


CASE REPORT

A huge Ewing's sarcoma of the rib presenting with superior vena cava syndrome and dysphagia

Dan Lin¹ | Xiaomu Zhu² | Long Tian¹ | Changlong Qin¹ | Jingsi Dong¹  | Qinghua Zhou¹

¹Lung Cancer Center, West China Hospital of Sichuan University, Chengdu, China

²Department of Pharmacy, West China Hospital of Sichuan University, Chengdu, China

Correspondence

Jingsi Dong and Qinghua Zhou
Lung Cancer Center, West China Hospital,
Sichuan University, No.37 Guoxue Alley,
Chengdu 610041, China.
Email: dongjingsi@wchscu.cn and zhouqh135@163.com

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Abstract

A 24-year-old male patient was admitted to our center complaining of dizziness (superior vena cava syndrome [SVCS]), dysphagia and pain in the right chest wall. At the initial diagnosis, the patient had been found to have an irregular shaped 35 × 30 × 27 cm mass in the right side of his chest. On November 12, 2019, this patient received surgery in our center. The right sixth rib and the tumor were completely removed (R0), while preserving all the lung tissue and other organs in the chest. The patient recovered well after surgery, and his right lung was fully expanded.

KEYWORDS

dysphagia, Ewing's sarcoma of the rib, superior vena cava syndrome, surgery

INTRODUCTION

Tumors of the Ewing's sarcoma (ES) family represent a spectrum of malignant tumors of bone or soft-tissue origin, histologically composed of small, round cells which include undifferentiated typical Ewing's sarcoma, poorly differentiated atypical Ewing's sarcoma, and differentiated peripheral primitive neuroectodermal tumors.¹ Ewing's sarcoma (ES) is a rare malignant primitive neuroectodermal tumor, primarily affecting the long bones in young children and adolescents, accounting for 6%–16% of chest wall malignancies.² The rarity of ES of rib has resulted in limited published reports with other primary sites of the chest wall.³ Surgery remains an important modality of treatment for ES of rib in multidisciplinary therapy including induction chemotherapy and radiotherapy.⁴

CASE REPORT

A 24-year-old male patient was admitted to our center complaining of dizziness (superior vena cava syndrome

[SVCS]), dysphagia and pain in the right chest wall. At the initial diagnosis, the patient was found to have an irregular shaped 35 × 30 × 27 cm mass in the right side of his chest. The mass invaded the sixth rib (Figure 1a,b) on the right and compressed the right lung (Figures 1c and FIGURE 3), heart, superior vena cava, inferior vena cava, diaphragm (Figures 1c,d and 2a–d) and esophagus (Figure 1e–h). This was confirmed as Ewing's sarcoma on puncture pathology. The patient received four cycles of chemotherapy in Beijing from July 2019 to October 2019, but the tumor did not shrink. After seeking medical help in Beijing, Shanghai and Guangzhou, he eventually consulted our center.

