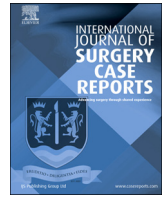


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Esophageal carcinosarcoma in which the sarcomatous element has sloughed off: A case report

Noriyuki Sasaki^{a,*}, Takeshi Iwaya^{a,*}, Yuji Akiyama^a, Shigeaki Baba^a, Fumitaka Endo^a, Haruka Nikai^a, Ryosuke Fujisawa^a, Toshimoto Kimura^a, Takeshi Takahara^a, Koki Otsuka^a, Hiroyuki Nitta^a, Yusuke Kimura^b, Keisuke Koeda^c, Ryo Sugimoto^d, Noriyuki Uesugi^d, Tamotsu Sugai^d, Akira Sasaki^a

^a Department of Surgery, Iwate Medical University School of Medicine, 2-1-1 Idaidori, Yahaba, Iwate 028-3695, Japan

^b Department of Palliative Medicine, Iwate Medical University School of Medicine, 2-1-1 Idaidori, Yahaba, Iwate 028-3695, Japan

^c Department of Medical Safety Science, Iwate Medical University School of Medicine, 2-1-1 Idaidori, Yahaba, Iwate 028-3695, Japan

^d Department of Molecular Diagnostic Pathology, Iwate Medical University School of Medicine, 2-1-1 Idaidori, Yahaba, Iwate 028-3695, Japan

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ABSTRACT

INTRODUCTION: Most esophageal carcinosarcoma (ECS) tumors present as a polypoid tumor that is continuous with the superficial lesion and suspended by a pedicle. Here, we report a case of ECS in which a polypoid lesion sloughed off before surgery.

PRESENTATION OF CASE: A 76-year-old man with dysphagia was admitted to our hospital. Esophagogastroscopy revealed a 20-mm polypoid tumor continuous with a superficial lesion and attached to the lesion by a thin pedicle in the mid-thoracic esophagus. Histopathological examination of the endoscopic biopsy showed that the superficial lesion was a moderately differentiated squamous cell carcinoma and that the polypoid tumor contained a sarcomatous element. He was diagnosed with ECS and underwent radical esophagectomy with three-field lymph node dissection. In the resected specimen, no polypoid tumor was found, and only a superficial lesion was observed. The histopathological findings revealed only squamous cell carcinoma, and the pathological diagnosis was esophageal squamous cell carcinoma, pT1bN0M0, pathological stage I. The patient was discharged from the hospital 22 days after surgery and did not experience any complications. He is currently alive and remained cancer-free for three years since surgery was performed.

DISCUSSION: Due to the distinctive configuration in which the polypoid lesion was connected to the superficial cancerous lesion by a very thin pedicle, researchers suggested that the polypoid tumor, which consisted of a sarcomatous element, was sloughed off before surgery.

CONCLUSION: We encountered a rare case of ECS in which the sarcomatous element sloughed off prior to surgical resection.

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1. Introduction

Carcinosarcoma is a neoplasm that contains both carcinomatous and sarcomatous elements. Esophageal carcinosarcoma (ECS) is a rare disease that accounts for 1–2% of all esophageal malignant neoplasms [1,2]. Many ECSs present as a polypoid configuration with the tendency towards endoluminal growth. The polypoid lesion of this tumor often consists of a sarcomatous element that is continuous with superficial squamous cell carcinoma [3]. The car-

cinomatous element is typically limited to only a small portion of the ECS tumor and is often located at the base of or around the polypoid lesion. In addition, the depth of tumor invasion in ECS is often limited to the superficial tumor layer [4]. On the contrary, ulcerative-type ECSs are often associated with a poor prognosis due to deeper tumor invasion compared with polypoid-type tumors [4,5]. Therefore, tumor type may be an important factor that can predict prognosis and guide the treatment plan.

