

Imaging Findings of Esophageal Sarcomatoid Carcinoma: Case Report

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ABSTRACT

OBJECTIVE: To investigate the imaging findings and pathological features, differential diagnosis of esophageal sarcomatoid carcinoma.

METHODS: Three cases of esophageal sarcomatoid carcinoma (ESC) proved by histology was retrospectively analyzed, and related literature were reviewed.

RESULTS: Two cases of barium in the digestive tract showed an irregular filling defect in the thoracic esophagus. The edges are clear, the surface of the tumor is irregular, and the niche is visible. The long axis of the tumor is the same as the esophagus, esophageal expansion, and the mucosa of the lesion area is destroyed. CT scan of 1 case showed a ellipse-like pedicle solid mass in the lower esophagus, the pedicle, and back wall is linked together, the back wall is slight thick, and esophageal eccentric stenosis, homogeneous density, lesions showed moderate enhancement, and the peripheral enhancement was relatively obvious in the central region, and the pedicle was not clear with the right posterior wall of esophagus. Intraoperative observation showed that the tumor was solid, rubbery, and the pedicle was closely related to the right posterior wall of esophagus. All 3 cases were pathologically diagnosed as esophageal sarcomatoid carcinoma.

CONCLUSION: ESC is uncommon, the imaging features have some characteristics, accurate diagnosis depends on the pathological examination.

KEYWORDS: Esophagus, sarcomatoid carcinoma, computed tomography, tomography

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Introduction

Sarcomatoid carcinoma can attack the whole body and commonly occur in the lungs, livers, kidneys, bladders, and skins, but primary ESC is very rare and is often misdiagnosed as esophageal carcinoma or other lesions.¹ ESC is clinically featured by low metastasis rate, high excision rate, and good prognosis. Imaging findings can facilitate the diagnosis of ESC and offer some valuable information for clinical surgical therapy and postoperative follow-up. In this study, 3 cases of ESC were reported, and their clinical, imaging, and pathological data were analyzed. Finally, the imaging manifestations of ESC were summarized by combining with literature, and aiming to improve the diagnostic level of ESC and aiming to improve the diagnostic level of ESC.

Case Presentation

All procedures were performed in accordance with the guidelines and regulations of the First Affiliated Hospital of Dali

University. All protocols were approved by the Department of Medical Imaging, the First Affiliated Hospital of Dali University. The patient provided written consent on standard forms and also provided informed consent for publication of this report.

In case 1, a 58-year-old man sought medical help after suffering progressive dysphagia accompanied with retrosternal pains for 1 month. He had no nausea, vomiting, dizziness, or fever. The upper digestive tract endoscopy showed that a swelling mass bulged on the esophageal posterior wall at 30 cm from the fore-tooth. The mass had a short pedicle and was $30 \times 15 \times 25 \text{ mm}^3$ in size. It was partially rotten and ulcerative on the surface, and was brittle and bled upon touching. Laboratory blood and urine routine examinations showed no abnormality. The digestive tract tumor indices were all within normal ranges, including carcinoembryonic antigen (CEA), alpha-fetoprotein (AFP), and cancer antigen (CA)125/19-9. Physical examination showed body temperature was 36.4°C ;

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heart rate, blood pressure, and pulse were all normal, and no superficial enlarged lymph node was found in the whole body. The digestive tract barium meal imaging (Figure 1) showed some irregular filling defects with clear edges in the middle esophageal segment, and the tumor surface was irregular with niche signs. The long-axis of the tumor was consistent with the esophageal long-axis, and the esophagus at the lesion was expanded in a spindle shape, but the mucosa of the lesion was destroyed and disappeared.

In case 2, a 68-year-old man felt slight retrosternal obstruction upon swallowing for over half a year, especially when he ate hard foods. He had no hematemesis or tarry stool. The upper digestive tract endoscopy showed a mass at the esophageal lower-segment soft tissues and luminal stenosis, and the mass was brittle, stiff, and dark red on surface. Laboratory blood and urine routine examinations showed no abnormality. The digestive tract tumor indices were all within normal ranges, including CEA, AFP, CA125, and CA19-9. Physical examination revealed body temperature was 36.6°C, and heart rate, blood pressure, and pulse were all normal. The digestive tract barium meal imaging displayed (Figure 2) some filling defects at the lower segment of the esophagus, and the barium agent was coated nonuniformly, showing “ring signs,” but it stopped when the barium agent passed, without evident obstruction, and the lesion expanded to the upper segment.

