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Case Report
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Robot-Assisted Esophageal Leiomyosarcoma Resection

Arámbulo RH[1], Ya-Lyn AN[2], Hillegersberg RV[2] and Ruurda J[2]*

[1]Department of General and Digestive Surgery, Bellvitge University Hospital, Spain

[2]Department of Surgical Oncology - Upper Gastrointestinal Surgery, UMC Utrecht, Netherlands

Abstract

Background: Esophageal leiomyosarcomas are rare, they represent approximately 0.1% to 0.5% of malignant esophageal tumors and were described for the first time in 1902, since then only a few cases have been reported. Robot-assisted thoracoscopic surgery is a relatively new technique for treatment of esophageal lesions. We report a case of robot-assisted thoracoscopic resection of an esophageal leiomyosarcoma.

Case Report: A 64-year-old man with a past medical history of smoking initially presented to our unit with a 10-months history of retrosternal chest pain. The pain had to be controlled with opiods. However, symptoms persisted and a surveillance Computerized Tomography performed 6 months later demonstrated a new 2.1 cm left paraesophageal mass without thoracic lymphadenopathy and close contact with the thoracic aorta. Further investigations performed included a Positron Emission Tomography-Computerized Tomography, and an Endoscopic Ultrasonography (EUS). EUS biopsies revealed the mass to be a leiomyosarcoma.

The patient subsequently underwent robot-assisted left sided thoracoscopic resection. The tumor was located in the mid-thorax arising from the esophagus with a pedicle to the esophageal wall and it was resected en bloc along with the surrounding pleura. There was no invasion into the aorta or pulmonary vein. The patient made a full recovery and was sent home 2 days after the surgery. Pathology report described a Grade 2 Sarcoma with a diameter of 4.0 cm, with free resection margins.

Conclusion: The robot-assisted minimally invasive thoracoscopic approach should be considered a feasible option for resection of esophageal leiomyosarcoma due to its benefits on dissection in a limited space, improved precision and complications reduction.

Keywords: Leiomyosarcoma; Esophageal tumor; Robotic surgery; Thoracoscopic; Robot-assisted surgery; Minimally invasive

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Correspondence:
Rodrigo Hermoza Arámbulo, Department of General and Digestive Surgery, Bellvitge University Hospital, Carrer de la Feixa Llarga, s/n, 08907 L'Hospitalet de Llobregat, Barcelona, Spain, Tel: +34 674 84 01 67; E-mail: rhermoza@bellvitgehospital.cat

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Introduction

Leiomyosarcoma is a high-grade slow-growing smooth muscle connective tissue tumor [1]. Its location in the esophagus was described for the first time in 1902 by Howard, since then only a few cases have been reported [2]. Esophageal leiomyosarcomas are rare, they represent approximately 0.1% to 0.5% of malignant esophageal tumors [3] and are characterized by sheets of spindle-shaped cells associated with increased number of mitotic figures [4].

Robot-assisted thoracoscopic surgery is a relatively new technique for treatment of esophageal lesions. It improves visualization and facilitates the surgical procedure using minimally invasive incisions [5]. We report a case of robot-assisted thoracoscopic resection of an esophageal leiomyosarcoma.

Case Presentation

A 64-year-old man with a past medical history of smoking initially presented to our unit with a 10-months history of retrosternal chest pain. The pain had to be controlled with opioids such as fentanyl patch and oral morphine. Initial thoracic Computed Tomography (CT) scan showed no explanation for his pain. However, symptoms persisted and a surveillance CT performed 6 months later demonstrated a new 2.1 cm left paraesophageal mass without thoracic lymphadenopathy and close contact with the thoracic aorta (Figure 1). Further investigations performed included a Positron Emission Tomography (PET)-CT, showing a 3 cm left paraesophageal solid mass with high FDG uptake (Figure 2); and an Endoscopic Ultrasonography (EUS) demonstrating a 3.4 cm × 2.7 cm esophageal mass at 38 cm from the incisors, separate from the muscularis propria layer without any involvement of the aorta (Figure 3). EUS biopsies revealed the mass to be a leiomyosarcoma

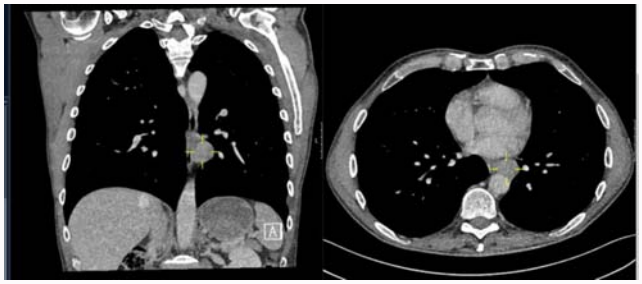

Figure 1: CT scan showing 2.1 cm left paraesophageal mass in close connection with the aorta and pulmonary vein.