Due to the distinctive configuration of ECS such as large tumor size and strong endoluminal growth, patients with this tumor type typically complain of dysphagia starting at the early stage. Therefore, ECS is often discovered earlier than esophageal squamous cell carcinoma. However, several reports suggested that the frequency of lymph node metastasis in ECS is equivalent to that of squamous cell carcinoma [6–8]. Although the prognosis of ECS patients was

Abbreviations: CT, Computed tomography; ECS, Esophageal carcinosarcoma.

* Corresponding authors.

E-mail addresses: nsasaki@iwate-med.ac.jp (N. Sasaki), tiwaya@iwate-med.ac.jp (T. Iwaya).

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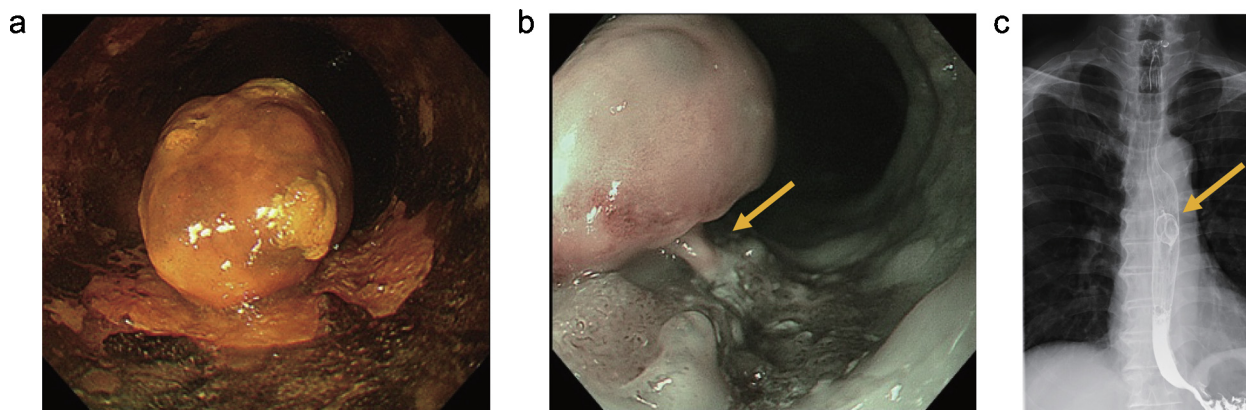


Fig. 1. Esophagogastroscopy and barium esophagography were performed for esophageal carcinosarcoma on the patient's first visit. (a) Esophagogastroscopy with Lugol staining revealed a superficial lesion with a polypoid mass in the middle thoracic esophagus. (b) The polypoid mass was continuous with the superficial lesion and was attached by a thin pedicle (arrow). (c) A barium esophagogram revealed a 20-mm mass in the middle thoracic esophagus (arrow).

considered equivalent to or slightly better than that of patients with squamous cell carcinoma, this concept is still controversial [5,6,9,10]. The possibility exists that the stratification of patients with ECS according to tumor type may provide clearer evaluation of treatment outcomes in these patients. Because ECS is rare, the accumulation of information on each type of ECS is likely important.

Here, we report a case of ECS in which a polypoid tumor that was continuous with the superficial lesion and attached by a thin pedicle sloughed off before surgery. This work has been reported in accordance to the Surgical Case Report (SCARE) guidelines [11].

2. Case report

A 76-year-old man with dysphagia was admitted to our hospital. Esophagogastroscopy revealed a polypoid tumor that was continuous with the superficial lesion and attached by a thin pedicle in the mid-thoracic esophagus 28 cm from the incisor teeth (Fig. 1a, b). A barium esophagogram revealed a polypoid tumor, 20 mm in diameter, in the mid-thoracic esophagus (Fig. 1c). A computed tomography (CT) scan revealed a tumor with contrast enhancement in the mid-thoracic esophagus. No apparent lymph node metastasis or distant metastasis was observed. Histopathological examination of the endoscopic biopsy specimen showed moderately differentiated squamous cell carcinoma in the superficial lesion and a sarcomatous element consisting of atypical spindle-shaped cells in the polypoid portion of the tumor (Fig. 2). The

patient was diagnosed with ECS, clinical T1N0M0, stage I (according to the TNM Classification of Malignant Tumors, 8th Edition). Radical esophagectomy with three-field lymph node dissection and gastric tube reconstruction was performed using a thoracoscopic approach with the patient in the prone position; laparotomy and bilateral cervical approaches were also used. Posterior mediastinal route reconstruction was achieved using a gastric tube. The surgical duration was 451 min, and total blood loss was 53 ml.

