


CASE REPORT

Epithelioid hemangioendothelioma of the sternumWenpeng Song^{1,2†}, Zeng Zhen^{1†}, Lin Li³, Jing Ye², Sicheng Zhou², Qiyu Wu², Liu Xu², Huilin Li² & Feng Lin¹ ¹ Department of Thoracic Surgery, West China Hospital, Sichuan University, Chengdu, China² West China School of Medicine, Sichuan University, Chengdu, China³ Pathology Department, West China Hospital, Sichuan University, Chengdu, China**Keywords**

Epithelioid hemangioendothelioma; follow-up; sternum; surgical resection.

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Thoracic Cancer **11** (2020) 1741–1745**Introduction**

Epithelioid hemangioendothelioma (EHE) is a rare sarcoma composed of dendritic and endothelial cells. The tumor is clinically and histologically intermediate between benign hemangioma and angiosarcoma and accounts for less than 1% of all vascular tumors. It is most commonly found in the liver, bone, and skin. EHE in the chest is extremely rare. This condition occurring in the sternum has not been reported until now. Given that EHE is aggressive, surgical resection with adjuvant chemotherapy and radiotherapy is recommended to prevent recurrence or metastasis.

Case report

A 43-year-old man presented to our hospital with a chest mass accompanied by chest pain for over five months. The patient complained of slight haphalgnesia and apparent tenderness radiating into the neck and shoulders, which was relieved by administering fenbid. The patient denied any

Abstract

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor composed of dendritic and endothelial cells, and accounts for less than 1% of all vascular tumors. The tumor may arise from any location in the body, but it has a predilection for veins of the extremities, the liver, bone, and skin. Herein, we report a case of a 43-year-old man who presented with a chest mass accompanied by chest pain. Computed tomography (CT) scan of the chest showed a 3 cm × 2 cm sternal mass in the chest wall. We performed tumor resection and reconstruction of the chest wall. EHE was confirmed via postoperative pathology. The patient underwent postoperative adjuvant radiotherapy, and no evidence of tumor recurrence was discovered during the one-year follow up.

history of trauma, and had no relevant medical history. On physical examination, a tumor 3 cm in diameter was found in the anterior chest wall. The preoperative thoracic computed tomography (CT) scan revealed a 3 cm × 2 cm mass in the sternum manubrium (Fig 1a). In addition, three-dimensional (3D) reconstruction of the chest CT confirmed the presence of a sternal tumor (Fig 1b). A percutaneous thin-needle aspiration biopsy was performed before the operation, and the primary diagnosis was mesenchymal tumor. Considering the particularity of sternal site and function, resection of the sternal tumor and a 3D-printed titanium alloy graft implantation was planned which was approved by the Institutional Review Board of West China Hospital (No. 2018–45). Informed consent of the patient was obtained before surgery and the design of the 3D-printed graft was performed using 3D reconstruction of the preoperative chest CT (Fig 2a). Consequently, the patient underwent an enlarged excision of the manubrium sternum with the surrounding muscle tissues and reconstruction of the chest wall with 3D-printed titanium alloy sternal prosthesis under general anesthesia. The 3D-

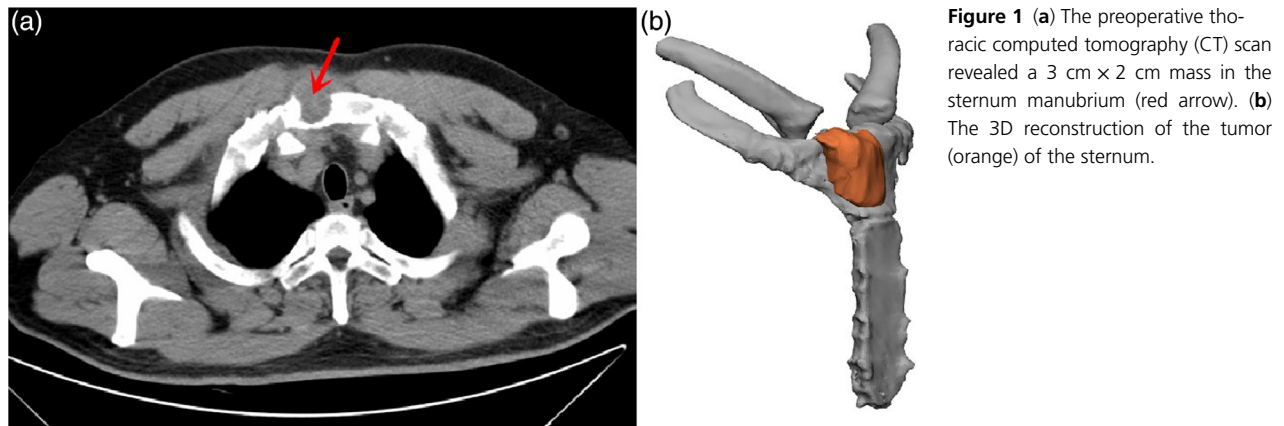


Figure 1 (a) The preoperative thoracic computed tomography (CT) scan revealed a 3 cm × 2 cm mass in the sternum manubrium (red arrow). (b) The 3D reconstruction of the tumor (orange) of the sternum.

