CASE REPORT

A huge intraluminally growing polypoid tumor of the cervical esophagus: A case report and literature review of spindle cell (undifferentiated pleomorphic) sarcoma

Che-Jui Yang a, Sen-Ei Shai a,b,*, Wan-Shan Li c, Chung-Ping Hsu a,b

a Division of Thoracic Surgery, Department of Surgery, Taichung Veterans General Hospital, Taichung, Taiwan
b National Yang-Ming University, School of Medicine, Taipei, Taiwan
c Department of Pathology, Taichung Veterans General Hospital, Taichung, Taiwan

Received 10 July 2012; received in revised form 31 August 2012; accepted 1 October 2012
Available online 6 April 2013

KEYWORDS
esophagectomy; spindle cell sarcoma

Summary Primary sarcomas of the esophagus are rare. We report the case of a 43-year-old man with a high-grade spindle cell (undifferentiated pleomorphic) sarcoma of the cervical esophagus just below the esophageal inlet. A huge tumor initially measuring 10 cm in length abutting the esophageal inlet was found. It was an intraluminal tumor with only a transverse pedicle stalk connected to the esophageal mucosa and had only invaded the submucosal layer. After six courses of neo-adjuvant chemotherapy (over 1 year), the patient underwent total laryngectomy, esophagectomy, and three-field radical lymph node dissection. This patient was disease-free at the most recent follow-up. Although there have been several case reports of undifferentiated pleomorphic sarcoma of the alimentary tract, undifferentiated pleomorphic sarcoma of the esophagus is extremely rare. The behavior of sarcomas of the esophagus and treatments are discussed, and the literature is reviewed.

1. Introduction

Esophageal cancer is the sixth most common cancer in the world, and is especially common among Asian populations. Most esophageal cancers are carcinomas. Primary sarcomas
of the esophagus are rare. Sarcomas and carcinosarcomas have been found to account for approximately 0.1–1.5% of all esophageal tumors,¹ and these comprised only 5% of all gastrointestinal sarcomas.² High-grade undifferentiated pleomorphic sarcoma or so-called pleomorphic malignant fibrous histiocytoma (MFH) in the esophagus is very rare. We report the case of a 43-year-old man with a huge high-grade undifferentiated pleomorphic spindle cell sarcoma of the cervical esophagus abutting the esophageal inlet, who had chemotherapy followed by total esophagectomy. Unlike carcinoma, esophageal sarcoma has a favorable prognosis with radical resection.³ In this article, we describe the features of our case and review the literature.

2. Case report

A 43-year-old man presented with odynophagia, progressive swallowing difficulty, and weight loss of 4 kg in 3 months. The patient had been smoking one pack of cigarettes per day for more than two decades and had a peptic ulcer, which had been under medical control for 5 years. Esophagography disclosed a huge tumor with severe mucosal destruction about 10 cm in length in the upper esophagus (Fig. 1). Endoscopic examination of the esophagus showed a huge tumor with irregular mucosa with ulceration 15 cm from the incisors, and the endoscope could not pass through this region (Fig. 2). Bronchoscopy demonstrated external compression on the posterior wall in the upper to middle trachea with obvious narrowing of up to 50% of the lumen. A computed tomography (CT) scan of the chest and upper abdomen showed a huge mass in the upper third of the esophagus with luminal widening (Fig. 3), but no direct invasion of adjacent structures or regional lymphadenopathy. No evidence of metastasis was found from the positron emission tomography (PET) and whole-body bone scans. The tumor markers α-fetoprotein (AFP), squamous cell carcinoma antigen (SCC), and carcinoembryonic antigen (CEA) were all within normal limits. Histopathological examination via endoscopic biopsy showed high-grade pleomorphic spindle cell proliferation with an intersecting fascicular growing pattern and increased mitotic activities. These tumor cells showed no immunoreaction to AE1/AE3, EMA, desmin, CD117, S100, or CD34. A high-grade pleomorphic sarcoma was diagnosed.

The patient first received neo-adjuvant chemotherapy with the MAID [mesna (700 mg), doxorubicin (15 mg/m²), ifosfamide (2.5 mg/m²), and dacarbazine (250 mg/m²)] regimen. After completion of six courses of chemotherapy, a CT scan of the chest demonstrated that the tumor had mildly shrunk in size. Although smaller, it still caused obvious symptoms such as odynophagia and dysphagia. In this period, the patient could tolerate a liquid diet. His body weight was within an acceptable range. The patient underwent radical surgery 1 year after diagnosis. Rightsided video-assisted minithoracotomy was carried out. The operation consisted of total esophagectomy, cardiectomy, and total laryngectomy with three-field radical lymph node dissection. Reconstruction was performed using a gastric tube transposed to the neck via the posterior mediastinal route. Pylomyotomy and feeding jejunostomy were also performed. During the operation, one huge tumor was noted in the upper esophagus just below the esophageal inlet. Enlarged lymph nodes were found in the paraesophageal and parapancreatic areas.