In the resected specimen, only a Lugol-unstained superficial lesion 20 × 16 mm in size was observed in the upper thoracic esophagus, whereas the polypoid tumor already sloughed off (Fig. 3). An ulcer, 9 × 7 mm in size, was observed in the center of the superficial lesion, where the polypoid tumor was piled up by its root. Pathologically, only squamous cell carcinoma that invaded the muscularis mucosae was observed around the central ulcer (Fig. 4), as the sarcomatous element was not observed in the resected specimen. In addition, no lymph node metastasis was seen. Finally, the tumor was histologically diagnosed as esophageal squamous cell carcinoma, pT1bN0M0, pathological stage IB (according to the TNM Classification of Malignant Tumors, 8th Edition). The patient was discharged from the hospital 22 days after surgery and did not experience any complications. He is currently alive and has been relapse-free for 41 months. The patient communicated a change in gastrointestinal symptoms, namely, that he experienced constipation one week before surgery and that he passed a spherical hard stool.

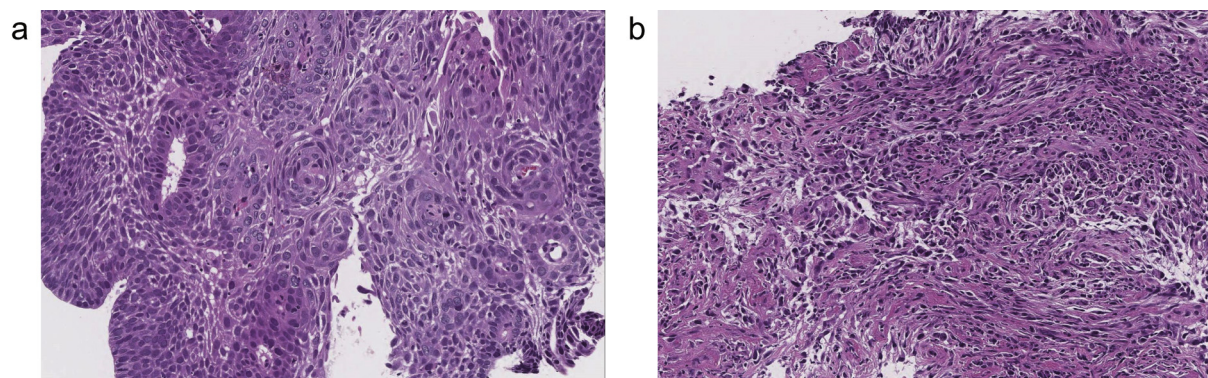


Fig. 2. Histopathological examination of endoscopic biopsy specimens from the esophagus (hematoxylin-eosin stain; high-power field). (a) Pathological finding of the biopsy specimen from the superficial lesion showed moderately differentiated squamous cell carcinoma. (b) Pathological finding of the biopsy specimen from the polypoid mass showed atypical spindle-shaped cells.

