

# Clear cell sarcoma of the rectus sheath

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## ABSTRACT

**We report a 35-year-old Chinese woman with clear cell sarcoma of the rectus sheath aponeurosis presenting as a tender anterior abdominal mass. She was treated with wide local excision. Local recurrence and distant metastasis occurred within two months of the onset of the complaint. Clear cell sarcoma is a rare cancer with a propensity for slow progressive invasion. They occur most commonly in the extremities, and the majority of patients are young adults. This case report demonstrates an unusual site of occurrence for clear cell sarcoma.**

**Keywords:** clear cell sarcoma, malignant melanoma, rectus abdominis muscle, rectus sheath tumour

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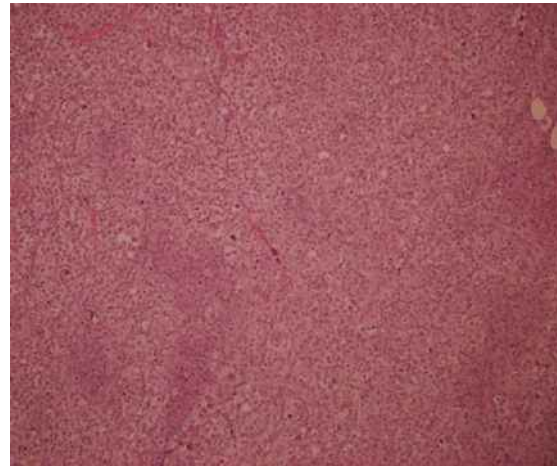
## INTRODUCTION

Clear cell sarcoma (CSS) is a rare tumour seen in adults. It occurs in a variety of unusual locations, and should be thought of as a differential diagnosis for soft tissue tumours.

## CASE REPORT

A 35-year-old Chinese woman presented with a painful central abdominal mass, which was increasing in size for one and a half months. There was no change in bowel habits, urinary symptoms or fever. She gave a history of laparotomy at 12 years of age for a benign condition. Physical examination showed a midline laparotomy scar and an 8 cm × 8 cm anterior abdominal mass that was firm, tender and erythematous. There was absence of skin pigmentation. The mass appeared to arise from the underlying rectus sheath. Cough impulse was negative. The rest of the abdomen was soft and non-tender. There was no organomegaly. An incarcerated incisional hernia was thought likely and an emergency laparotomy was planned.

Operative findings showed a 10 cm × 8 cm soft tissue tumour in the left rectus sheath involving the rectus abdominis muscle. The frozen section tested positive for malignancy, and was suggestive of an epithelial tumour. Laparotomy and careful search for



**Fig. 1** Photomicrograph shows an irregular nest of cells with areas of tumour necrosis separated by thin fibrous septae (Haematoxylin & eosin, × 40).

potential intra-abdominal primary tumour revealed an incidental left ovarian cyst. Oophorectomy was performed. Histology showed a mature teratoma. The liver, stomach, spleen, and bowels were otherwise normal. The rectus tumour was removed by wide en-bloc resection. Margins sent for frozen section were clear. Tumour markers were normal. She had an uncomplicated recovery and was discharged well on the fourth postoperative day.

Histology revealed a circumscribed tumour in the reticular dermis with a nesting pattern in some areas containing diffuse sheet-like pattern in other areas. The tumour cells were separated by delicate connective tissue containing vascular channels lined by flattened endothelium (Fig. 1). Loss of cellular cohesion and necrosis of the centrally-located cells within the nests were noted. The individual cells were large, round and exhibited cellular and nuclear pleomorphism, and contained vesicular nucleus with prominent nucleolus and high mitotic activity (Fig. 2). Cytoplasm appeared abundant and eosinophilic, was faintly periodic acid Schiff (PAS) positive, and Mucicarmine and PAS-digested stain negative. Immunostains tested the tumour cells positive for vimentin, S100, Melan A, HMB45 and calponin (Fig. 3). They were negative for cytokeratin AE1/3, PLAP, HCG, MyoD1, SMA and caldesmon. Diagnosis was a malignant

