Case report

Giant liposarcoma of the esophagus with Li-Fraumeni-like syndrome

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Abstract

Primary esophageal liposarcoma is a rare malignancy, with only 25 cases reported in the English literature. In this report, we present a new case of giant esophageal liposarcoma in a 49-year-old woman with a family history of upper gastrointestinal tract cancer. This was a transmural tumor located in the lower esophagus. Subtotal esophagectomy was successfully performed, and histopathology revealed a well-differentiated liposarcoma. The patient was followed-up for 20 months without recurrence. However, asynchronous ovarian serous adenocarcinoma was found 1 year after the esophageal surgery, and Li–Fraumeni-like syndrome was diagnosed.

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1. Introduction

1.1. Case summary

In November 2008, a 49-year-old woman was admitted to our hospital complaining of a 3-year history of dysphagia, which had been aggravated in the previous 2 months. There was no weight loss or change of voice. No remarkable medical history was noted. However, her father had died of esophageal carcinoma and her mother had undergone a subtotal gastrectomy for a gastrointestinal stromal tumor. Physical examination and laboratory data were unremarkable. An initial esophagoscopy showed an extrinsic compression of the lumen of the middle third of the esophagus with an erosive overlying mucosa, starting at 25 cm from the incisors, down to the gastroesophageal (GE) junction (Fig. 1(a)). Barium swallow showed a dilated proximal esophagus with an ellipsoid mass on the wall of the lower esophagus (Fig. 1(b)). A chest computed tomography (CT) scan showed a giant, heterogeneous mass with fat-like density (−48 Hounsfield units (HU)), inseparable from the lateral wall of the mid-lower esophagus (Fig. 1(c)). An enhanced CT scan showed that the mass was mildly enhanced after administration of intravenous contrast material.

The patient underwent a subtotal esophagectomy through a left posterior–lateral thoracotomy, with intrathoracic esophagogastric anastomosis. Intra-operatively, the giant soft-tissue mass was found within the esophageal wall, originating at the level of the aortic arch and extending downward to the cardia (Fig. 2(a)).

1.2. Pathology

Macroscopically, the tumor measured 12 cm × 6 cm × 4 cm, and was covered with normal esophageal mucosa (Fig. 2(b)). A longitudinal section of the mass revealed that its color ranged from ivory to deep yellow (Fig. 2(c)). Histologically, the neoplasm consisted predominantly of vacuolated adipocytes that were similar to well-differentiated adipose tissue. Areas of spindle cells were observed. Myxoid tissue was present, with scattered lipoblasts. Pathologic evaluation of the resected specimen confirmed the presence of a well-differentiated liposarcoma. The resection margins were negative.

The postoperative course was uneventful; on the 14th postoperative day, the patient was discharged with complete resolution of her dysphagia, and was prescribed a soft diet. No further treatment was required, and the patient was followed-up for 20 months without recurrence.

However, in October 2009, the patient was re-admitted, again for persistent abdominal distention. Bilateral ovary lesions were revealed by gynecological ultrasound. Thus, surgical treatment (total abdominal hysterectomy with bilateral salpingo-ophorectomy (TAH-BSO) + subtotal omentectomy) was performed. Pathological evaluation confirmed the presence of a bilateral, poorly differentiated, serous ovarian carcinoma with omental metastasis. The patient received adjuvant chemotherapy, a combined regimen of...
intra-peritoneal carboplatin and intravenous taxane, which was administered for one cycle, and intravenous taxane/carboplatin, which was administered for seven cycles, with partial response. On the basis of the patient’s clinical findings and family history, Li—Fraumeni-like syndrome was suspected [1]. However, the patient refused further genetic tests.
2. Discussion

Primary esophageal liposarcoma is rare type of tumor. The first case was described by Mansour in 1983 [2], and only 25 cases have been reported in the English literature [3—9], with a male predominance (72%). The average age of patients was 58.4 years (range: 38–73 years), and the average tumor size was 13 cm length (range: 4–23 cm). Most lesions were polypoid (24 cases), with only one transmural case. Eighty percent of the liposarcomas were located in the cervical portion of the esophagus. Our case is the second case of a transmural type, and the fourth case located in the distal esophagus.

The reported histologic types of esophageal liposarcoma have included well-differentiated, dedifferentiated, myxoid cell, and pleomorphic types [3—9]. Well-differentiated liposarcoma was the most common, comprising 68% (17/25) of all the lesions. Myxoid liposarcoma was reported in five cases. Dedifferentiated and pleomorphic liposarcomas were reported with only one case each. Our case is the only case to date of a transmural type of well-differentiated tumor; all the others have been polyps.

The therapy of choice for liposarcoma is surgical resection. The surgical approaches are varied and include transcervical, transthoracic, and trans-abdominal resections. Endoscopic resection and a minimally invasive approach have also been reported [4,6,8]. Polypectomy is the most common approach due to the growth pattern and biological behavior of the tumors [3]. We performed an en bloc resection for this transmural tumor with negative margins. Liposarcomas at other sites have been shown to be radiosensitive, and adjuvant radiotherapy is recommended in such cases [4,10]. In the long term, however, only one in 17 followed-up patients with well-differentiated liposarcomas, who had a polypoid type with a polypectomy, showed local recurrence [3—9]. Thus, no adjuvant radiotherapy was administered as we had to perform a radical resection. However, close follow-up was definitely necessary.

Li–Fraumeni-like syndrome is a cancer predisposition syndrome associated with soft-tissue sarcoma, and is a variety of malignancy that shares some, but not all, features of Li–Fraumeni syndrome [1]. We suspected that our patient was an individual with Li–Fraumeni-like syndrome, due to her clinical course and her family’s tumor history. However, we were unable to test for the p53 mutation due to her refusal to permit further genetic testing.

References