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

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Abstract

BACKGROUND

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

CASE SUMMARY

A 31-year-old male was admitted for an uncertain mass in left pleural cavity in routine physical examination without any symptoms. A VATS surgery was performed to remove the tumor. The pathological finding displayed a cystic mass with 6.5 cm at the longest diameter, dark red in section and cysts could be found locally. A strong expression of S-100, HMB45 and Vimentin was detected in immunohistochemical staining, which was inclined to the diagnosis of the 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 at the 90th postoperative day and the 180th postoperative day, and no obvious sign of recurrence was found till now.

CONCLUSION

CCS of soft tissue also can be found in pleural cavity although in 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 (CCS) is a rare malignant tumor of soft tissue usually arising from extremities. Now we reported a case of CCS located on the chest cavity, confirmed

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by the positive stain of HMB-45 and S-100 mark in immunohistochemistry. Radical resection of the tumor was performed and the short-term outcome was good.

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INTRODUCTION

Clear cell sarcoma (CCS) of soft tissue is a rare malignant soft tissue sarcoma that represents about 1% of all sarcoma^[1]. The true origination of CCS is still uncertain, which 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 similar morphology with malignant melanoma, SSC seems more likely to derive from neural crest cells, which produces melanin typically and has positive HMB-45 and S-100 mark stained in immunohistochemistry^[2]. CCS has been usually reported to locate at distal end of extremities and rarely at trunk^[3]. According to the literature review, there is no report of CCS in pleural cavity till now.

CASE PRESENTATION

Chief Complaints

A 31-year-old male was admitted to our hospital for an uncertain mass in 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

Physical examination is normal.

Laboratory examinations

Routine blood parameters were within the normal range. Tumor marker measurement results were as follows: Carcinoembryonic antigen, 0.64 ng/mL; alphafetoprotein, 1.77 ng/mL; CA199, 5.80 U/mL. The enhanced thoracic computed tomography (CT) demonstrated a posterior mediastinal mass about 6 cm diameter in the left pleural cavity, right nearby the 9th and 10th thoracic vertebrae beside the thoracic descending aorta, with an uneven enhancement (Figure 1). A neurologic tumor was suspected as the first diagnosis.

A 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 degree with unilateral ventilation under general anesthesia. Three trocars were placed at 3th, 4th and 8th intercostal space. Under the 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 side by the 9th thoracic vertebra, close to the aorta while no obvious adhesion was found between the tumor and the aorta or vertebra. Careful dissection was done with the electrocautery and the harmonic 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 deal with electrocoagulation to ensure no residual tumor (Figure 2). The drain tube was put and the chest wall wound was close routinely. The chest tube was removed on the 1st postoperative day and the patient was discharged on the 5th postoperative day with no events.