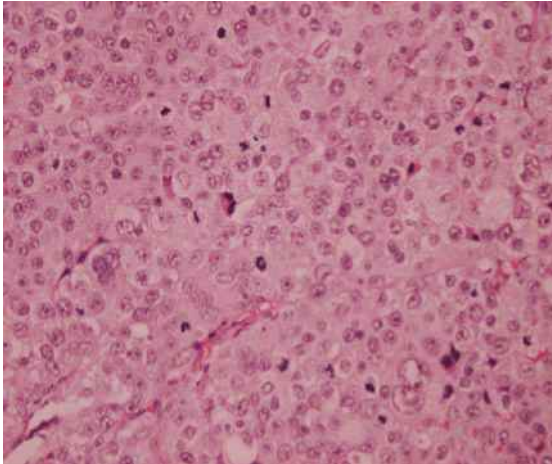
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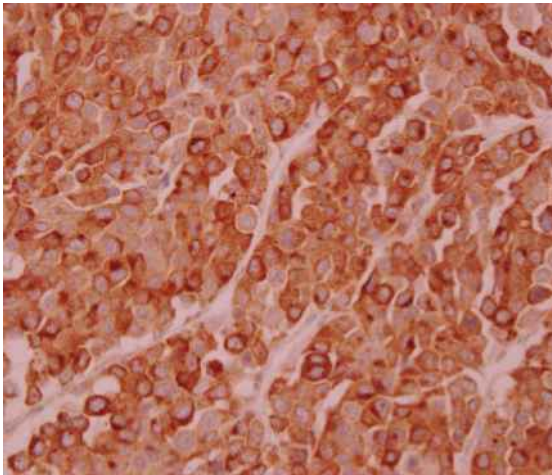
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**Fig. 2** Photomicrograph shows tumour cells with enlarged nuclei, prominent nucleoli with clear and eosinophilic cytoplasm and atypical mitoses (Haematoxylin & eosin,  $\times 400$ ).



**Fig. 3** Photomicrograph shows tumour cells with immunopositivity to HMB-45 (HMB45 immunostain,  $\times 400$ )

tumour with histological and immunohistochemical features of a CCS.

Computed tomography (CT) of the abdomen performed postoperatively showed multiple irregular hepatic lesions consistent with hepatic metastasis. Bone scintiscan and CT of the thorax were negative. She was admitted again two months later for abdominal pain. Repeat CT showed free fluid in the pelvis, multiple enlarged intra-abdominal lymph nodes, and three subcutaneous nodules in the anterior abdominal wall. She was referred to medical oncology for palliation.

## DISCUSSION

CCS, also known as malignant melanoma of soft parts, is a tumour with a propensity for lymphatic

spread. It was described by Enzinger as “clear-cell sarcoma of tendons and aponeuroses”.<sup>(1)</sup> It occurs in the foot, hand and wrist of young adults,<sup>(2)</sup> but has been described in the penis<sup>(3)</sup> and gastrointestinal tract. Mean age of diagnosis is 44 years.<sup>(2)</sup> Grossly, such tumours are firm, gray/white with a gritty sensation and well-circumscribed. Microscopical features show solid nests and fascicles of pale fusiform or cuboidal cells. Nucleoli are large and deeply basophilic. Multinucleated giant cells are often present. Iron is present within and outside of cells. Some cells contain cytoplasmic melanin. This suggests a neuroectodermal derivation that is characteristic of malignant melanoma of soft parts. The tumour exhibits immunosensitivity to S-100 protein,<sup>(4)</sup> HMB-45,<sup>(4)</sup> Leu7, NSE, and vimentin, which is seen in conventional melanoma.<sup>(5)</sup> Melanosomes and features consistent with neural derivation are seen ultrastructurally. CCS is more likely to be diploid or show lesser degrees of aneuploidy, compared to conventional skin melanoma with metastasis to soft parts on DNA ploidy analysis. An association with translocation  $t(12;22)(q13;q12)$  is seen. EWS-ATF1 gene fusion and expression of melanocyte-specific splice form of the MIFT transcript is the result.<sup>(6)</sup> ERBB3 is being studied as a new marker gene.<sup>(7)</sup>

CCS is slowly progressive. Frequent local recurrences and eventual nodal and distant metastasis (usually pulmonary) characterise the disease. CCS has a high propensity for distant spread. Once regional lymph node metastasis or haematogenous spread has occurred, prognosis is adverse. Other poor prognostic indicators include large tumour size, necrosis, and local recurrence. Patients with a tumour  $\leq 2$  cm have better survival rates compared to patients with a larger/still-localised tumour.

Early diagnosis and initial radical surgery are important for a good outcome. Sentinel lymph node biopsy is being investigated in the management of CCS as they help avoid radical lymphadenectomy in node negative patients.<sup>(8)</sup> Adjuvant radiotherapy may be used when resection margins are  $< 1$  mm.<sup>(9)</sup> However, other series showed that aggressive multi-agent chemotherapy and radiotherapy have little impact on outcome.<sup>(10)</sup> A French series of 36 patients showed three- and five-year survival rates at 72% and 62%, respectively.<sup>(2)</sup> In another series of eight patients, two- and five-year disease-free rates were 68% and 50%, respectively, while two- and five-year survival rates were 86% and 68%, respectively.<sup>(10)</sup> CCS remains a rare tumour with a potentially good long-term outcome, and may be considered as a rare differential in soft tissue tumours.

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