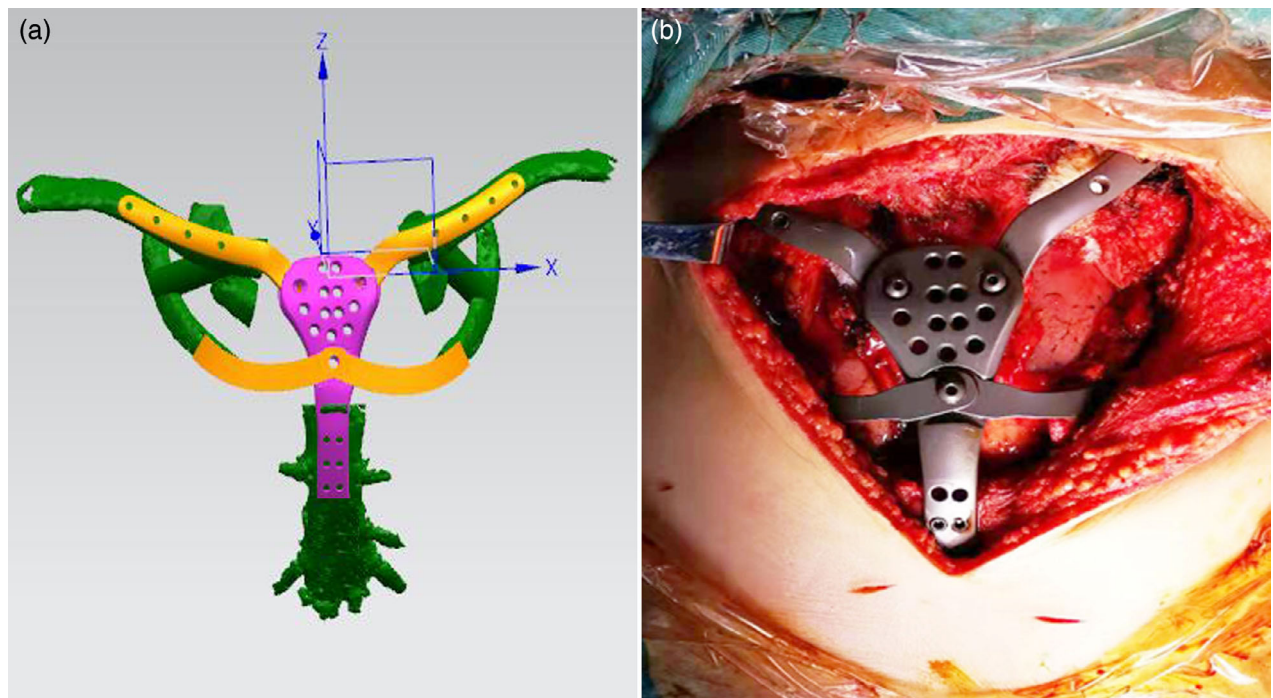


Figure 2 (a) The 3D printed design drawing before operation. (b) The 3D-printed titanium alloy sternal prosthesis was implanted during the operation.