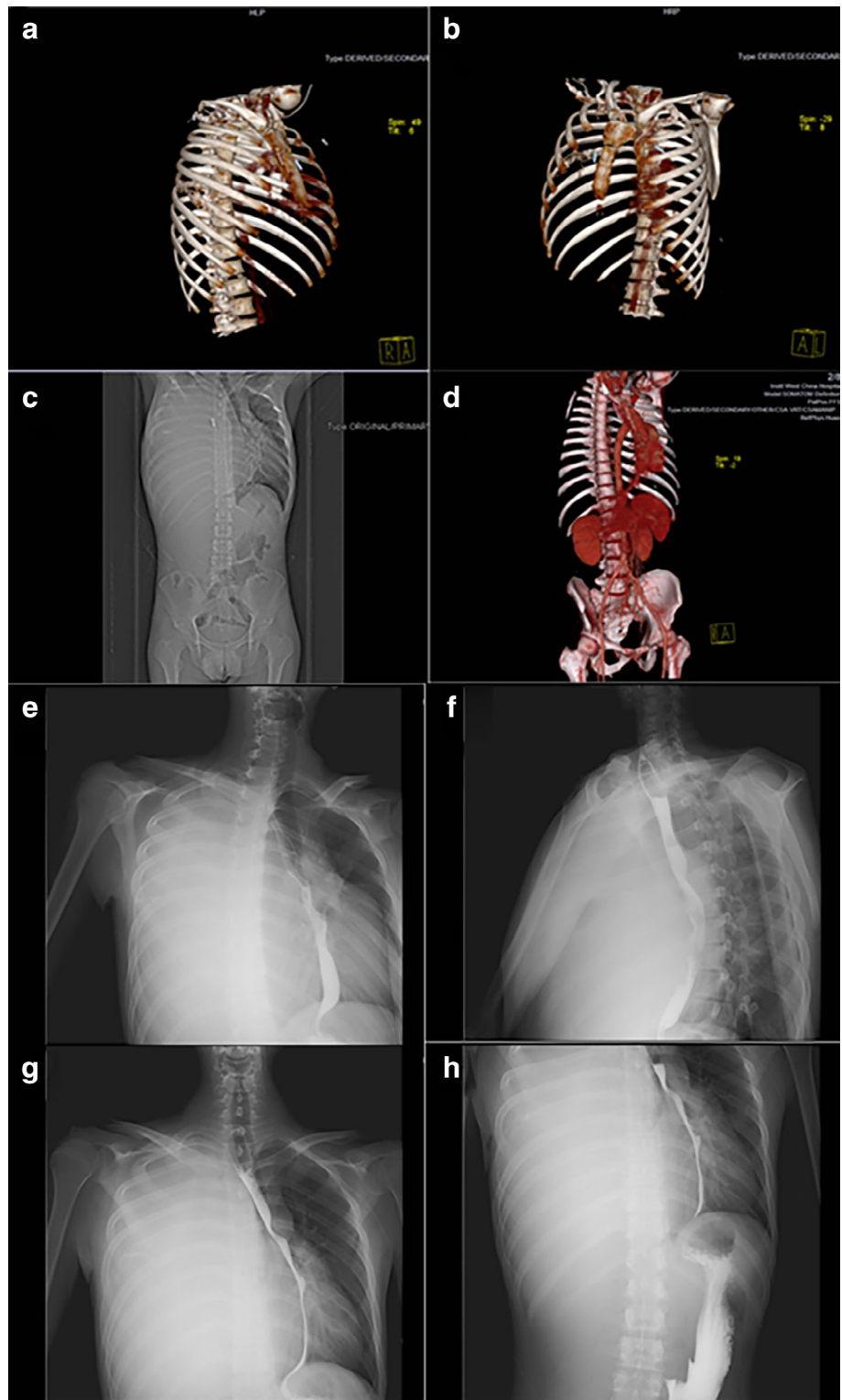
On November 12, 2019, the patient received surgery in our center. The right sixth rib and the tumor were completely removed (R0), while preserving all the lung tissue and other organs in the chest (Figure 2e,f). During the operation, we could see that the tumor invaded the middle and anterior segments of the right sixth rib. The tumor compressed the right lung, heart, superior vena cava and inferior vena cava, the mediastinum was significantly shifted to the left, and the right lung was completely atelectatic. The patient recovered well after surgery, with full expansion of his right lung (Figure FIGURE 3a–c).

Dan Lin and Xiaomu Zhu contributed equally.

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FIGURE 1 (a, b) Rib destruction. (c, d) The heart, superior vena cava, inferior vena cava were compressed. (e–h) The tumor compressed the esophagus



The tumor originated from the right sixth rib with a complete membrane. The tumor was excised, and it weighed 11.3 kg. Postoperative pathology: right thoracic round cell tumor; immunohistochemistry: CD99 (partial +), BCOR (–), TLE-1 (–), EMA (–), CgA (–), desmin (–), myoD1

(–), TdT (–), FLI1 (–), INI1 (+). FISH detection: *EWSR1* gene unbalanced translocation (+), *SS18* gene translocation (–), *DICER1* gene 22–24 exon mutation (–) (Figure 4a–c). The tumor was eventually diagnosed as Ewing's sarcoma of the rib.

