

## Malignant Extramucosal Esophageal Tumor - Yolk Sac Tumor - Case Report

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### Rezumat

*Tumoră malignă extramucoasă esofagiană – tumoră de sinus endodermal – prezentare de caz.*

Tumorile esofagiene extramucoase maligne sunt rare. Publicăm un caz de tumoră esofagiană extramucoasă care s-a dovedit a fi o tumoră de sinus endodermal. Acest tip de tumoră este extrem de rar. Prezentăm managementul inițial și, de asemenea, managementul la distanță a acestui caz (de la diagnostic până la gestionarea complicațiilor tardive). Pacientul a fost internat pentru disfagie. Tomografia computerizată toraco-abdominală cu substanță de contrast relevă o importantă masă retrocardiacă; eco-endoscopia cu biopsie a confirmat o tumoră esofagiană extramucoasă malignă. Am efectuat o esofagectomie prin triplu abord (McKeown) - toracotomie dreaptă, laparotomie și cervicotomie stângă pentru anastomoză. Examenul histopatologic clasic a ridicat suspiciunea de tumoră mediastinală cu celule celule germinale sau carcinom slab diferențiat. Examenul imuno-histochimic a precizat diagnosticul pozitiv de tumora de sinus endodermal. Bolnavul a urmat tratament citostatic cu etoposid și cisplatin. Postoperator tardiv, pacientul dezvoltă insuficiență respiratorie secundară unei hernii hiatale gigante care a fost operată prin abord laparoscopic.

Cuvinte cheie: tumoră esofagiană extramucoasă, tumoră de sinus endodermal, esofagectomie McKeown, hernie hiatală, laparoscopie

### Abstract

Malignant extramucosal esophageal tumors are rare. We publish

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a case of an extramucosal esophageal tumor which turned out to be a yolk sac tumor. This type of tumor is extremely rare. We present the initial and also late management of this case (from the diagnostic to the management of the late complications). The patient was admitted for dysphagia. Contrast thoraco-abdominal CT scan revealed an important retrocardiac mass; echo-endoscopy with biopsy confirmed one malignant extra-mucosal esophageal tumor. We performed an esophagectomy through triple approaches (McKeown) – right thoracotomy, laparotomy, and left cervicotomy for anastomosis. The pathological report raised the suspicion of a mediastinal tumor with germ cells or a poorly differentiated carcinoma. The immunohistochemistry tests confirmed a yolk sac tumor. The patient was referred to the oncologist and was treated with etoposide and cisplatin chemotherapy. Subsequently, the patient developed respiratory failure secondary to giant hiatal hernia which was surgically treated via laparoscopy.

**Key words:** extramucosal esophageal tumor, yolk sac tumor, McKeown esophagectomy, hiatal hernia, laparoscopy

## Introduction

Extramucosal esophageal tumors are a rare entity in esophageal tumor pathology; the diagnosis is either incidental or as a result of the onset of symptoms; the diagnosis is mainly based on endoscopy and tomography, but both barium X-ray and echo-endoscopy find their place in the diagnosis. Surgical treatment is indicated only in the presence of symptoms or if the malignancy is certain. Surgical treatment in extramucosal tumors mainly refers to enucleation. Esophageal resection for benign lesions is an extremely rare event, but for malignant lesions it is the procedure of choice; esophageal resection can be performed through a variety of approaches: right/ left thoracotomy, laparotomy, complete minimally invasive approach (laparoscopy + thoracoscopy / VATS) or mixed (laparotomy + thoracoscopy/ VATS, laparoscopy + thoracotomy). Also, the minimally invasive robotic approach, both abdominal and thoracic, can be used (1). In the literature we found few articles concerning extramucosal esophageal tumors and fewer concerning malignant ones (2-14).

## Case Report

We present the clinical case of 72-year-old

patient with hypertension, atrial fibrillation, ischemic cardiomyopathy, heart failure NYHA II, admitted for dysphagia and involuntary weight loss, approximately 15 kilograms in 3 months. Four years before this hospitalization, the patient was investigated for pain in the right hypochondrium. Computed tomography suspected a small paraesophageal tumor that remained incompletely investigated. The patient received surgery in another health care unit for chronic alithiasic cholecystitis.

