Successful resection of esophageal carcinosarcoma in presence of right aortic arch: A case report

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ABSTRACT
We herein report a giant esophageal carcinosarcoma (ECS) occupying the entire intrathoracic esophagus with a coexisting right aortic arch (RAA). The patient was admitted with severe dysphagia and weight loss. Subtotal esophagectomy was performed via left thoraco-cervical approach with cervical esophagogastric anastomosis. The aortopulmonary ligament was ligated and dissected at first for better exposure. Patient recovered uneventfully and there was no evidence of recurrence during the 1 year follow-up period. To our knowledge, this is the first report of such a giant ECS with coexisting RAA. For this kind of patient, a left thoracotomy approach and cervical anastomosis is recommended, and the dissection of aortopulmonary ligament is usually required.

Key Words: Esophageal carcinosarcoma, Right aortic arch, Case report

1. INTRODUCTION
Esophageal carcinosarcoma (ECS) is a rare esophageal malignant tumor. It comprises approximately 0.36%-2.8% of all esophageal malignancies.[1,2] According to a literature search using PubMed, most reported cases were smaller than 17 cm in length, while a tumor occupying the entire esophagus was quite less frequently seen.[3] Right aortic arch (RAA) is an intrathoracic great vessel anomaly that has been reported to be present in 0.04%-0.1% of autopsy cases. Moreover, the coexistence of ECS with RAA is extremely rare but warrants special awareness during the operation.[4-6] The present case of an ESC occupying the entire esophagus with coexisting RAA was even more unusual.

2. CASE PRESENTATION
A 53-year-old man was admitted to our hospital with complaints of a 20-days history of progressive dysphagia, accompanied with a weight loss of nearly 10 kg during the past month. Preoperative TPN was used sometimes in local hospital. Past medical and family history did not include any relevant disease, but was positive for heavy smoking and alcohol consumption. Barium esophagography (see Figure 1A) showed an irregularly-shaped stenosis in the beginning of esophagus. Endoscopy revealed a tumor located at 20 cm from the incisors, extruding into the esophageal lumen with surface ulceration that prevented passage of the endoscope. The computed tomography scan demonstrated an abnormal intrathoracic great vessel anomaly, the RAA (see Figure 2).
thoracotomy through the 5th intercostal space. Firstly, the esophagus was explored and a huge bulk of the tumor was identified with a pedicle originating from the cervical esophagus. The aortopulmonary ligament was dissected before mobilizing the esophagus, in order to obtain satisfactory exposure. The left recurrent laryngeal nerve was not observed around the aortic arch. After mobilization of the esophagus, the left diaphragm was opened and the stomach was mobilized. The gastric tube was formed using a linear stapler along the greater curvature with a diameter of 3 cm-4 cm. Through left cervical incision, a hand-sewn esophagogastric anastomosis was performed.

Figure 1. Upper gastrointestinal barium swallow
A: the tumor occupies the entire length of the esophagus (preoperative); B: normal anastomosis and intrathoracic mucosa of stomach at two-months follow-up

![Figure 1](image1.jpg)

Figure 2. CT scan shows the giant thoracic esophageal tumor with RAA. No enlarged lymph nodes are seen.

Macrosopically, the tumor was 26 cm × 7 cm × 7 cm in size (see Figure 3). The resected tumor manifested as a lobulated polyloid lesion with a bulk base and submucosal thickening of the esophageal wall. Histologically, the tumor consisted of two components: the squamous cell carcinomatous element and spindle cell sarcomatous element. The tumor infiltrated into muscularis propria and only one of the 23 resected lymph nodes was positive for tumor metastasis. The tumor was then staged as pT2N1M0 (IIB). Immunohistochemistry disclosed that the carcinomatous component was positive for P63, PCK, CK5/6 and the sarcomatous component was positive for SMA. Both components were negative for desmin, S-100, CD117, CD34 and CAM52.

The patient had total parenteral nutritional support for five days and then resumed oral intake. He was discharged eventfully on the 13th postoperative day. No severe hoarseness was observed. Barium esophagography at 2 months after operation showed a smooth lumen at the anastomosis site without stricture (see Figure 1B). There was no evidence of recurrence at 1 year follow-up.

Figure 3. Macroscopic appearance of ECS, measuring 26 cm × 7 cm × 7 cm in size

3. DISCUSSION

ECS is a rare esophageal malignancy, thus there are many aspects that remain unelucidated. Currently, there is no firm conclusion about the histological origin of carcinosarcoma. There are three main hypotheses: collision concept, metaplastic concept and reaction concept,[2] but none of them could clarify all reported cases. In our case, the expression of cytokeratin and SMA were mutually exclusive in the carcinomatous or sarcomatous elements, which may support the collision concept. Immunohistochemistry is the most accurate method for diagnosing carcinosarcoma.[7] The two different components of carcinosarcoma have different specific markers. The cytokeratin and epithelial membrane antigen are the most common markers expressed in the carcinomatous element, while vimentin and SMA are expressed in the sarcomatous element.[8,9] Histological features in the carcinomatous component have variously been described as squamous cell carcinoma or adenocarcinoma. In our case, the carcinomatous component was positive for CK5/6 and the sarcomatous component was positive for SMA, which supported the diagnosis.

In our case, the carcinosarcoma involved the entire thoracic esophagus, thus the patient had severe dysphagia and extreme weakness before surgery. Furthermore, the patient presented with a congenital RAA, thus we chose a left thoracotomy approach and cervical anastomosis for better safety and visibility. To the best of our knowledge, this is the first case in the literature that describes the coexistence of an ECS of this formidable size along with a RAA.

This patient had smoked approximately thirty cigarettes everyday for the past fifteen years. The close link between smoking and esophageal cancers is well-established and doc-
The authors declare no conflicts of interest.

REFERENCES