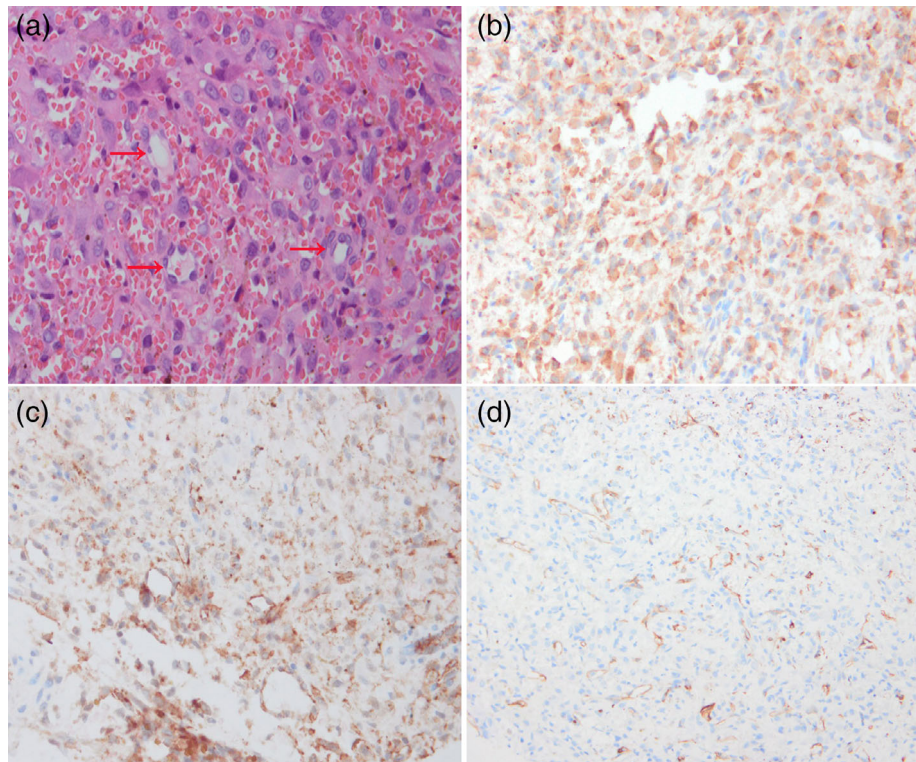
printed sternal prosthesis consisted of two pairs of transverse limbs and a body. The limbs were fixed in the clavicle and the second rib, respectively, and the body in the sternum with screws (Fig 2b). Postoperative pathological examination confirmed the diagnosis of EHE. Hematoxylin and eosin staining showed some dendritic and endothelial cells formed various size vessels with red blood cells contained (arrows). There was also a dense inflammatory infiltration in some areas. (Fig 3a). Immunohistochemical staining was positive for CAMTA1, CD31, and CD34 (Fig 3b–d). Postoperative chest radiographs showed that

the metal graft was in a normal position (Fig 4). After a successful postoperative recovery, he underwent radiotherapy, and no evidence of tumor recurrence was discovered during the one-year follow up.

Discussion

EHE, a rare low-moderate malignant hemangioma, was first described by Weiss in 1982.¹ Clinically and histologically, EHE is considered an intermediate malignancy between angiosarcoma and hemangioma. According to

Figure 3 (a) Hematoxylin and eosin staining showed some dendritic and endothelial cells formed various size of vessels with red blood cells contained (arrows). There was also a dense inflammatory infiltration in some areas. (b–d) Immunohistochemical staining was positive for CAMTA, CD31 and CD34 proteins.



reports in the literature, the incidence is less than one in a million,² and it can occur at any age, especially in the 20s. Clinically, EHE mostly occurs in soft tissues, the liver, bone, and other organs.³ Review of the literature revealed rare involvement of the chest wall. To the best of our knowledge, our case is the first report of sternal EHE. The symptoms of EHE usually depend on the site of the disease, such as abdominal pain (liver) and dry cough (lung). The clinical presentation of sternal EHE is nonspecific and varies depending on the size of the tumor, which is similar to other common chest wall tumors. Patients may present with swelling and pain in the chest wall.

Radiological examination has great clinical value for the early discovery of ETE. The CT image of osseous EHE presents a multifocal lytic abnormality with osseous expansion, sclerotic margins, and a lack of periostitis. However, the diagnosis of EHE needs to be confirmed by pathology and immunohistochemistry. Pathologically, the tumor is composed of dendritic and endothelial cells with positive nuclear staining, and positive immunohistochemistry for vascular markers (CD31, CD34, and vimentin) can show better specificity. In addition, the WWTR1-CAMTA1 characteristic fusion gene is regarded as an auxiliary diagnosis. The main differential diagnosis of EHE includes metastatic signet-ring cell adenocarcinoma and epithelioid angiosarcoma.⁴