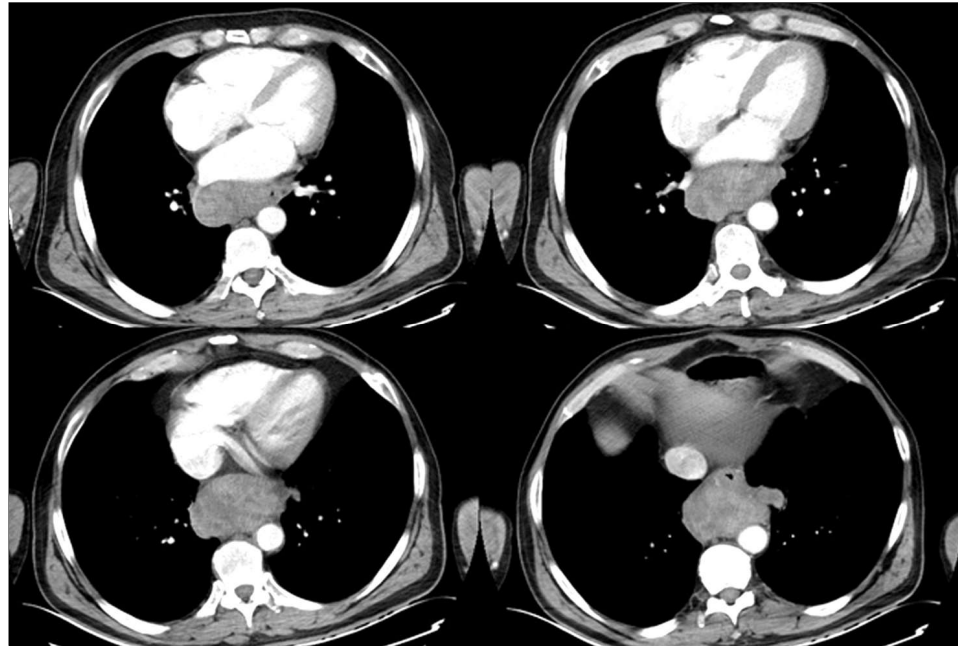
At the time of admission to our department, the hematology and biochemistry tests were almost normal.

Contrast CT scan of the thorax and abdomen (May 2020) revealed a retrocardiac tumor mass, which enclosed and displaced the esophagus laterally (on the left side), with important dimensional progression compared to previous examination (2017); in observation esophageal leiomyoma, without excluding a gastrointestinal stromal tumor; mediastinal (subcarinal) and subdiaphragmatic (celiac) lymphadenopathy were also found (*Fig. 1*).

Upper digestive endoscopy performed on the occasion of hospitalization for the symptoms presented above showed an extrinsic compression of the esophageal mucosa, without its invasion; correlating with the imaging aspects, EUS-FNA was practiced.

The pathology result from endoscopic

**Figure 1.** Preoperative CT of the thorax and abdomen with contrast agent – retrocardiac tumor mass that encloses and pushes the lateral esophagus (to the left), having important dimensional progression; mediastinal lymphadenopathy (subcarinal)