In case 3, a 63-year-old man suffered dysphagia for 3 months and was admitted after 3 days of aphagia accompanied with vomiting. He showed dysphagia without evident inducement 1 month before admission, and the symptoms were more severe when he ate hard foods, but were alleviated when he ate fluid foods. He had no nausea, vomiting, abdominal pains, diarrhea or tarry stool. He lost 2 Kg of weight after the occurrence of the disease. Physical examination showed body temperature was 37.1°C, and heart rate, blood pressure, and pulse were all normal. No superficial enlarged lymph node was found in the whole body, and no special change occurred in the heart, lungs, or abdomen (Figures 3-7).

The 3 cases were all pathologically diagnosed as sarcomatoid carcinomas, and their tumor tissues consisted of epithelioid cancer cells and sarcomatoid spindle cells (Figure 8). The cells were heteromorphic significantly and involved the mucosae and submucosae.

Discussion

Esophageal sarcomatoid carcinoma (ESC) is a special type of esophageal epithelia tumors and is defined by WHO2002 *Pathology and Genetics of Tumours of the Digestive System* as squamous cell carcinoma with unequal quantities of sarcomatoid spindle cells, naming it spindle cell carcinoma and marking it 8074/3 in ICD-O.² As is well-known, its synonyms include spindle cell carcinoma, sarcomatoid carcinoma, carcinosarcoma, polypoid cancer, and pseudo sarcomatoid squamous carcinoma. Though the names of such lesions are different among different systems of WHO, a relatively explicit definition has been

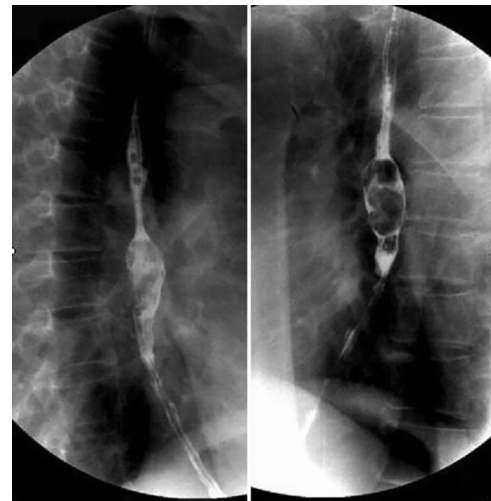


Figure 1. Male, 58 years old, esophageal angiography showed some irregular filling defects with clear edges in the middle esophageal segment, and the tumor surface was irregular with niche signs.

