**Name of journal:** *World Journal of Gastroenterology*

**ESPS Manuscript NO: 2751**

**Columns: CASE REPORT**

**A case of rapidly progressing leiomyosarcoma combined with squamous cell carcinoma in the esophagus**

Jang SS *et al*. A rapidly progressing esophageal leiomyosarcoma

Su Sun Jang, Woo Tae Kim, Bong Suk Ko, Eun Hae Kim, Jong Ok Kim, Kuhn Park, Seung Woo Lee

**Su Sun Jang, Woo Tae Kim, Bong Suk Ko, EunHae Kim, Seung Woo Lee,** Division of Gastroenterology, Department of Internal Medicine, Daejeon St. Mary’s Hospital, Catholic University of Korea, Daejeon 301-723, South Korea

**Jong Ok Kim**, Department of Pathology, Catholic University of Korea, Daejeon St. Mary’s Hospital, Daejeon 301-723, South Korea

**Kuhn Park**, Department of thoracic and cardiovascular surgery, Catholic University of Korea, Daejeon St. Mary’s Hospital, Daejeon 301-723, South Korea

**Author contributions:** Jang SS designed and wrote the case report; Kim WT, Ko BS, Kim EH and Park K revised the report critically for important intellectual content; Kim JO provided the figures and discussion of the pathology; Lee SW approved the final version to be published.

**Correspondence to: Seung Woo Lee, MD,** Division of Gastroenterology, Department of Internal Medicine, Daejeon St. Mary’s Hospital, Catholic University of Korea, 520-2 Dae Heung Dong, JoongGu, Daejeon 301-723, South Korea. leeseungw00@hanmail.net

**Telephone:** +82-42-220-9501 **Fax:** +82-42-252-6807

**Received:** March 12, 2013 **Revised:** June 15, 2013

**Accepted:** July 18, 2013

**Published online:**

**Abstract**

Esophageal leiomyosarcoma is a rare tumor that accounts for less than 1% of all malignant esophageal tumors. Esophageal leiomyosarcoma combined with squamous cell carcinoma is even rarer than solitary leiomyosarcoma. We experienced a case of leiomyosarcoma combined with squamous cell carcinoma that progressed very rapidly.

© 2013 Baishideng. All rights reserved.

**Key words:** Leiomyosarcoma, Carcinoma, Squamous cell, Esophagus, Sarcoma

**Core tip:** We performed esophagectomy with esophago-gastrostomy to resect the tumor. The pathologic examination of surgical specimen revealed that it was combined with squamous cell carcinoma. It should be considered whether there is a combined carcinoma when leiomyosarcoma shows rapid progression.

Jang SS, Kim WT, Ko BS, Kim EH, Kim JO, Park K, Lee SW. A case of rapidly progressing leiomyosarcoma combined with squamous cell carcinoma in the esophagus

**Available from:**

**DOI:**

**INTRODUCTION**

Leiomyosarcoma of the esophagus is a rare malignant tumor, accounting for less than 1% of all malignant esophageal tumors[[1-3](#_ENREF_1)]. Esophageal leiomyosarcoma combined with squamous cell carcinoma is even rarer than solitary leiomyosarcoma. Simultaneous esophageal leiomyosarcoma and squamous cell carcinoma were first described by Ovens *et al*[[4](#_ENREF_4)] in 1951. Leiomyosarcomas are characterized by slow growth and late metastases and hence have a better prognosis than squamous cell carcinomas of esophagus[[5](#_ENREF_5),[6](#_ENREF_6)]. But we experienced a case of leiomyosarcoma combined with squamous cell carcinoma that progressed very rapidly. We report this case and review the literature.

**CASE REPORT**

A 72–year–old male visited to our hospital with chest pain of a months’ duration. The patient diagnosed with colon cancer and received laparoscopic surgeryone year ago. The physical examination was unremarkable. He admitted to the cardiology department, and received electrocardiogram and cardiac single photon emission computed tomography, but no cardiac problem was found. Endoscopic examination demonstrated an intraluminal polypoid mass with stalk in the mid esophagus, 30 cm from the incisor (Figure 1). The tumor was large to fill the esophageal lumen, but allowed passage of the gastrofiberscope(Q260, Olympus, Tokyo, Japan) to the distal part of the esophagus. Endoscopic biopsy was performed, and the patient was suspected of leiomyoma and leiomyosarcoma. The computed tomography scan showed a large, well enhancing soft tissue mass in the mid esophagus (Figure 2), but no regional lymph node enlargement or liver metastasis. Positron emission tomography/computed tomography (PET-CT) showed intense segmental F-18 fluorodeoxyglucose (FDG) uptake [Standardized Uptake Value (SUV) max 17.3] at mid thoracic esophagus. Compared with the previous PET-CT for colon cancer follow-up from 3 mo prior, there was only physiologic FDG uptake at esophagus (Figure 3). The patient underwent surgery, anesophagectomy with esophago-gastrostomy. Macroscopically the resected specimen was a polypoid tumor measuring 9.8 cm × 5.0 cm × 2.5 cm (Figure 4). Histopathologically, the tumor consisted of pleomorphic spindle cells with mitosis and cell necrosis compatible with leiomyosarcoma (Figure 5A). Tumor invasion involved the muscularis propria, submucosa and mucosa. Nine regional lymph nodes were free of metastasis. Immunohistochemical examination stained positive for smooth muscle actin, but negative for cytokeratin and S-100 protein (Figure 5B). These were stained by automated Ventana immunohistochemical / in situ hybridization staining platforms machine (BenchMark XT). Also squamous severe dysplasia and focal stratified squamous epithelial invasion into lamina propria was noted in mucosa (Figure 5C). We diagnosed the patient as leiomyosarcoma combined with squamous cell carcinoma.