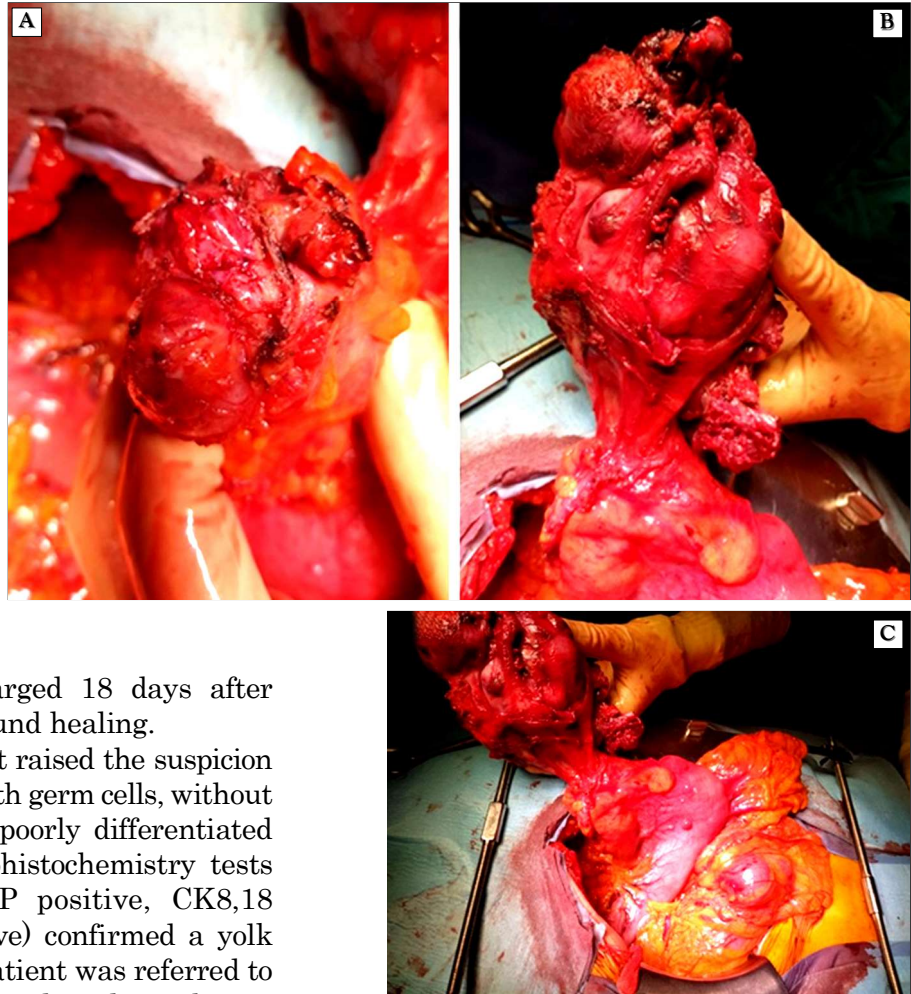


biopsy – malignant round cell tumor (epithelioid GIST, seminoma or round cell tumor).

The case was discussed in the tumor board session and the recommendation was for surgical therapy as initial management (considering the symptomatology and malignancy; diagnosis of germ cell tumor, which would have required neoadjuvant chemotherapy, was unexpected). We started the McKeown procedure via right posterolateral muscle sparing thoracotomy (5<sup>th</sup> intercostal space). No disseminated lesions in the pleural cavity observed. At the level of the posterior mediastinum, from the carina through esophageal hiatus to esogastric junction, a massive tumor invasive in lung was observed, adherent to the pericardium, aorta and right main bronchus and inferior pulmonary vein. We performed a wedge resection at the level of the pulmonary right lower lobe. Using blunt and sharp dissection we completed the esophageal release from the inferior pulmonary vein, pericardium, right main bronchus, carina and aorta, with minimal breach at the level of the left mediastinal pleura (left chest tube drainage inserted at the end of the surgical procedure). Before right pleural cavity drainage and thoracic wound

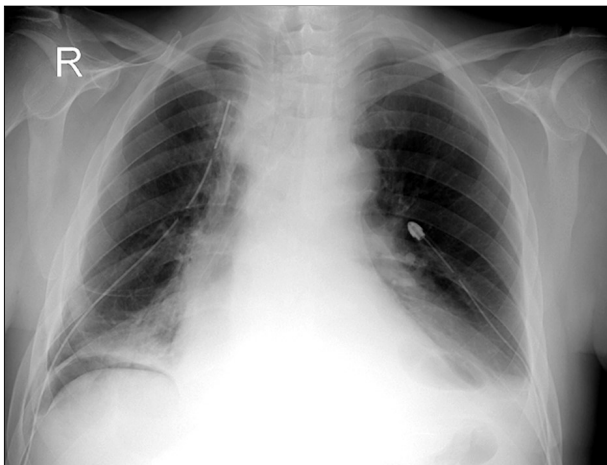
closure, the esophagus was stapled and cut in the apex of the pleural cavity. After that, the patient was repositioned on the operating table in a supine position. A laparotomy was performed, no metastatic lesions were observed in the peritoneal cavity or at the level of abdominal organs. A celiac lymphadenectomy (large lymphadenopathy) was performed; completed the adhesiolysis between the anterior part of the esophageal hiatus and tumor; the right and the left diaphragmatic crus were resected because of the tumor invasion and the specimen was retrieved from the thorax and finalizing the subtotal esophagectomy and partial superior gastric resection (*Fig. 2*); an Akiyama gastric tube was prepared and pulled up through the posterior mediastinum at the cervical level after left cervicotomy; we performed a modified Collard cervical esophagogastric anastomosis (hybrid technique – posterior wall by stapler and anterior wall hand-sewn). After the cervical wound closure, a jejunostomy was performed. Abdominal drainage and wound closure finalized the surgical procedure (*Fig. 3*). On postoperative day 7 we observed small anastomotic leakage (less than 100 ml / 24 h) that was treated conservatively.