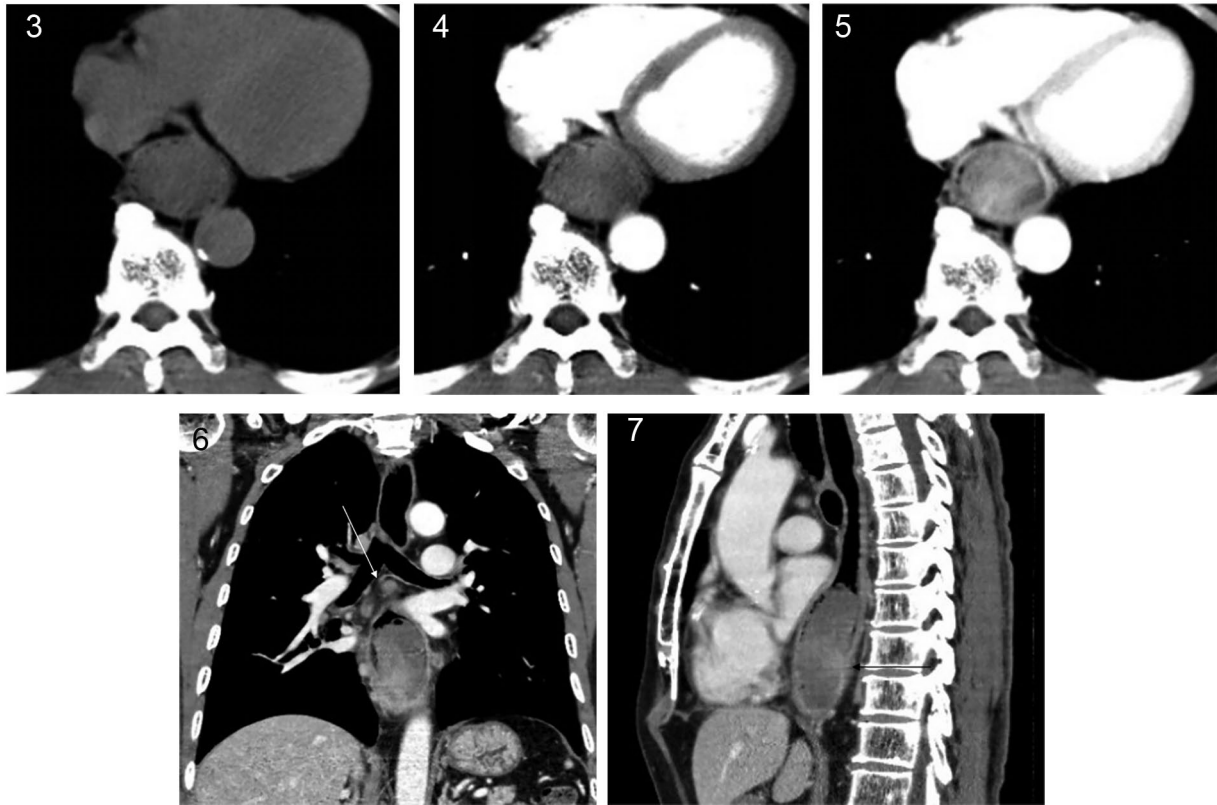


Figure 2. Male, 68 years old; filling defects at the lower esophageal segment, and the barium agent was coated nonuniformly, and the lesion expanded to the upper segment.

formed in recent years.³ In brief, if clear heterologous sarcoma components appear in sarcomatoid interstitia (eg, fibrosarcoma, rhabdomyosarcoma, osteosarcoma), it is carcinosarcoma; otherwise, it is sarcomatoid carcinoma. The 3 cases in this study were all dominated by sarcomatoid spindle cell components, without clear heterologous sarcomal components, and hence were classified as sarcomatoid carcinomas.

So far, the views about the original tissues of ESC are still controversial and include multipotential stem cells, metaplasia, embryo residues, and collision, but there are two mainstream views.⁴ In our study, 2 cases of immunohistochemistry showed that some sarcomatoid components could express markers of epithelial components, so we believed that sarcomatoid components might be derived from metaplasia of carcinoma components.

As reported,⁵ ESCs account for 0.36% to 2.4% of esophageal malignant tumors and mainly attack middle-aged and old



Figures 3-7. Male, 63years old. CT plain scan (Figure 3) showed a low-density soft tissue mass growing intracavitarily, with unclear boundary and uneven edges. Uniform persistent reinforcement occurred at the arterial phase (Figure 4) and venous phase (Figure 5). The reinforcement was more obvious in the center than in the periphery. The boundary between the tumor and the right posterior esophageal walls was unclear, and local Hampton lines were thickened and interrupted (black arrow). Coronal (Figure 6) and sagittal (Figure 7) reconstruction displayed a short pedicle at the right posterior was connected with the esophageal walls (black arrow), and multiple slightly-enlarged lymph nodes (white arrow) were found under the mediastinal jugum.