Figure 1  Esophagography: a huge tumor with severe mucosal destruction about 10 cm in length in the upper esophagus.

Figure 2  Endoscopic findings of esophagus: tumor with irregular mucosa and ulceration 15 cm from the incisors.
The surgical specimen contained a huge intraluminal tumor with only a thin transverse pedicle stalk 3 cm in length connecting it to the esophageal mucosa. The gross size of the tumor was $7 \times 5 \times 3$ cm. It was located near the esophageal inlet (Fig. 4) and had a firm and highly vascular nature. The tumor had only invaded the submucosal layer, and there were adequate peripheral margins of uninvolved tissue in all planes. The second histopathological examination revealed the same tumor morphology as that of the initial examination, and no lymph node metastasis was found (Fig. 5). Immunohistochemical staining was AE1/AE3 (e$\cdot$), EMA (e$\cdot$), desmin (e$\cdot$), CD34 (e$\cdot$), CD-117 (e$\cdot$), S-100 (focal +), CD68 (focal +), CD163 (focal +), CD35 (e$\cdot$), and actin-M851 (focal +). The immunohistochemical staining showed complete absence of reactions with any lineage-selective markers. The diagnosis of high-grade undifferentiated pleomorphic sarcoma was confirmed.

The postoperative course was uneventful, and the patient was discharged in a stable condition. One year after the operation, a CT scan of the chest and an upper gastrointestinal endoscopy showed no tumor recurrence or distant metastasis.

3. Discussion

Primary sarcomas of the esophagus are rare neoplasms. In a previous study, sarcomas and carcinosarcomas accounted for approximately 0.1–1.5% of all esophageal tumors and comprised only 5% of all gastrointestinal sarcomas.$^{1,2}$ Leiomyosarcoma is the most common cell type of the esophageal sarcomas. The rarity of esophageal sarcoma is illustrated by the small number of previous reports, consisting of single or small groups of patients. The clinical symptoms of esophageal sarcomas are nonspecific and different from those of esophageal carcinoma, and include progressive dysphagia, loss of weight, regurgitation, retrosternal pain, respiratory distress, odynophagia, sensation of a lump in the throat, hemorrhage, anemia, sudden death (due to asphyxiation), vomiting (food or tumor fragments), fever, and cough. Our patient presented with odynophagia and dysphagia initially, which was consistent with symptoms in previously reported cases.

Esophageal leiomyosarcoma is classified as polypoid in 60% of cases and infiltrative in 40%.$^{4,5}$ Yamashita et al reviewed seven cases of malignant fibrous histiocytoma in the esophagus; all the cases involved pedunculated or polypoid tumors.$^6$ Those reports suggested that esophageal sarcomas usually present as a polypoid mass. Esophagography is a useful tool for evaluating the morphology of esophageal tumors. Endoscopic examination is useful for assessing the characteristics of the tumor and biopsy can be helpful in differentiating sarcomas from carcinoma.
However, a superficial biopsy may yield a false-negative result, due to the esophageal mucosa overlying the tumor. In our case, we could not completely evaluate the morphology of the tumor by endoscopic examination or esophagography on account of the huge size of tumor, so we were unaware that it was a polypoid lesion until surgical resection.

In our case, the diagnosis of high-grade undifferentiated pleomorphic sarcoma was made through histopathological examination, which revealed a pleomorphic spindle cell appearance and no definitive reaction to immunohistochemical staining. Undifferentiated high-grade pleomorphic sarcoma and so-called pleomorphic MFH are a group of pleomorphic sarcomas that do not demonstrate a definitive line of differentiation, even with immunohistochemistry. The pathological diagnosis of undifferentiated high-grade pleomorphic sarcoma is therefore one of exclusion, which is made in the absence of reaction with any lineage-selective markers in a high-grade pleomorphic sarcoma. Many undifferentiated spindle cell sarcomas were previously classified as MFH. However, with current advances in diagnostic techniques, it is expected that the number of tumors diagnosed as MFH will decrease. Although MFH is common in the extremities and retroperitoneal tissue, it is extremely rare in the esophagus with only a few cases documented so far.