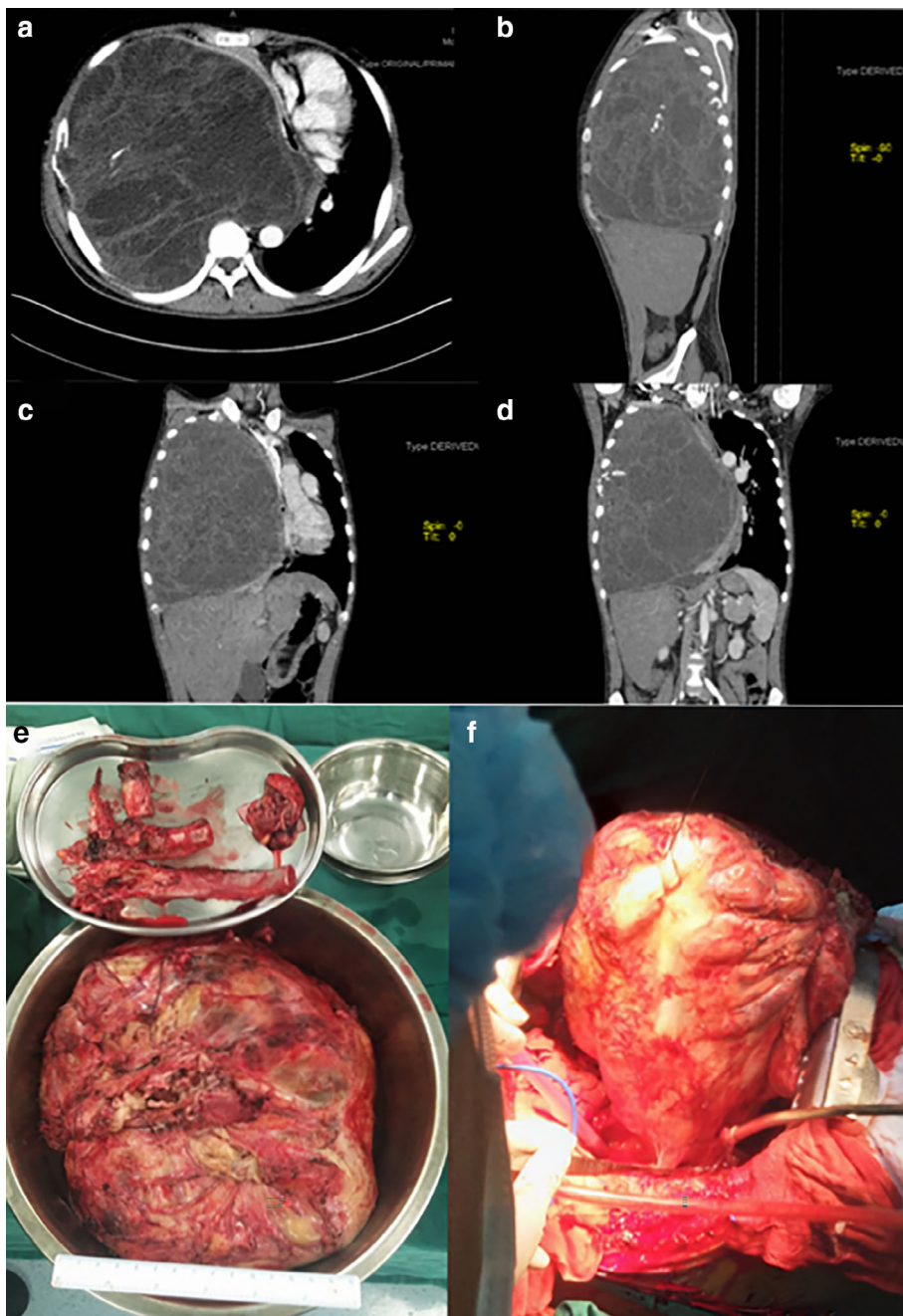


FIGURE 2 (a–d) The mass invaded the sixth rib on the right and compressed the right lung, heart, superior vena cava, inferior vena cava and diaphragm. (e–f) During the operation the patient was found to have an irregular shaped $35 \times 30 \times 27$ cm mass in the right chest. The tumor capsule was intact and had not invaded the superior vena cava, inferior vena cava or the heart

The patient was followed-up and remains well at present and on January 5, 2022, the patient's CT showed that the right sixth rib was absent and there was a postoperative scar with pleural thickening on the right lung (Figure [FIGURE 3d–f](#)).

