**Name of Journal: *World Journal of Clinical Cases***

**Manuscript NO: 47117**

**Manuscript Type: CASE REPORT**

**Clear cell sarcoma of soft tissue in the pleural cavity: A case report**

Chen YT *et al*. CCS of soft tissue

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**Author contributions:** Chen YT, Li H, and Ni CH were the patient’s thoracic surgeons, reviewed the literature, and contributed to manuscript drafting; Yang Z performed the pathologic analyses and interpretation; Chen YT and Ni CH were responsible for revising the manuscript for important intellectual content; All authors issued final approval for the version to be submitted.

**Informed consent statement:** Informed written consent was obtained from the patient for publication of this report and any accompanying images.

**Conflict-of-interest statement:** The authors declare that they have no conflicts of interest.

**CARE Checklist (2016) statement:** The authors have read the CARE Checklist (2013), and the manuscript was prepared and revised according to the CARE Checklist (2016).

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**Manuscript source:** Unsolicited manuscript

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**Telephone:** +86-591-86250214

**Received:** March 14, 2019

**Peer-review started:** March 15, 2019

**First decision:** July 30, 2019

**Revised:** August 20, 2019

**Accepted:** August 25, 2019

**Article in press:**

**Published online:**

**Abstract**

***BACKGROUND***

Clear cell sarcoma (CCS) of soft tissue is a rare malignant soft tissue sarcoma usually located at the distal end of extremities and rarely at the trunk.Herein, we report a case of CCS in the pleural cavity*.*

***CASE SUMMARY***

A 31-year-old male was admitted for an uncertain mass in the left pleural cavity in routine physical examination without any symptoms. Video-assisted thoracoscopic surgery was performed to remove the tumor. The pathological finding displayed a cystic mass of 6.5 cm at the longest diameter, dark red sections, and cysts that could be found locally. Strong expression of soluble protein-100, human melanoma black-45, and vimentin was detected by immunohistochemical staining, which was inclined to the diagnosis of CCS of soft tissue. The patient refused chemotherapy, radiotherapy, and targeted therapy because of the personal financial situation. Follow-up computed tomography scans were done on the 90th and 180th postoperative days, and no obvious sign of recurrence was found until now.

***CONCLUSION***

CCS of soft tissue can be found in the pleural cavity, although with an extremely rare incidence. Radical resection is useful to improve the prognosis.

**Key words:** Sarcoma; Clear cell sarcoma; Clear cell sarcoma of soft tissue; Treatment; Case report

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**Core tip:** Clear cell sarcoma is a rare malignant tumor of soft tissue usually arising from the extremities. Here, we report a case of clear cell sarcoma located on the chest cavity, confirmed by positive staining of human melanoma black-45, and soluble protein-100 markers by immunohistochemistry. Radical resection of the tumor was performed and the short-term outcome was good.

Chen YT, Yang Z, Li H, Ni CH. Clear cell sarcoma of soft tissue in pleural cavity: A case report. *World J Clin Cases* 2019; In press

**INTRODUCTION**

Clear cell sarcoma (CCS) of soft tissue is a rare malignant soft tissue sarcoma that represents about 1% of all sarcomas[1]. The true origination of CCS is still uncertain; it might be one type of synovial sarcoma because SSC usually arises in association with tendons and aponeuroses and has the same bidirectional differentiation ability as synovial sarcoma. However, on account of sharing a similar morphology with malignant melanoma, SSC is more likely derived from neural crest cells, which typically produce melanin and stain positive for human melanoma black-45 (HMB-45) and soluble protein-100 (S-100) markers by immunohistochemistry[2]. CCS is usually located at the distal end of extremities and rarely at the trunk[3]. According to the literature review, to date, there has been no report of CCS in the pleural cavity.

**CASE PRESENTATION**

***Chief complaints***

A 31-year-old male was admitted to our hospital for an uncertain mass in the left pleural cavity in routine physical examination without any symptoms.

***History of past illness***

His past history was unremarkable.

***Personal and family history***

His family history was unremarkable.

***Physical examination upon admission***

His physical examination was normal.

***Laboratory examinations***

Routine blood parameters were within the normal range. Tumor marker measurement results were as follows: carcinoembryonic antigen, 0.64 ng/mL; alpha-fetoprotein, 1.77 ng/mL; and carbohydrate antigen 19-9, 5.80 U/mL. Enhanced thoracic computed tomography (CT) demonstrated a posterior mediastinal mass about 6 cm diameter in the left pleural cavity, near the 9th and 10th thoracic vertebrae beside the thoracic descending aorta, with uneven enhancement (Figure 1). A neurologic tumor was suspected as the first diagnosis.