**Figure 2.** Intraoperative appearance of the tumor mass  
**(A)** esophageal stump; **(B)** giant tumor mass pulled out from mediastinum; **(C)** laparotomy view: stomach and esophagus



The patient was discharged 18 days after surgery with cervical wound healing.

The pathological report raised the suspicion of a mediastinal tumor with germ cells, without being able to exclude a poorly differentiated carcinoma. The immunohistochemistry tests (AE1/AE3 positive, AFP positive, CK8,18 positive, Glypican positive) confirmed a yolk sac tumor (*Fig. 4*). The patient was referred to the oncologist. Etoposide and cisplatin chemotherapy was applied. The follow-up was in normal parameters for 9 months when the patient presented with intensive cough and respiratory failure with significant

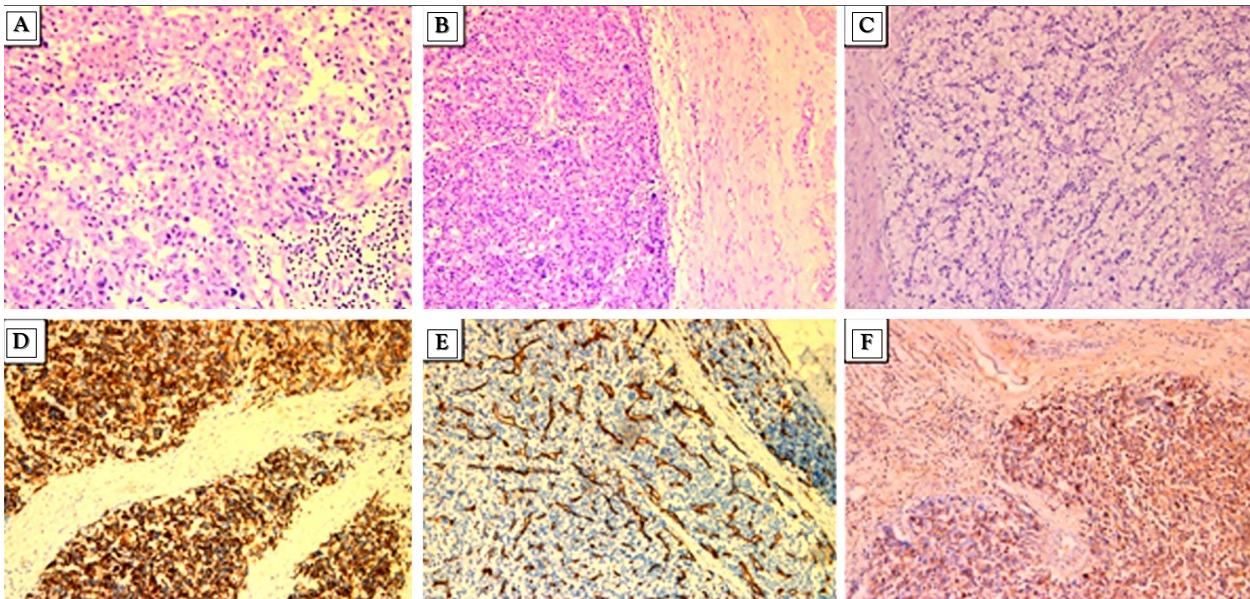


**Figure 3.** Day 3 post-esophagectomy - chest X-ray

impairment of quality of life. Thoraco-abdominal CT scan revealed important hiatal hernia (containing the transverse colon) with left lower lobe collapsed. PET-CT without oncological lesions (*Fig. 5*). We performed a diaphragmatic hernia repair via laparoscopy (*Fig. 6*). Surgery consisted of reducing the hernia contents into the abdominal cavity, followed by suturing of the hiatal defect with non-absorbable interrupted suture. The patient was discharged 6 days after surgery (*Fig. 7*).