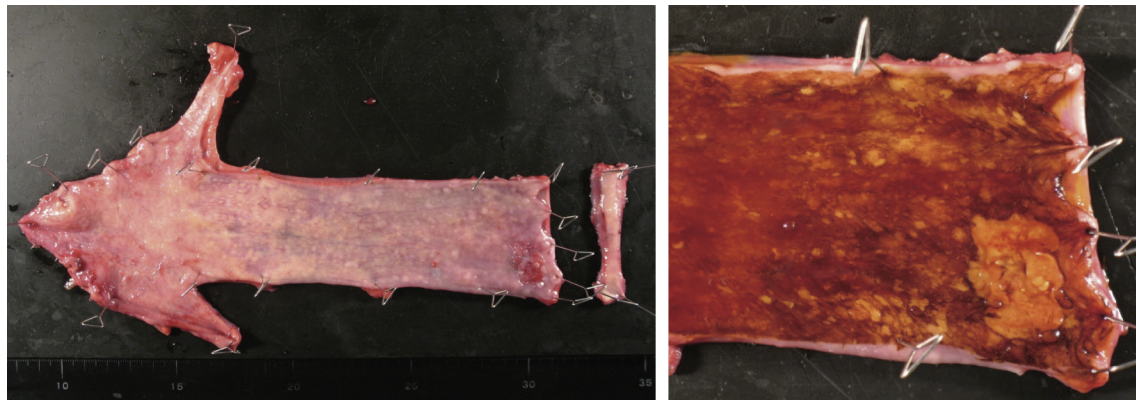


Fig. 3. Esophageal surgical specimen. No polypoid mass was observed, and only the superficial lesion with erosion was detected.

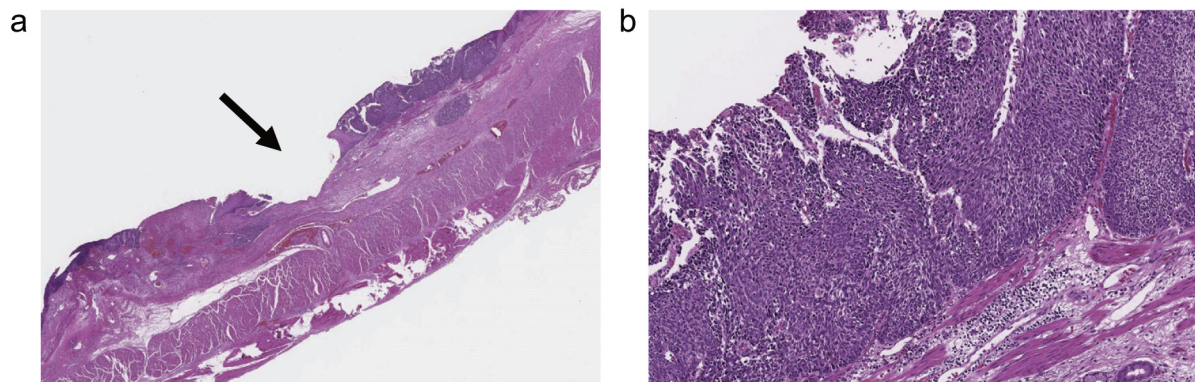


Fig. 4. Pathological examination of surgical specimens. A slightly depressed lesion with erosion was observed (arrow). Atypical squamous cells invaded the muscularis mucosae. (a) Hematoxylin-eosin stain; 40× magnification. (b) Hematoxylin-eosin stain; high-power field.

3. Discussion

Our patient was diagnosed with ECS according to the preoperative biopsy from both superficial and polypoid lesions. As the polypoid tumor was not present in the resected specimen, the final pathological diagnosis of the tumor was esophageal squamous cell carcinoma. Due to the distinctive configuration in which the polypoid lesion was suspended from the superficial cancerous lesion by a very thin pedicle, researchers easily recognized that the polypoid tumor consisting of a sarcomatous element was sloughed off before surgery. The polypoid tumor in the middle thoracic esophagus was detected by preoperative barium esophagogram, as shown in Fig. 1c, but in the resected specimen, the superficial cancerous lesion was located in the upper thoracic esophagus. This suggested that the polypoid tumor was suspended from the superficial cancerous lesion by a thin pedicle due to gravity and the peristaltic motion of the esophagus when the patient was in an upright position.