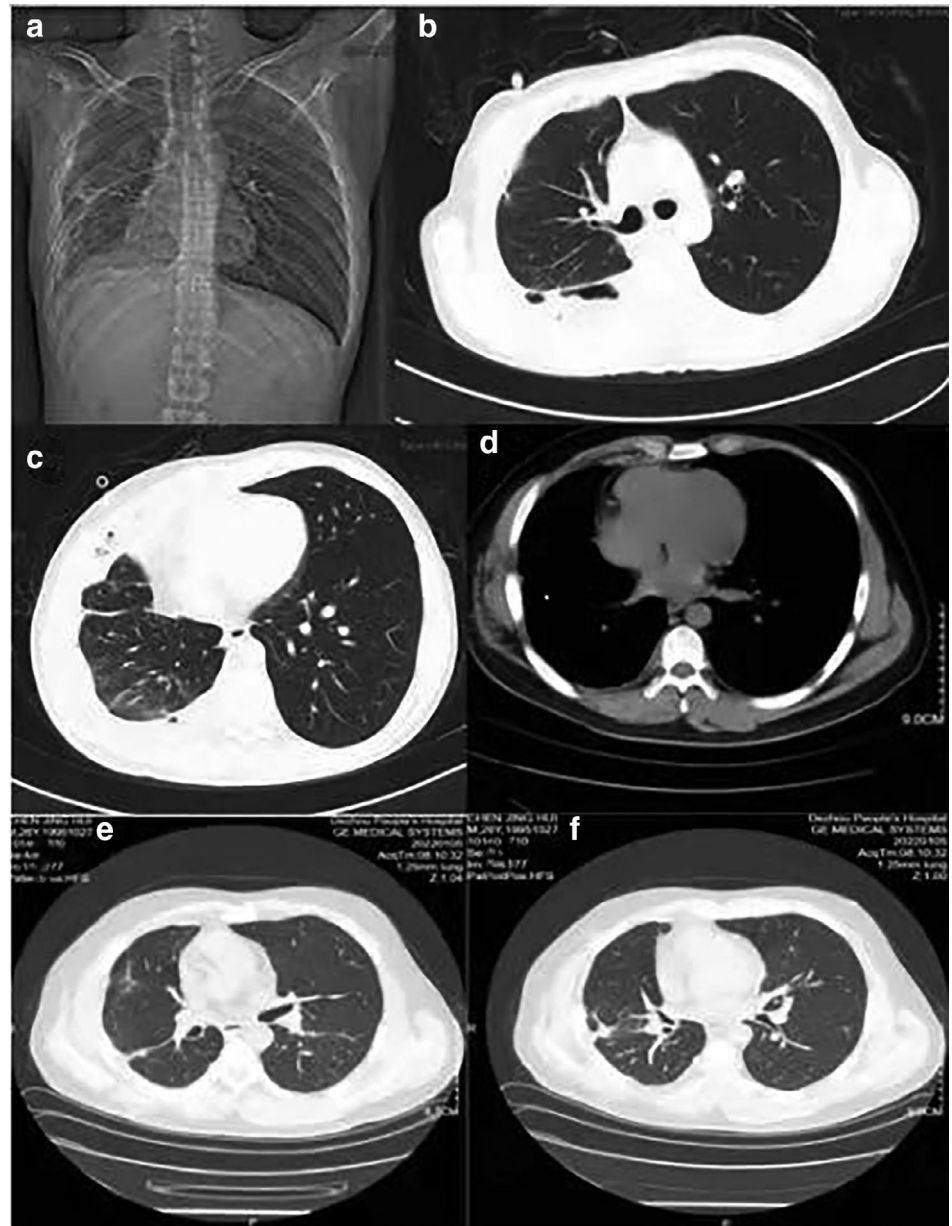
DISCUSSION

Ewing's sarcoma occurs in the metaphysis of long bones in approximately 80% of cases, with a peak incidence between the first and second decades.⁵ About 10% of cases of Ewing's sarcoma arise from the rib as first reported by Franseen in

1946.⁶ The common symptoms of Ewing's sarcoma of the rib are chest pain, fever and malaise, and the most important clinical findings are a palpable mass and pleural effusion.⁷ Devika et al.⁸ reported a male patient who developed facial puffiness and tachypnea which suggested superior vena cava obstruction. However, Ewing's sarcoma of rib presenting with superior vena cava syndrome (dizziness) and dysphagia has not yet been reported.

In most cases, Ewing's sarcoma depends on a combination of morphology, immunohistochemistry, and molecular findings to realize accurate classification. The most common chromosomal translocation is between the *EWSR1* (22q12) and *FLI1* (11q24) genes. CD99 has also been reported to be

FIGURE 3 (a–c) The patient recovered well after surgery, and his right lung was fully expanded. (d–f) The patient was followed-up and remains well. The right sixth rib was absent and there was a postoperative scar with pleural thickening on the right lung



highly sensitive but lowly specific.⁷ In the case reported here, this turned out to be a Ewing's tumor of the rib by puncture pathology and postoperative pathology.