## Discussion

Extramucosal esophageal tumors (EmET) develop in the esophageal wall, beyond its mucosa (with origin in submucosa or muscularis propria). Intramural tumors are usually

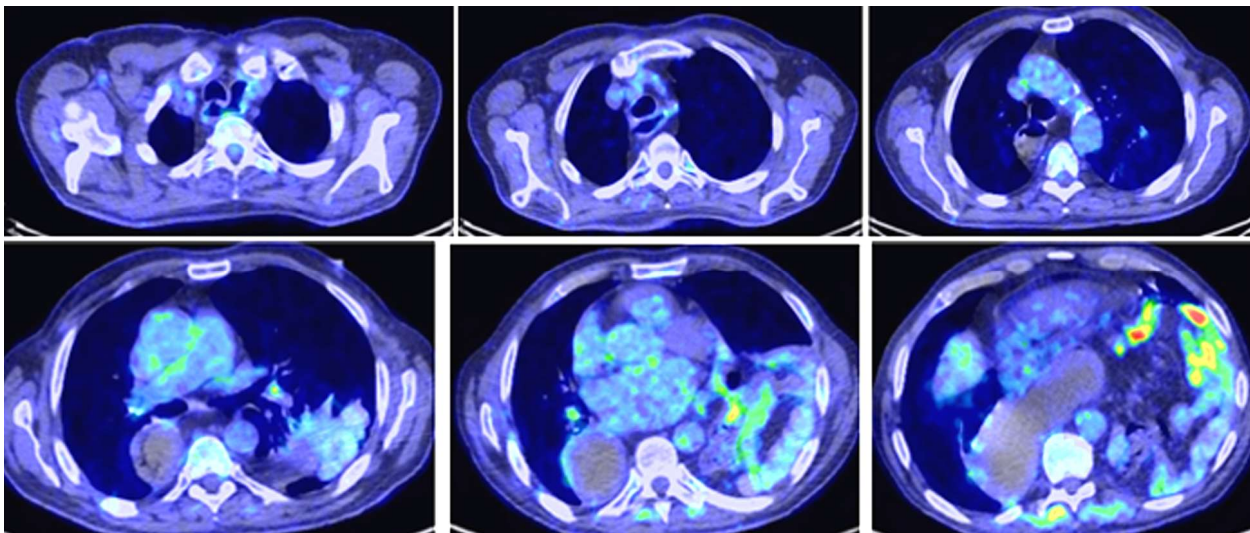


**Figure 4.** Microscopic and immunohistochemical findings of the yolk sac tumor; **(A)** HE staining, x20, trabecular architecture weakly eosinophilic cytoplasm, solid and trabecular architecture; **(B)** HE staining, x10, tumor adjacent to the esophageal wall represented by smooth muscles, on the right side; **(C)** HE staining, x10, clear cell areas, solid and microcystic architecture, abundant vascularization; **(D)** cytokeratin AE1-AE3, diffuse positive; **(E)** CD34, negative in tumor cells, positive in vessels, emphasizing the trabecular architecture of proliferation; **(F)** AFP, zonal expression in tumor cells

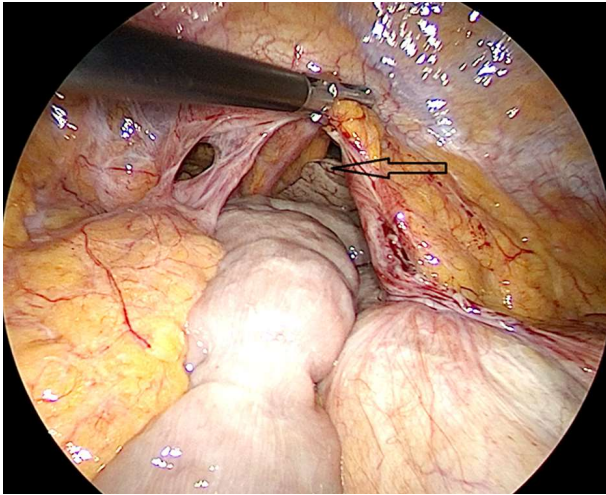
well defined and they do not produce complete stenosis of the esophagus. Extramucosal esophageal tumors are usually asymptomatic and very rare (probably because they are underdiagnosed). Because of their infrequency, they can present a diagnostic dilemma in patients with dysphagia, odynophagia, or

radiologic abnormalities on imaging studies (2,15-17).