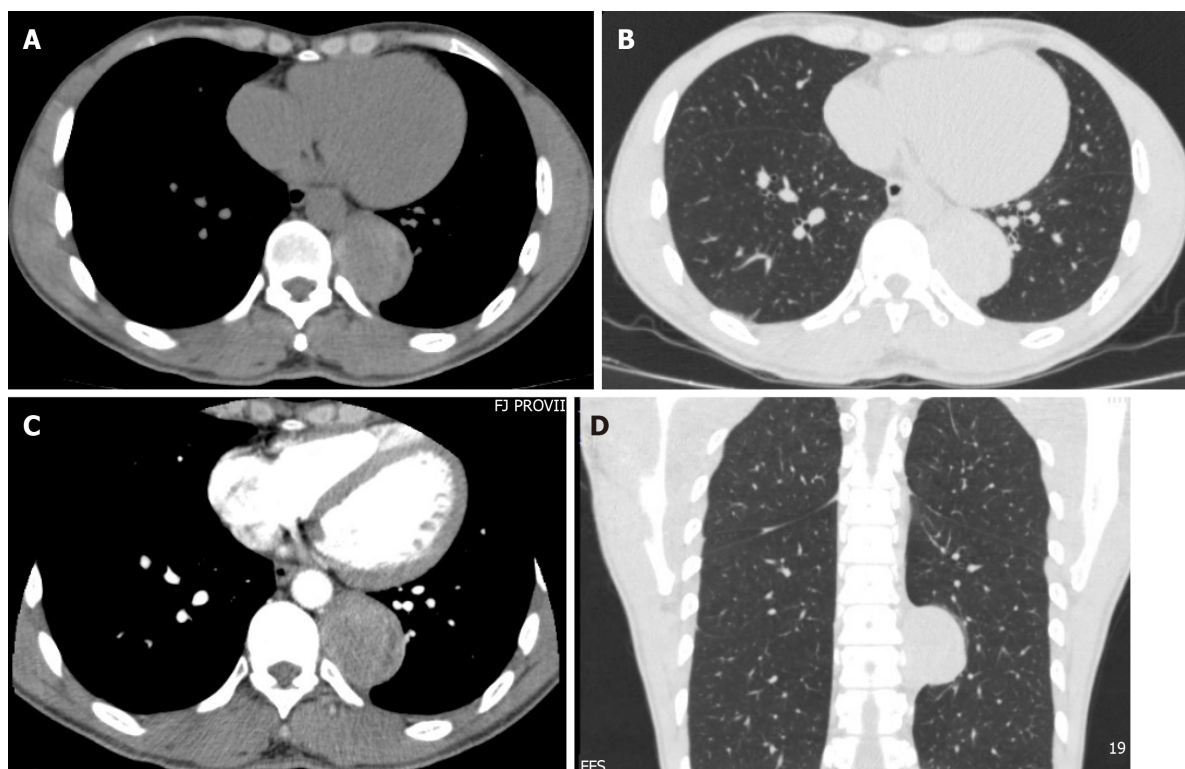


Figure 1 The computed tomography scan indicated a paravertebral mass close to the thoracic aorta. A: Transverse view in mediastinal window of normal computed tomography (CT) scan; B: Transverse view in lung window of normal CT scan; C: Transverse view in mediastinal window of enhanced CT scan; D: Horizontal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

FINAL DIAGNOSIS

The pathological finding displayed a cystic mass with 6.5 cm at the longest diameter, dark red in section and cysts could be found locally. A strong expression of S-100, HMB45 and Vimentin was detected in immunohistochemical staining (Figure 3), which was inclined to the diagnosis of the CCS of soft tissue^[2,3].

TREATMENT

A video-assisted thoracoscopic surgery was performed to dissect the tumor. After the surgery, the patient refused the suggestion of all the 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 scan were done on the 90th postoperative day and the 180th postoperative day, and no obvious sign of recurrence was found till 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 lack of pathologic immunohistochemical results. Although CCS has the similar morphologic appearance with malignant melanoma, it is possible for pathologists today to distinguish them correctly by the difference of histopathologic and cytogenetic features between them^[2]. CCS usually locates 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 foot and knee, rarely at the trunk, with no epidermal involvement^[4-9]. Differentiation diagnosis from other sarcoma

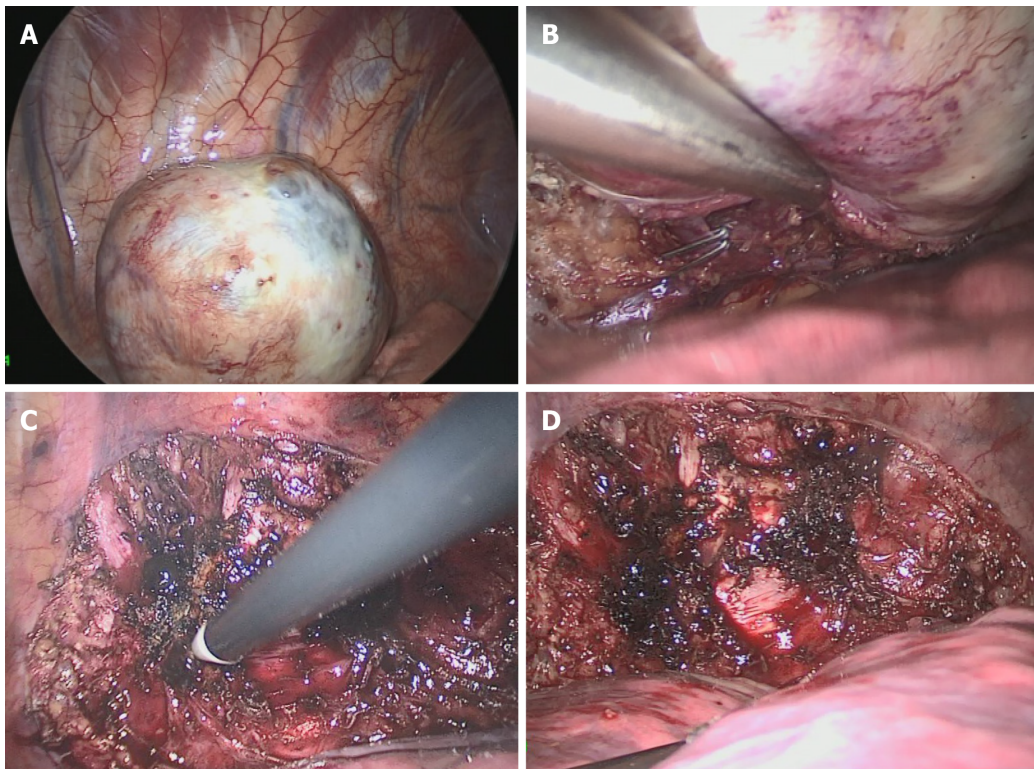


Figure 2 A video-assisted thoracoscopic surgery was performed to remove the mass. A: The tumor located on the posterior chest wall nearby the 9th thoracic vertebra; B: The drain vein to the hemiazygos vein was ligated by hemoclips; C: The tumor was dissected carefully by the electrocautery and Harmonic in turns; D: The base plane on the chest wall was deal with the electrocoagulation.

