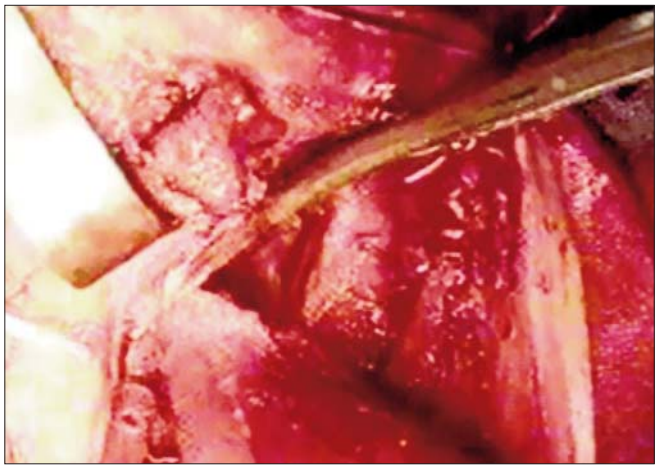

Figure 16. *The vascular pedicle of the cyst is being ligated and sectioned*

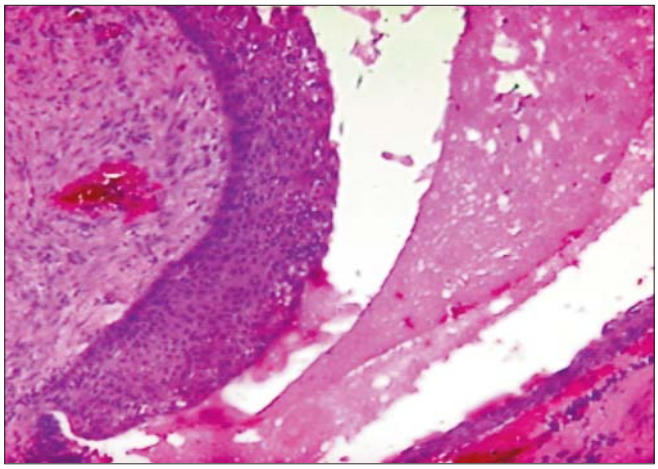

Figure 17. *Histological examination – esophageal cyst (smooth muscle, amorphous contents)*

bronchial glands. Conclusion: esophageal cyst (Figs. 17, 18, 19 and 20).

Postoperative evolution was favorable, without any major complications. The patient was discharged the seventh day after surgery.

Discussion

Esophageal cysts and duplications are defined as the presence of a double structure, partial or complete, with variable length, localized at cervical, mediastinal or abdominal level. Esophageal cysts, duplications or diverticula can express the same embryological defect.

A classification of mediastinal cysts proposed by several authors (1) divides them in bronchogenic and esophageal cysts. Some authors state that bronchogenic cysts may be differentiated by the presence of cartilage or bronchial glands in the cyst wall. Salyer and his group (2) and Nobuhara et al. (3) proposed that esophageal duplications may be differentiated by

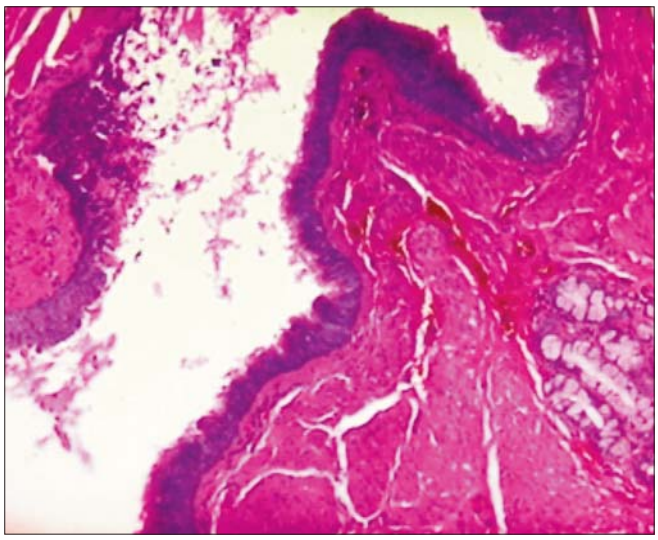

Figure 18. *Histological examination – esophageal cyst (smooth muscle, skeletal muscle, cystic contents)*

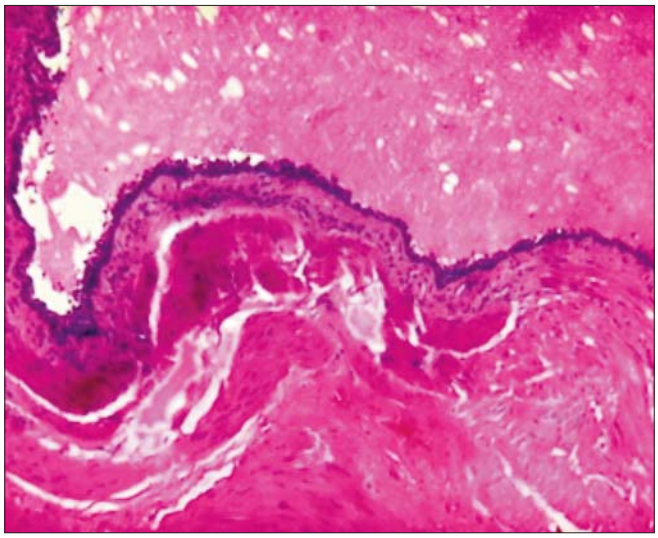

Figure 19. *Histological examination – esophageal cyst (squamous epithelium, stratified squamous epithelium, cystic contents)*