Esophagoscopy is limited to partial mucosal visualization and evaluation of the external compression. Classical biopsy allows histopathological assessment of the mucosa because it is limited to the epithelium and lamina



**Figure 5.** PET scan surveillance - no oncological lesions; the herniated transverse colon is observed in the left hemithorax



**Figure 6.** Hiatal hernia repair - lung (black arrow)

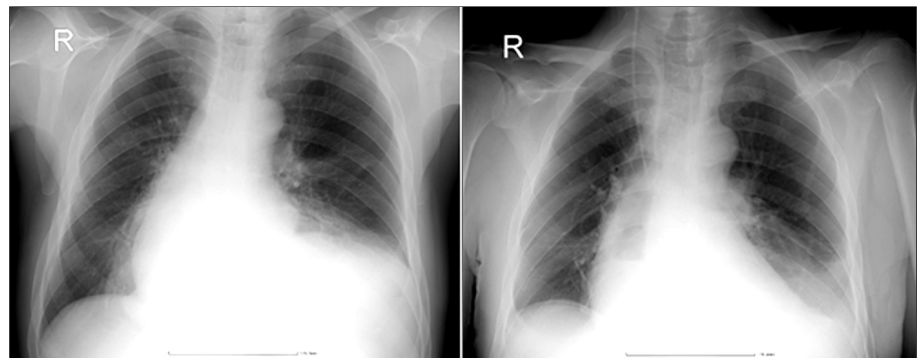
propria. Endoscopic ultrasound (EUS) is essential in the diagnosis of extramucosal esophageal tumors. However, if the overlying mucosa is normal, biopsy should be avoided for it is likely to be nondiagnostic and may complicate enucleation (prior biopsy with secondary impaired mucosal integrity as well as tissue remodeling produced, would predispose to a fistula in case of enucleation of an extramucosal tumor). Because the majority of extramucosal esophageal tumors are asymptomatic, slow growing, discovered incidentally, and have a low malignant potential, most require nothing more than diagnosis and, occasionally, surveillance. However, large and/or anatomical narrowing located tumors are more likely to become symptomatic and require removal. If symptomatic, patients with benign esophageal tumors typically present with dysphagia; however, odynophagia, reflux,

regurgitation, chest pain, hiccups, anorexia, or respiratory complaints including cough, dyspnea, or pneumonia may be present (2,15-18).

A heterogeneous aspect of an EmET on echo-endoscopy or a tumor greater than 3 to 4 cm in largest diameter is suspicious for malignancy. Malignant extramucosal esophageal tumors (MEMET) are very rare conditions. Any suspicion of malignancy requires surgical management (2,15-18).

Yolk sac tumors (also known as primitive endodermal tumors or endodermal sinus tumors) are malignant primitive germ cell tumors. They are histologically similar to the mesenchyme of the primitive yolk sac. Yolk sac tumor is a type of germ cell tumor. Germ cell tumors commonly arise in the gonads, but can sometimes occur outside the gonads and are called extragonadal germ cell tumors. Anatomical distribution of extragonadal germ cell tumors: abdomen and pelvis (uterus, pancreas), anterior mediastinum, esophagus, brain (including pineal gland). To avoid the high morbidity and mortality associated with this condition, it must be promptly diagnosed and treated. This activity reviews the evaluation and treatment of the yolk sac tumors and highlights the role of the interprofessional team in evaluating and treating patients with this condition (2,15-18).

If enucleation is the surgical treatment of choice for symptomatic extramucosal esophageal tumors, when we are talking about malignant or suspiciously malignant extramucosal tumors, surgical treatment must impose oncological principles. Thus, various types of approaches for esophagectomy and esophago-



**Figure 7.** Chest Xray aspects: before and after hiatal hernia repair

plasty can be used: thoracotomy (left or right side), thoracoscopy or VATS, laparotomy, laparoscopy, left side cervicotomy for anastomosis or mixed (combined) approaches (2-14, 18).