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 (MRI) of thoracic vertebra is done to exclude the 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 the reconstruction of chest wall should be made.

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 tumor residual after surgery to achieve better outcome^[11]. The targeted therapy seems to be a good treatment to 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 MRI 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 also can be found in pleural cavity although in an extremely rare incidence. Radical resection is useful to improve the prognosis.

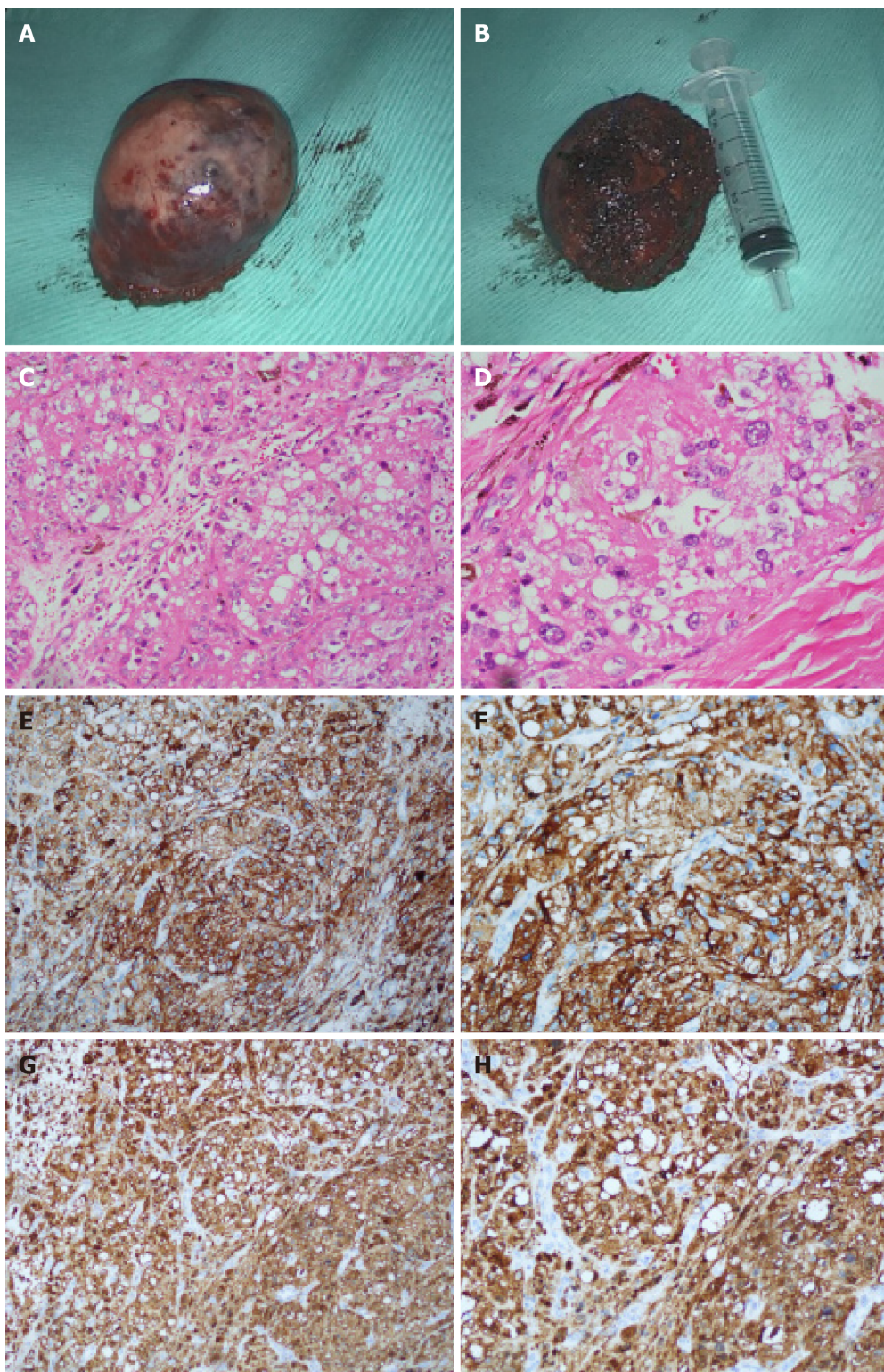


Figure 3 Cystic mass and immunohistochemical staining. A, B: The tumor was removed under *en bloc* dissection with a relative clear margin; C, D: Hematoxylin-eosin stain: Neoplastic cells are arranged in irregular nests separated by fibrous septa. Cells are round or oval in shape with regular vesicular nuclei and prominent nucleoli, moderate to abundant eosinophilic or clear cytoplasm; E, F: Neoplastic cells showed a strong immunohistochemical expression of melanocytic marker: S100 (+); G, H: Neoplastic cells showed a strong immunohistochemical expression of melanocytic marker: HMB-45 (+).

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