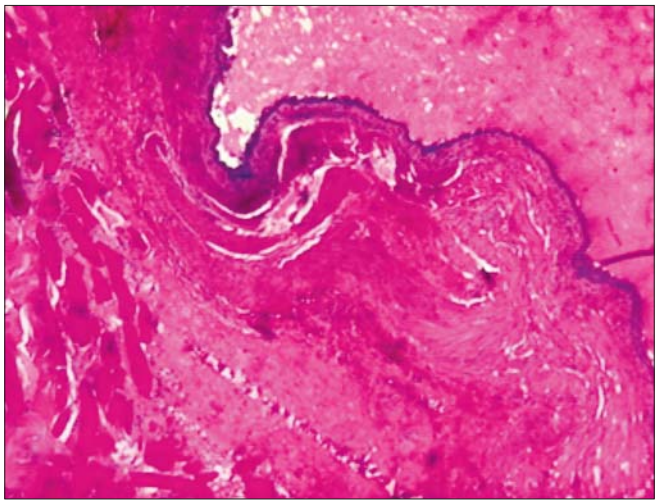

Figure 20. *Histological examination – esophageal cyst (squamous epithelium, respiratory epithelium, bronchial glands, smooth muscle)*

the presence of two distinct muscle layers or longitudinally oriented muscle fibers on histologic examination. Because all these cysts arise from a ventral anomaly of the embryonic foregut, differentiation into distinct entities may not be of any clinical significance.

Clinical presentation usually associates symptoms of chest pain or respiratory complaints of dyspnea, cough, pneumonia, recurrent asthma, hemoptysis, and cyanosis with gastrointestinal complaints of vomiting, dysphagia, anorexia, weight loss, and/or gastrointestinal bleeding. The presentation may be asymptomatic, or it may present incidentally on radiographic examination. The manner of presentation seems to depend on several factors, including the anatomic location, age at presentation, mass effect of the lesion, and potential complications that may result from the lesion (1). Some of these complications are infections, dysphagia, hemorrhage from ectopic mucosa, perforation, and peptic esophageal stricture. There is cited the potential for fistulous erosion of the cyst to the adjacent airway tract. Rare and lethal complications may include superior vena cava syndrome, arrhythmias, pulmonary artery stenosis, malignant degeneration, fatal air embolism, and myocardial infarction from left coronary artery compression (1).

Complete surgical excision is recommended for all foregut cyst malformations. Bratu and colleagues (4) suggest that a staged approach may be necessary to initially drain an infected cyst, treat the infection, and then implement a definitive resection. Prompt surgical intervention is mandatory in the infant with severe respiratory distress. The cyst is generally approached through a posterolateral thoracotomy or via thoracoscopy. Considering the localization of the esophageal cyst in our patient (upper left mediastinum), we decided median cervico-sternotomy will be the best approach, giving a wide exposure of the mediastinum and of the lower cervical region if necessary. The lesion should be carefully dissected from the surrounding structures, preserving them when possible (1). Concerns for recurrence and potential malignant degeneration, as expressed by DiLorenzo et al., (5) argue against partial surgical excision. Intraoperatively, St-Georges et al. (6) report concerns for tracheal, bronchial, esophageal, or vagus nerve injury.

Postoperative complications reported by Suen et al., (7) St-Georges and associates, (6) and Zhang et al. (8) include recurrence, hemothorax, atelectasis and postoperative pneumonia, persistent air leak, Clostridium difficile enterocolitis, pleural effusion, wound infections, gastroesophageal reflux, and Horner's syndrome. In this case, we didn't notice any of these complications.

The prognosis overall is excellent after complete resection of the lesions.

Conclusions

The diagnosis and treatment of esophageal cysts is still evolving. Diagnosis is aided by the relatively recent developments of CT scans and endoscopic ultrasonography. Current treatment of choice is still surgery through open thoracotomy, but the trend to less invasive procedures, including video-assisted thoracic surgery (VATS) and endoscopic treatment is gaining more and more support (9,10).

All esophageal cysts should be evaluated and, eventually, resected. Radiographs guided by history and physical examination findings usually confirm the diagnosis. Other useful investigations are CT scan, which reveals a fluid-filled cystic structure originating from the esophagus, endoscopy demonstrates extrinsic compression with intact mucosa and endoscopic ultrasonography reveals a cystic, filled structure in connection with the esophagus.

Most patients with esophageal cysts eventually become symptomatic; therefore, cysts should be resected when they are diagnosed.

Surgical excision is the treatment of choice for congenital esophageal cysts. The short-term results after surgical removal of congenital esophageal cysts was excellent. However, the long-term follow-up revealed esophagus-related symptoms and pathological findings such as moderate heartburn or reflux symptoms.

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