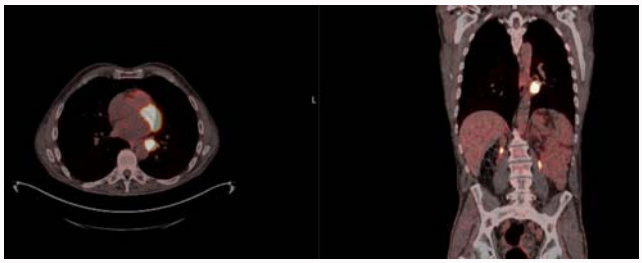

Figure 2: PET-CT showing 3 cm left paraesophageal solid mass with high FDG uptake.

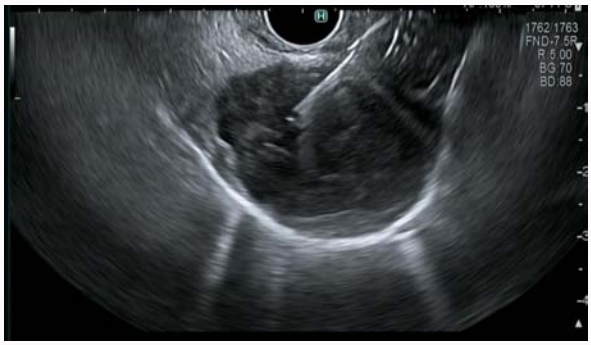

Figure 3: EUS did not show invasion of the aorta or pulmonary vein. Fine Needle Aspiration was performed.

(Immunohistochemistry staining was positive for desmin, Smooth Muscle Actin (SMA), CD34, and low Ki-67 expression).

The patient subsequently underwent robot-assisted left sided thoracoscopic resection. An intraoperative gastroscopy was performed prior to resection which showed no obvious intramucosal extension of tumor. Surgery was performed in the right lateral semi-prone position due to the location of the lesion. Trocars were placed as follows: Camera in the 6th Intercostal Space (IC) and 3 working trocars in the 4th, 8th and 10th ICs with an assistant port in the 5th IC. The tumor was located in the mid-thorax arising from the esophagus with a pedicle to the esophageal wall and it was resected en bloc along with the surrounding pleura. There was no invasion into the aorta or pulmonary vein. Connections to the thoracic duct were clipped and the pedicle was secured with a Hem-o-Lok. The tumor was removed with an endobag *via* extension of the 6th IC space wound. Clips were applied to the resection base in the event postoperative radiotherapy was needed (Figure 4). The patient made a full recovery and was sent home 2 days after the surgery without any complications. Pathology report described a Grade 2 Sarcoma according to FNCLCC with a diameter of 4.0 cm, with free resection margins.

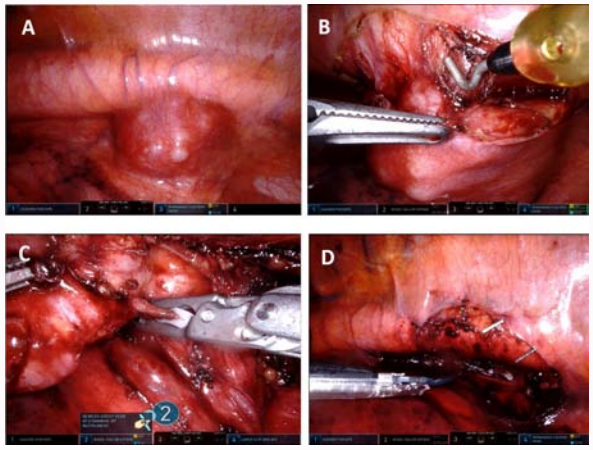

Figure 4: A) Thoracoscopic view of the tumor in the middle esophagus, near to the aorta. B) Dissection of mediastinal pleura and liberation of attachments mobilizing the tumor. C) Identification of esophageal pedicle which is clipped with Hem-o-Lok to proceed to final resection. D) Resection base marked with clips.