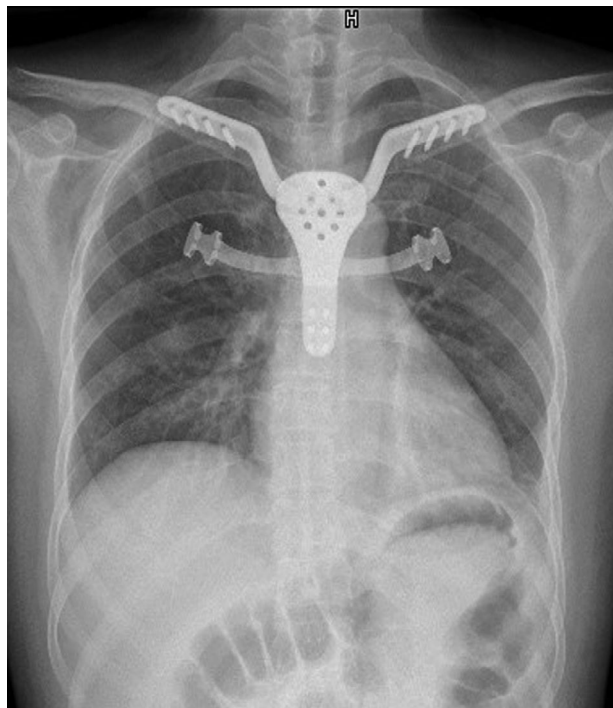


Figure 4 Postoperative chest radiographs showed that the metal graft was in a normal position.

Table 1 Reported cases of epithelioid hemangioendothelioma (EHE) in the chest

N	Author	Age	Sex	Location	Size (cm)	Symptoms	Treatment	Follow-up
1	Kumar et al. ⁵	63	M	Lung	15 × 15 × 4	Chest pain, cough	Surgical resection	No recurrence
2	Agaimy et al. ⁶	62	M	Mediastinum	10 × 9 × 7	Dyspnea, cough	Surgical resection	No recurrence
3	Isowa et al. ⁷	41	M	Mediastinum	3.4 × 2.7	No	Surgical resection	No recurrence
4	Val-Bernal et al. ⁸	69	F	Heart	4 × 3 × 2	Dyspnea, orthopnea	Surgical resection	No recurrence
5	Lisy et al. ⁹	61	M	Heart	3 × 3	Abdominal pain	Surgical resection	No recurrence
6	Safirstein et al. ¹⁰	51	F	Heart	5 × 4 × 4	None	Surgical resection	No recurrence
7	Márquez-Medina et al. ¹¹	85	M	Pleura	15 × 15 × 4	Chest pain, cough	Surgical resection	No recurrence
8	Lee et al. ¹²	31	F	Lung, bone	5 × 3 × 2.5	Dull pain	Chemotherapy	No recurrence
9	Kim et al. ¹³	46	F	Pleura	No stated	chest discomfort, cough	Pleurectomy; Chemotherapy	Died after 23 months
10	Mizota et al. ¹⁴	59	F	Lung	2.5 × 3.5	Cough, dyspnea	Surgical resection; chemotherapy	Recurrence
11	Bansal et al. ¹⁵	51	F	Pleura	No stated	Chest pain, weight loss	Surgical resection; chemotherapy	Died after 14 months
12	Campos et al. ¹⁶	36	F	Mediastinum	5.4	Dyspnea, dry cough	Surgical resection	No recurrence
13	Kumar et al. ¹⁷	33	M	Mediastinum	3.9 × 7.3 × 5.8	Chest and back pain	No	Died
14	Wethasinghe et al. ¹⁸	41	M	Mediastinum	1.2 × 2.5	Chest pain, weight loss	radiotherapy	No recurrence

EHE in the chest is extremely rare. The literature review (Table 1) reveals that the most common sites in the chest are lungs, mediastinum, and pleura.^{5–18} Rare cases occurring in the brachiocephalic vein and the superior vena cava have also been reported.^{19,20} To date, this condition occurring in the sternum has not been reported. Surgical resection is generally accepted as the definitive treatment of choice for tumor of the chest wall. After resection of the sternal tumor, sternal reconstruction is necessary to protect the exposed thoracic viscera, repair the appearance of the chest wall, and maintain arm function. Available materials include titanium mesh or plate, autogenous bone graft, cryopreserved sternal allograft, or homograft flap.^{21,22}

However, 3D-printed titanium alloy grafts have the advantages of individualization, easy fixation, and light tissue rejection. The clinical course of EHE is usually somewhat indolent compared with other sarcomas, and the five-year survival is about 73%. Postoperative adjuvant radiotherapy is usually required to prevent recurrence and metastasis.^{3,23}

In conclusion, EHE in the chest are extremely rare, and there are no cases in the sternum having been described in the literature to date. Preoperative diagnosis of sternal EHE is difficult, since clinical symptoms and radiology features overlap with other tumors arising from the chest wall. Here, we report a very rare case of EHE occurring in the sternum. Due to the lack of the evidence in the current

literature, the therapeutic effect cannot be exactly assessed and close follow-up is necessary.

Disclosure

The authors report that there are no conflicts of interest.

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