We searched PubMed and the Japan Medical Abstracts Society database for literature on ECS in which the polypoid tumor was sloughed off during the course of the disease. Three other reported cases are listed in the Table 1. Nakano et al. reported two cases in which exfoliation of the polypoid tumors was confirmed in the resected specimens, similar to our case [12]. Yamashita et al. reported a case in which exfoliation of the polypoid tumor was noted by esophagoscopy [13]. Although the frequency of exfoliation of the polypoid lesion of ECS is unknown, many factors such as tumor diameter, location, thickness/length of the pedicle, and esophageal caliber may be involved. The size of the sloughed off polypoid tumors ranged from 20 mm to 60 mm in diameter (Table 1). ECS cases with a larger polypoid lesion connected by a thin

pedicle with no exfoliation were also reported [12]. The possibility exists that a larger size of polypoid lesions and tumor location just above the natural constriction sites of the esophagus reduce tumor mobility and prevent its exfoliation. In our case, only squamous cell carcinoma components were observed in the resected specimens. Yamashita et al. reported an ECS case in which another subpedunculated tumor grew at the same site one month after exfoliation of the original tumor [13]. Therefore, the possibility exists that a remnant sarcomatous component after exfoliation may induce regrowth of another polypoid lesion.

ECS is generally treated in the same manner as squamous cell carcinoma. For early-stage ECS, esophagectomy with lymphadenectomy is typically performed. For advanced-stage ECS, some reports demonstrated remission of the polypoid tumor as a result of chemotherapy or radiation therapy [14,15]. However, the efficacy of chemotherapy and radiation therapy was not proven as many ECS cases are resistant [16]. Notably, it is possible that some ECS cases in which exfoliation of the polypoid lesion occurred during chemotherapy or chemoradiotherapy are misdiagnosed as responders to therapy.

In terms of prognosis, although ECS exhibits a significantly lower 5-year survival rate than esophageal squamous cell carcinoma in T1 cases (47.6% vs. 84.2%) [8], a significant difference was not observed in the 5-year survival between advanced ECS and squamous cell carcinoma, which is probably due to the high recurrence rate late in the disease course (26.7% vs. 22.4%) [10]. These reports suggested that ECS might exhibit oncologic characteristics that differ from those of squamous cell carcinoma. The pathogenesis of ECS was not elucidated due to its rarity. Although ECS accounts for only 2% of all esophageal malignancies, some ECS cases may be misdiagnosed as squamous cell carcinoma if the polypoid lesion is naturally

Table 1
Reports of esophageal carcinosarcoma in which the sarcomatous element sloughed off.

Case	Author	Sex	Age	Chief complaint	Location	Longest diameter (mm)	T*	N*	M*	Pathological stage*	Recognition of sloughing	Treatment	Prognosis	Survival (M)
1	Yamashita et al.	M	55	Dysphagia	Lt	NA	1b	0	0	I	Preoperative esophagoscopy	Surgery	Alive	69
2	Nakano et al.	M	58	Dysphagia	Mt	60	1b	0	0	I	Surgical specimen	Surgery	Death due to cerebral hemorrhage	1
3	Nakano et al.	M	80	Dysphagia	Lt	30	1b	2	0	II	Surgical specimen	Surgery	Alive	27
4	Our case	M	76	Dysphagia	Mt	20	1b	0	0	I	Surgical specimen	Surgery	Alive	36

* TNM Classification of Malignant Tumors, 8th Edition; NA, not available.

sloughed off by the time the patient first visits a hospital, similar to the present case. In other words, the frequency of ECS may actually be higher. Because the oncologic characteristics of ECS are unclear, collecting more detailed information on this rare tumor type is important due to the possibility of a diagnosis of ECS where the polypoid lesion sloughed off.

4. Conclusion

Here, we reported a case of ECS in which the polypoid tumor consisting of a sarcomatous element sloughed off before surgery.

Declaration of Competing Interest

None.

Funding

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Ethical approval

Ethic approval has been exempted by Clinical Research Ethics Committee of Iwate Medical University.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

Author contribution

NS and TI conceived the case presentation and drafted the manuscript. RS, NU, and TS interpreted pathological aspects of the case. YA, TK, TT, KO, HN, YK, and KK participated in the design of the case presentation. SB, FE, HN and RF took care of the management of the patient. AS read and approved the final manuscript.

Registration of research studies

This case report does not require registration as a research study.

Guarantor

Takeshi Iwaya.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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