In the postoperative period, the patient recovered uneventfully and was discharged 18 d after operation. No adjuvant radiotherapy or chemotherapy was administered. At the last follow up visit to our hospital 5 mo after surgery, the patient wasin good condition without any recurrence or distant metastasis.

**DISCUSSION**

Leiomyosarcoma is a high grade, smooth muscle soft tissue tumor that can occur in any tissue containing smooth muscle fibers. A leiomyosarcoma combined with squamous cell carcinoma is an extremely rare disease of the esophagus. Very few cases have been described in esophagus[[4](#_ENREF_4),[5](#_ENREF_5),[7-9](#_ENREF_7)]. Leiomyosarcomas are most commonly located in the middle and lower thoracic esophagus because smooth muscle predominates in that area[[10](#_ENREF_10)]. Esophageal leiomyosarcomasare typically divided into two types: polypoid type in 60% of cases and infiltrative type in 40% of cases[[11](#_ENREF_11),[12](#_ENREF_12)]. Our case was the polypoid type. The prognosis of esophageal leiomyosarcoma is better than esophageal squamous cell carcinoma because of its characteristics of slow growth and late metastases[[5](#_ENREF_5),[6](#_ENREF_6)]. Patients with polypoid and intramural tumors, tumors in an intrathoracic location and well differentiated tumors have a better prognosis than patients with infiltrating lesion, tumors in cervical location and poorly differentiated tumors[[13](#_ENREF_13),[14](#_ENREF_14)]. Koga *et al*[[13](#_ENREF_13)]reported a case of esophageal leiomyosarcoma that grew rapidly and had a poor prognosis. We think this case is unique because the tumor had good prognostic factors such as the polypoid type and intrathoracic location but it grew very rapidly and was combined with squamous cell carcinoma. Erogluet al. suggested that intermutability or metaplasia between mesenchymal and epithelial tissues or multipotent stem cells with the ability to undergo biphasic differentiation toward mesenchymal and epithelial elements could be a mechanism of this combined malignancy[[7](#_ENREF_7)]. It is possible that these are separate entities that have arisen independently and combined squamous cell carcinoma may affect the growth of leiomyosarcoma by cytokines or growth factors.

The role of FDG-PET-CT in the diagnosis of leiomyosarcoma was reported recently[[15-17](#_ENREF_15)]. Our case showed intense FDG uptake on PET-CT. The standard treatment is esophagectomy but the role of adjuvant radiotherapy or chemotherapy is controversial with some authors[[2](#_ENREF_2),[6](#_ENREF_6),[14](#_ENREF_14),[18](#_ENREF_18)]. In our case, leiomyosarcoma grew exceptionally rapidly and was combined with squamous cell carcinoma, so further research will be needed to reveal the relationship of leiomyosarcoma and squamous cell carcinoma.

**REFERENCES**

1 **Almeida JM**. Leiomyosarcoma of the esophagus. *Chest* 1982; **81**: 761-763 [PMID: 7075315 DOI: 10.1378/chest.81.6.761]

2 **Choh JH**, Khazei AH, Ihm HJ. Leiomyosarcoma of the esophagus: report of a case and review of the literature. *J Surg Oncol* 1986; **32**: 223-226 [PMID: 3736064]

3 **Galandiuk S**, Hermann RE, Cosgrove DM, Gassman JJ. Cancer of the esophagus. The Cleveland Clinic experience. *Ann Surg* 1986; **203**: 101-108 [PMID: 3942414]

4 **OVENS JM**, RUSSELL WO. Concurrent leiomyosarcoma and squamous carcinoma of the esophagus. *AMA Arch Pathol* 1951; **51**: 560-564 [PMID: 14818533]

5 **CAMISHION RC**, GIBBON JH, TEMPLETON JY. Leiomyosarcoma of the esophagus: review of the literature and report of two cases. *Ann Surg* 1961; **153**: 951-956 [PMID: 13690145]