Discussion

Leiomyosarcoma is the most common of the esophageal sarcomas with only 200 cases reported in the literature and three case series published [6]. Esophageal leiomyosarcoma represents approximately 0.1% to 0.5% of malignant esophageal tumors [3]. The most common presentation is with progressive dysphagia secondary to intraluminal compression, other symptoms are odynophagia, chest pain, weight loss, cough, emesis [4]. As Zhang et al. described in his series of 12 cases, 66.7% of esophageal leiomyosarcoma occur in the fifth and sixth decades of life and are most commonly located in the middle and lower thoracic esophagus. They have been typically divided into two kinds: Polypoid type in 60% of cases and infiltrative type in 40% of cases [2]. Metastatic disease and lymph node involvement is uncommon but regional lymph node metastases may rarely occur [2,7].

The diagnosis of esophageal leiomyosarcoma is very rare, only 20% to 30% are diagnosed before surgery [3]. A CT scan and EUS are very useful in this kind of tumors, they allow to describe the growth pattern (intraluminal and/or exophytic) [2], size and extent of the tumor and to take samples for preoperative histopathological characterization performing a Fine-Needle Aspiration (FNA) [3,4]. EUS-FNA may be a safe and accurate method in distinguishing the leiomyosarcoma from other various esophageal tumors and may help guide the therapy. F-18 fluorodeoxyglucose Positron Emission Tomography/CT (PET/CT) has taken a role in the diagnosis of leiomyosarcoma, but its differentiation from leiomyoma needs further assessment [2].

The treatment of choice for the majority of patients with esophageal leiomyosarcoma is surgical esophagectomy [7]. Even in the cases that resectable metastasis exists, salvage surgery should be performed to prolong survival [2]. Radical excision performing an esophagectomy has been the standard treatment, since the tumor is not chemo-radio sensitive. However, a few reports describe that local excision of polypoid tumors could achieve good survival rate [2,7]. As regional node metastases may occur in some cases, adjacent lymph node dissection should be considered to reduce the locoregional relapse [2]. The growth type was reported to be associated with survival, polypoid or intramural growth types have a better prognosis

than the infiltrating type [3]. Zhang et al., reported in his group that 3-, 5-, and 10-year survival rates after complete resections were 80.0%, 58.3%, and 31.1%, respectively [2]. Some authors claimed that adjuvant therapy and postoperative radiation may be helpful in controlling the disease and prolong survival in some cases [8].

The minimally invasive approach for the surgical treatment of the esophageal leiomyosarcoma is poorly described in the literature and the majority have been resected through an open approach. Manipadam et al., stated that the reason why the thoracotomy is preferred is that these tumors have been known to reach sizes as large as 16 cm and present as mediastinal masses with deep ulcerations [6]. In other hand, other case series describe that the dissection of the expanded esophagus was easily completed in most cases [2]. Hatch et al. reported that mortality associated with surgery for leiomyosarcoma of the esophagus is high in the immediate postoperative period due to pulmonary and anastomotic complications [9]. This emphasizes the importance of approaching esophageal leiomyosarcoma by minimally invasive approach even in the presence of large tumors [6].

In our case we show that a 3 cm to 4 cm esophageal polypoid leiomyosarcoma can be safely enucleated using a robot-assisted approach, with correct oncological results, minimal postoperative morbidity and a full recovery after surgery. Robotic-assisted minimally invasive surgery offers significant advantages with three-dimensional vision, hand-eye coordination, tremor filtration, resulting in decreased surgical mortality, intraoperative blood loss, hospital stay and quicker postoperative return to normal activities than laparotomy [5].

Conclusion

Esophageal leiomyosarcoma has a good prognosis, and radical resection achieves good results. Polypoid type, early stage, and well differentiation of the tumor has been shown to be favorable prognostic factors [2].

This report shows that the robot-assisted minimally invasive thoracoscopic approach should be considered a feasible option for resection of esophageal leiomyosarcoma due to its benefits on dissection in a limited space, improved precision and complications reduction [5].

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