The most frequently used surgical procedures are Ivor Lewis, McKeown and transhiatal esophagectomy (Orringer). Analyzing the data from the specialized literature regarding the three types of approach, McKeown esophagectomy is associated with more unplanned intubation, increased difficulty weaning from the ventilator, incisional surgical site infections, anastomotic leak, and higher length of stay, but is associated with improved overall survival and a decreased risk of disease recurrence in patients with stage T3 tumors. Rates of pneumonia, ventilator dependence, and septic shock are increased with the length of thoracic incision (thoracotomy versus VATS); transthoracic approach is an independent risk factor for the development of new-onset atrial fibrillation (AF) after esophagectomy; new-onset AF is associated with severe postoperative complications and longer hospital stay; minimally invasive approach does not decrease the incidence of new-onset AF. However, transhiatal esophagectomy is not without complications. The transhiatal esophagectomy procedure requires a skilled surgeon and a trained interprofessional team that works together to provide quality patient outcomes (2-14,18,19).

Given the few similar cases presented in the literature, we emphasize that extragonadal yolk sac tumors are rare, those with digestive localization being extremely rare. Most of the cases presented in the literature have a mixed histopathological component (germinal cells and poorly differentiated adenocarcinoma). The oncological evolution of patients with extragonadal germ cells tumor is worse than those with gonadal germ cells tumor. The prognosis of patients is also given by the time of diagnosis, as most patients with yolk sac tumor with digestive localization are in metastatic stage. It should be mentioned here that the symptoms are initially nonspecific, being easily confused with other non-malignant pathologies. Even if

the treatment of gastric yolk sac tumor is represented by aggressive chemotherapy, we consider that a combination with radiation therapy and surgery can improve prognosis (14-16).

Cervical anastomotic leakages may manifest either cervically or intrathoracically. Intrathoracic manifestations are more commonly accompanied by fever and leukocytosis than cervical manifestations. Compared to patients with cervical manifestations, those with intrathoracic manifestations had a longer duration of hospital stay and higher incidence of tracheal fistula (which can be caused by damaging of the membranous portion during dissection, secondary to intubation or a cervicomedial abscess; treatment of the tracheal fistula requires a tracheal stent installation, up to surgical management). Management of leakage refers to conservative management (wait-and-see strategy), cervical wound drainage, mediastinal drainage (transthoracic or transnasal inner drainage), esophageal stent placement or surgical reintervention, if necessary (2-14,18-21).

Hiatal hernia is an uncommon postoperative complication after esophagectomy, with a reported incidence of 0.4% to 6%, mainly with herniation of intraabdominal organs into the left hemithorax. Hiatal hernia can occur in the early postoperative period or as a late complication. When it occurs early, its cause can most likely be attributed to the lack of peritoneal adhesions. With longer follow-up, other mechanisms such as progressive hiatal dilatation by the tumor, resection of the hiatal structures, increased intraabdominal pressure, and the pull of negative intrathoracic pressure may contribute to its development. Another reason might be the more frequent use of minimally invasive procedures in esophageal surgery, leading to fewer peritoneal adhesions, especially in the hiatal region. Diaphragmatic hernia is a possible complication to be sought in all patients who show signs and symptoms of respiratory or intestinal obstruction. The surgical treatment aims at the management of the hernia and the possible complications caused by it (ischemia, injury of the affected

organs, such as the spleen); this can be achieved by a classic or minimally invasive approach, depending on the experience of the surgical team in correlation with the severity of the case. Surgical treatment refers to the reduction of the hernial content in the abdominal cavity, followed by the repair of the diaphragmatic defect by non-absorbable interrupted suturing or in case of major defects, an addition of an absorbable mesh (2-14,18-21).

## Conclusions

Yolk sac tumor, which most frequently arises in the gonads as a type of germ cell tumor, is rare in children, but is highly malignant, whereas in adults it is even rarer.

Yolk sac tumors of the esophagus are extremely rare, most of the cases presented in the literature are pediatric ones.

This paper is the first of its kind presented in Romania, the uniqueness being one of the peculiarities of this case.

Although rare, malignant extramucosal esophageal tumors should be considered when certain characteristics, such as tumor size, symptomatology, and homogeneity are suspected. When malignancy is suspected, surgery should be oncologically radical.

Complications of postesophagectomy, immediate and late, require sustained management, consistent interdisciplinary collaboration, and consistent experience of the anesthetic-surgical team.

## Conflicts of Interest

The authors declare no conflicts of interests.

## Ethics Approval

All procedures performed were in accordance with the ethical standards of the 1964 Helsinki Declaration and its later amendments.

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