Video-assisted thoracoscopic surgery was performed to remove the mass. During the operation, the patient was placed in a right lateral decubitus position at 70 degrees with unilateral ventilation under general anesthesia. Three trocars were placed at the 3th, 4th, and 8th intercostal space. Under endoscopy, there was no obvious effusion in the pleural cavity. A spherical mass with broad base was found on the posterior chest wall, just left of the 9th thoracic vertebra, close to the aorta while no obvious adhesion was found between the tumor and aorta or vertebra. Careful dissection was done with electrocautery and harmonic scalpel in turn. A feeding branch from the intercostal artery and a draining vein to the hemiazygos vein were ligated and cut. Finally, the mass was removed under *en bloc* excision with a relatively clear margin. Furthermore, the base plane on local chest wall was managed with electrocoagulation to ensure no residual tumor (Figure 2). The drain tube was placed and the chest wall wound was closed routinely. The chest tube was removed on the 1st postoperative day and the patient was discharged on the 5th postoperative day with no events.

**FINAL DIAGNOSIS**

The pathological finding displayed a cystic mass of 6.5 cm at the longest diameter, dark red sections, and cysts that could be found locally. Strong expression of S-100, HMB-45, and vimentin was detected by immunohistochemical staining (Figure 3), which was inclined to the diagnosis of CCS of soft tissue[2,3].

**TREATMENT**

Video-assisted thoracoscopic surgery was performed to dissect the tumor. After surgery, the patient refused the suggestion of all following treatments such as chemotherapy, radiotherapy, and targeted therapy because of the personal financial situation.

**OUTCOME AND FOLLOW-UP**

Meanwhile, two postoperative follow-up CT scans were done on the 90th postoperative day and the 180th postoperative day, and no obvious sign of recurrence was found until now.

**DISCUSSION**

CCS is a rare malignant tumor of soft tissue with uncertain origination, which seems most likely to be diagnosed in adolescents and middle-aged people. The diagnosis of CCS always seems to be a challenge if there is a lack of pathologic immunohistochemical results. Although CCS has a similar morphologic appearance with malignant melanoma, it is possible for pathologists to distinguish them correctly by the difference of histopathologic and cytogenetic features between them[2]. CCS is usually located in the deep soft tissue or under the deep fascia, in close relation to tendons and aponeuroses at the distal end of extremities such as the foot and knee, rarely at the trunk, with no epidermal involvement[4-9] Differential diagnosis from other sarcomas should be made including epithelioid sarcoma, synovial sarcoma, epithelioid malignant peripheral nerve sheath tumor, alveolar soft part sarcoma, and so on[3].

Radical resection of the tumor might be the most effective way to treat CCS. Before the operation, magnetic resonance imaging of thoracic vertebra is done to exclude possible invasion of the tumor to the vertebra. The intraoperative exploration also ensures that there is no obvious adhesion between the spine and the tumor. If the vertebra or posterior ribs are invaded by tumor, the nearby vertebra and ribs should be removed with the tumor under *en bloc* excision and reconstruction of the chest wall should be done.

Adjuvant radiotherapy or chemotherapy might not be necessary for the poor evidence of therapeutic effect on SCC[10]. However, radiation therapy is recommended if there is possible residual tumor after surgery to achieve a better outcome[11].. Targeted therapy seems to be a good treatment for SSC with some encouraging evidence on malignant melanoma, but there is still a long way to go[12]. Regular follow-up including CT scan or magnetic resonance imaging should be made every 3 mo postoperatively on account of the high risk of early metastases and recurrence of SSC[13].

**CONCLUSION**

CCS of soft tissue can be found in the pleural cavity, although with an extremely rare incidence. Radical resection is useful to improve the prognosis.

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**P-Reviewer:** Kutlubay Z, Petrucciani N, Simone G **S-Editor:** Dou Y **L-Editor:** Filipodia **E-Editor:**

**Specialty type:** Medicine, Research and Experimental

**Country of origin:** China

**Peer-review report classification**

Grade A (Excellent): 0

Grade B (Very good): B

Grade C (Good): C

Grade D (Fair): D

Grade E (Poor): 0



**Figure 1 The CT scan indicates a paravertebral mass close to the thoracic aorta.** A: Transverse view in mediastinal window of the normal CT scan; B: Transverse view in lung window of the normal CT scan; C: Transverse view in mediastinal window of the enhanced CT scan; D: Horizontal view in lung window of the normal CT scan. CT: Computed tomography.



**Figure 2 Video-assisted thoracoscopic surgery was performed to remove the mass.** A: The tumor was located on the posterior chest wall near the 9th thoracic vertebra; B: The drain vein to the hemiazygos vein was ligated by hemoclips; C: The tumor was dissected carefully by electrocautery and harmonic scalpel in turn; D: The base plane on the chest wall was managed by electrocoagulation.



**Figure 3 Cystic mass and immunohistochemical staining.** A, B: The tumor was removed under *en bloc* dissection with a relative clear margin; C, D: Hematoxylin-eosin stain: Neoplastic cells were arranged in irregular nests separated by fibrous septa. Cells were round or oval in shape with regular vesicular nuclei and prominent nucleoli, moderate to abundant eosinophilic or clear cytoplasm; E, F: Neoplastic cells showed strong immunohistochemical expression of melanocytic marker S-100 (+); G, H: Neoplastic cells showed strong immunohistochemical expression of the melanocytic marker HMB-45 (+).