6 **Levine MS**, Buck JL, Pantongrag-Brown L, Buetow PC, Hallman JR, Sobin LH. Leiomyosarcoma of the esophagus: radiographic findings in 10 patients. *AJR Am J Roentgenol* 1996; **167**: 27-32 [PMID: 8659399]

7 **Eroğlu A**, Kürkçüoğlu C, Karaoğlanoğlu N, Erdoğan F, Polat P. Simultaneous leiomyosarcoma and squamous cell carcinoma of the esophagus: report of a new case. *Dis Esophagus* 2001; **14**: 245-246 [PMID: 11869330]

8 **Gaede JT**, Postlethwait RW, Shelburne JD, Cox JL, Hamilton WF. Leiomyosarcoma of the esophagus. Report of two cases, one with associated squamous cell carcinoma. *J Thorac Cardiovasc Surg* 1978; **75**: 740-746 [PMID: 642571]

9 **RELLA AJ**, FARRELL JT, COMER JV. CONCURRENT LEIOMYOSARCOMA AND SQUAMOUS CELL CARCINOMA OF ESOPHAGUS. *N Y State J Med* 1965; **65**: 1254-1256 [PMID: 14293557]

10 **Zhang BH,** Zhang HT, Wang YG. Esophageal leiomyosarcoma: clinical analysis and surgical treatment of 12 cases. *Dis Esophagus* 2012; [Epub ahead of print] [PMID: 23163522 DOI: 10.1111/j.1442-2050.2012.01444.x]

11 **Hatch GF**, Wertheimer-Hatch L, Hatch KF, Davis GB, Blanchard DK, Foster RS, Skandalakis JE. Tumors of the esophagus. *World J Surg* 2000; **24**: 401-411 [PMID: 10706912]

12 **Patel SR**, Anandarao N. Leiomyosarcoma of the esophagus. *N Y State J Med* 1990; **90**: 371-373 [PMID: 2385391]

13 **Koga H**, Iida M, Suekane H, Aoyagi K, Yao T, Kimura Y, Masuda N, Fujishima M. Rapidly growing esophageal leiomyosarcoma: case report and review of the literature. *Abdom Imaging* 1995; **20**: 15-19 [PMID: 7894289]

14 **Rocco G**, Trastek VF, Deschamps C, Allen MS, Miller DL, Pairolero PC. Leiomyosarcoma of the esophagus: results of surgical treatment. *Ann Thorac Surg* 1998; **66**: 894-86; discussion 897 [PMID: 9768947]

15 **Grover RS**, Kernstine K, Krishnan A. A case of diffuse large B-cell lymphoma in association with paraesophageal leiomyoma: highlighting false-positivity of PET scan and importance of tissue diagnosis. *J Natl Compr Canc Netw* 2012; **10**: 577-581 [PMID: 22570288]

16 **Kao YH**, Saad U, Tan AE, Magsombol BM, Padhy AK. Fluorine-18-fluorodeoxyglucose PET/CT for the evaluation of suspected recurrent uterine leiomyosarcomas. *Acta Radiol* 2011; **52**: 463-466 [PMID: 21498277 DOI: 10.1258/ar.2011.100509]

17 **Manohar K**, Mittal BR, Kashyap R, Bhattacharya A, Kakkar N, Mete UK. F-18 fluorodeoxy glucose positron emission tomography/computed tomography findings in a rare case of penile leiomyosarcoma. *J Clin Imaging Sci* 2011; **1**: 58 [PMID: 22267993 DOI: 10.4103/2156-7514.90955]

18 **Perch SJ**, Soffen EM, Whittington R, Brooks JJ. Esophageal sarcomas. *J Surg Oncol* 1991; **48**: 194-198 [PMID: 1658495]

**P-Reviewer** Yeudall WA **S-Editor** Zhai HH **L-Editor E-Edito**r

**Figure 1 Endoscopic finding.** A: Intraluminal polypoid mass; B: Stalk of the mass (arrow).

**Figure 2 Computed tomography scan showed a large, homogeneously enhancing soft tissue mass.**

**Figure 3 Positron emission tomography/computed tomography.** A: Positron emission tomography/computed tomography (PET-CT) showed intense segmental FDG uptake (SUV max 17.3) at mid esophagus; B: PET-CT performed at 3 mo ago.

**Figure 4 Resected specimen measured about 9.8 cm × 5.0 cm × 2.5 cm.**

**Figure 5 Pathologic images.** A: Pleomorphic spindle cells showing mitosis and cell necrosis compatible with leiomyosarcoma (HE stain, **×**200); B: Immunohistochemical stain was positive for smooth muscle actin (**×**12); C: Squamous severe dysplasia and focal stratified squamous epithelial invasion into lamina propria was noted in mucosa (HE stain, **×**100).