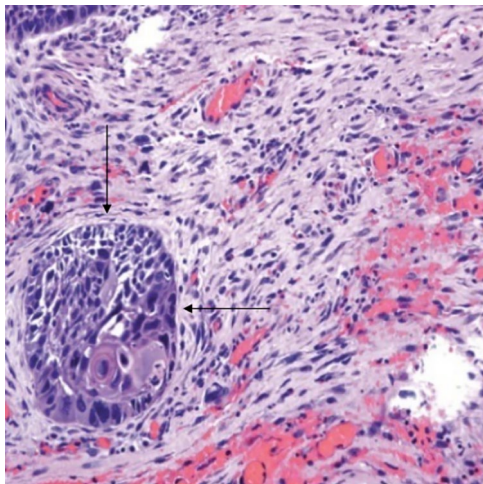


Figure 8. The mass under microscopy was composed of epithelioid cancer cells (black arrow) and sarcomatoid spindle cells ($\times 100$).

men, especially those with smoking or liquor addiction, and the age of pathogenic peak is about 60years old. ESCs most frequently occur at the middle segment of the esophagus (about 60%), followed by the lower segment (28%), the upper segment, and the cervical part. The average age of the 3 patients was 63years old, 2 patients were located in the middle part of

the esophagus, and 1 patient was located in the lower part, which was consistent with the literature report. The clinical manifestations are mostly progressive dysphagia or obstruction accompanied with retrosternal pains, and the symptoms are not significantly different from other esophageal malignant tumors, but since ESCs are mostly polypoid-like and grow intracavitarily, the symptom of obstruction occurs earlier, so the course of diseases is shorter.

Esophageal barium meal is the first choice for imaging examination. In this group, barium meal examination was performed for 2 patients, and the lesion size was not proportional to the degree of obstruction. In spite of its large size, the tumor presented a slow shunt or circumferential flow when barium passed through the lesion area, with no obvious obstruction. In addition, the adjacent wall of the lesion was more soft, and the mucosa was flat or there was small erosion between the lesions, presenting a change of "smear sign," which was also different from the annular wall growth pattern of esophageal cancer. The above signs were helpful to suggest the diagnosis.

Multislice CT (MSCT) with strong post-processing ability after 3D reconstruction can display the size, shape, and position of masses and the thickening degree of adjacent esophageal walls, and can help to judge whether the masses

invade outward and whether lymph node metastasis occurs.⁶⁻⁸ One patient in this group underwent CT examination. Plain CT scan showed the soft tissue masses were connected with the esophageal walls at the narrow base, and the lumina were eccentric crescent-like or crevice-like, while the adjacent walls were slightly thickened. The masses had clear boundaries and regular shapes and were slightly lobulated, with uniform density, but no necrotic cystic degeneration was observed. The above manifestations were significantly different from esophageal cancer, which was characterized by the growth of the annular wall and the thickening of the wall, accompanied by different degrees of dysphagia, and when the mass was large, the hemorrhage and necrosis were generally obvious. In addition, the tumor in this contrast-enhanced scan showed mild to moderate heterogeneous enhancement, with relatively obvious central enhancement compared with peripheral enhancement, which may be caused by sarcomatoid components and more abundant blood supply inside the tumor, while most esophageal cancers were significantly heterogeneous enhancement, with different intensification modes and degrees from ESC.

ESCs should be differentially diagnosed from the following diseases: (1) Fungating esophageal carcinoma: differentiation of the two is difficult on imaging and requires confirmed diagnosis by endoscopic pathological biopsy. We should carefully observe the imaging manifestations, and if massive pedicles can be observed through rotation at different directions, this will be valuable for the diagnosis of ESCs. (2) Esophageal leiomyoma and leiomyosarcoma: the leiomyoma is a benign tumor and intersects at an obtuse angle with the esophageal walls. It grows submucosally and shows clear “ring signs” or “mucosal bridge signs.” CT can clearly display the size, extent, and growing way of leiomyoma. When mucous layer completeness is observed

via enhanced scan, the possibility of ESC should be considered, and if intratumor bleeding or necrosis is obvious, leiomyosarcoma should be considered. (3) Lymphoma: lesions at esophageal walls usually are not obviously stiff and usually will induce the narrowing of esophageal lumina, with local expansion, and the imaging manifestation is intracavitary irregular filling defects.

In a word, esophageal sarcoma-like carcinoma is one of the rare malignant tumors of the esophagus, which has special pathological characteristics and has a good prognosis. Therefore, preoperative diagnosis is of great significance to improve the prognosis of patients. Upper gastrointestinal barium meal angiography is simple and easy to perform, which can be used as the preferred examination method. CT examination has certain characteristics, which can show the morphology, location, and thickening degree of the adjacent esophageal wall of the lesion, which is helpful for preoperative staging.

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