The multimodal therapy of ES has significantly improved patient prognosis. Doxorubicin-based neoadjuvant chemotherapy has been proven to reduce tumor volume, decrease the use of radiotherapy and increase the rate of negative margin resection (R0).^{9,10} Our patient was also treated with neoadjuvant chemotherapy, but the tumor did not shrink. The literature suggests that surgery is the best option for Ewing's sarcoma in the chest wall to achieve definitive local control and should be practiced by specialists experienced in complex thoracic oncology.¹¹ In this case, we were prepared to resect the superior vena cava and the inferior vena cava and reconstruct the artificial vessels before the operation, but the tumor capsule was intact and the tumor did not invade the above vessels or the heart, and

eventually we only removed the rib and tumor. In terms of the range of ribs that should to be resected, it is recommended to remove the involved rib and partially excise the adjacent ribs to achieve a tumor-free margin.^{12,13} Local radiotherapy is necessary in patients with pleural effusion and poor response to chemotherapy, but is not beneficial in patients who have undergone R0 resection and achieved a complete response to chemotherapy.^{14,15} The right sixth rib only was removed in this patient, protecting the function of the chest wall, and radiotherapy was not considered because of the R0 resection. With regard to prognosis, a poor response to chemotherapy and the infiltration of adjacent lung parenchyma are important adverse prognostic factors.¹³ In our case, although the patient had a poor response to chemotherapy, there was no evidence that the surrounding tissue had been invaded, and the patient has been followed-up and remains well at present.

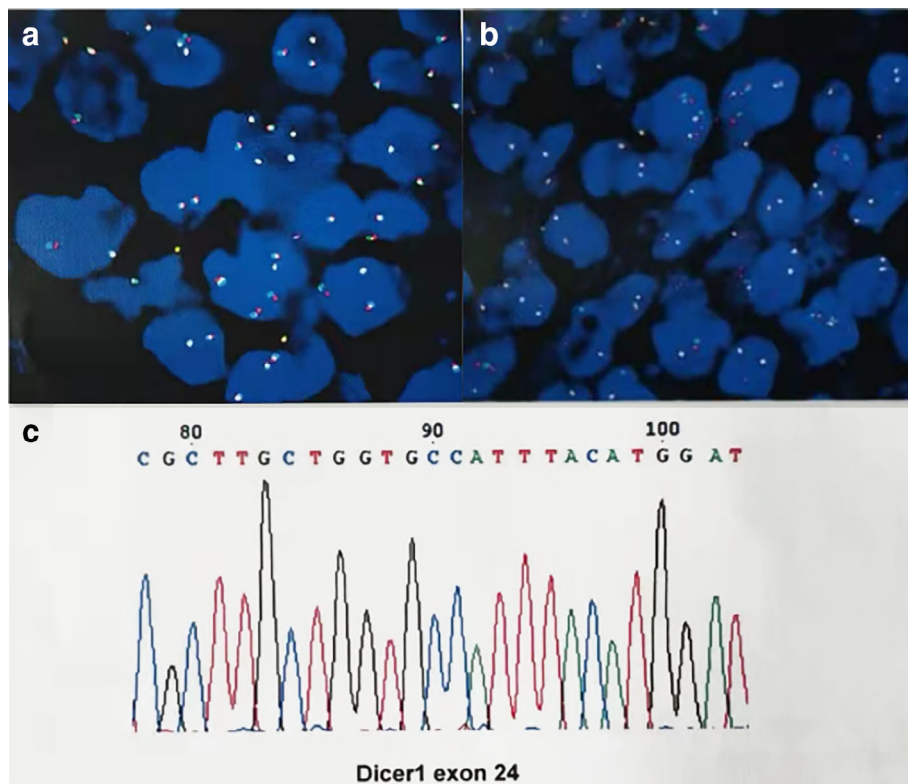


FIGURE 4 (a–c) FISH detection: *SS18* gene translocation (–), *EWSR1* gene unbalanced translocation (+), *DICER1* gene 22–24 exon mutation (–)

CONFLICT OF INTEREST

The remaining authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

ORCID

Jingsi Dong  <https://orcid.org/0000-0002-1922-5862>

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