Unlike carcinoma, esophageal sarcoma has a favorable prognosis following radical resection. DeMeester et al reported five cases of polypoid esophageal sarcoma. In their study, in spite of the large size (mean size 7.1 cm) of the tumors, they remained superficial within the esophageal wall, and nodal or distant organ metastasis was absent in four of the five patients. Rocco et al reviewed 17 cases of esophageal leiomyosarcomas and curative resection was carried out in 11 patients. The results showed that curative resection offered a significant survival benefit to the patients and the authors suggested that esophagectomy should be the standard management for most patients with esophageal leiomyosarcoma. We reviewed previous reports of undifferentiated pleomorphic sarcoma, and the eight cases, including ours, are summarized in Table 1. In our case, although the cell type was uncommon (high-grade undifferentiated pleomorphic sarcoma), the clinical presentation was similar to that in previous reports. Although the tumor was huge, it remained superficial within the esophageal wall. However, in our case, the preoperative images and endoscopic findings failed to confirm the degree of invasion of tumor completely; they showed only that the tumor was just at the opening of the esophagus. The total esophagectomy and total laryngectomy were performed to ensure adequate radical resection. After surgical resection, the patient remained disease-free at the most recent follow-up (1 year after surgery). The characteristics of intraluminal polypoid growth appear to allow endoscopic resection in certain selected patients. In some early cancer lesions which are small in size and with an obvious stalk, endoscopic polypectomy may be feasible.

Numerous modes of treatment, including surgical resection, radiation, and chemotherapy have been used either alone or in combination in limited cases. Chemotherapy does not appear to show any benefit in the treatment of esophageal sarcomas. In our case, the initial planned treatment was neo-adjuvant therapy for downstaging. But the neo-adjuvant chemotherapy with the MAID regimen only resulted in a partial response with an initial slight reduction in tumor size. Lokesh et al reported a case of spindle cell sarcoma of the esophagus in a 55-year-old woman managed with radical radiotherapy alone. However, the evidence for successful radiotherapy in esophageal sarcomas is limited and its role in treatment strategies for such lesions has not been well established.

In summary, we report a rare case of spindle cell sarcoma of the esophagus. Our experience and previous

### Table 1

Review of eight cases in men with primary undifferentiated pleomorphic sarcoma in the esophagus, including our case.

<table>
<thead>
<tr>
<th>Reference</th>
<th>Patient age (y)</th>
<th>Symptoms</th>
<th>Location</th>
<th>Appearance</th>
<th>Size (cm)</th>
<th>Surgical procedure</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Our case</td>
<td>43</td>
<td>Odynophagia, dysphagia, hoarseness</td>
<td>Ce-Tl (cervical part of esophagus)</td>
<td>Polypoid tumor</td>
<td>7</td>
<td>Total esophagectomy</td>
<td>Disease free for 4 mo</td>
</tr>
<tr>
<td>Yamashita et al</td>
<td>50</td>
<td>Dysphagia, cough, dyspnoea</td>
<td>Ce (cervical part of esophagus)</td>
<td>Dome-shaped tumor</td>
<td>12</td>
<td>Total esophagectomy</td>
<td>Death within 7 mo</td>
</tr>
<tr>
<td>Naganuma et al</td>
<td>78</td>
<td>Dysphagia</td>
<td>Tm (middle thoracic part of esophagus)</td>
<td>Polypoid tumor</td>
<td>7</td>
<td>Total esophagectomy</td>
<td>Alive within 1 y</td>
</tr>
<tr>
<td>Sápi et al</td>
<td>65</td>
<td>Dysphagia</td>
<td>Tm (middle thoracic part of esophagus)</td>
<td>Pedicled polypoid tumor</td>
<td>3.3</td>
<td>Polypectomy</td>
<td>Alive within 3 mo</td>
</tr>
<tr>
<td>Aagaard et al</td>
<td>67</td>
<td>Dysphagia</td>
<td>Tl (upper thoracic part of esophagus)</td>
<td>Exophytic tumor</td>
<td>12</td>
<td>Esophagogastrectomy</td>
<td>Death within 1 mo</td>
</tr>
<tr>
<td>Geboes et al</td>
<td>59</td>
<td>Dysphagia</td>
<td>Tm (middle thoracic part of esophagus)</td>
<td>Pedunculated exophytic tumor</td>
<td>9</td>
<td>Subtotal esophagotomy</td>
<td>Unknown</td>
</tr>
<tr>
<td>Geboes et al</td>
<td>57</td>
<td>Dysphagia</td>
<td>Tm (middle thoracic part of esophagus)</td>
<td>Pedunculated tumor</td>
<td>6</td>
<td>Esophagotomy</td>
<td>Alive within 1 y</td>
</tr>
<tr>
<td>Takada et al</td>
<td>46</td>
<td>Dysphagia, hoarseness</td>
<td>Tm-Tl (upper thoracic part of esophagus)</td>
<td>Pedunculated tumor</td>
<td>14</td>
<td>Subtotal esophagotomy</td>
<td>Unknown</td>
</tr>
</tbody>
</table>

Ce = cervical part of esophagus; Tl = lower thoracic part of esophagus; Tm = middle thoracic part of esophagus; Tu = upper thoracic part of esophagus.
Evidence suggests that surgical resection can now be carried out with low morbidity and mortality. Once the diagnosis has been established, complete resection of esophageal sarcomas may be performed to achieve optimal benefit. Further studies are required to establish the potential roles of chemotherapy and radiotherapy for spindle cell sarcoma of the esophagus.

References