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Clear cell sarcoma (melanoma of soft parts): The Royal Marsden Hospital experience

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Abstract

Introduction: Clear cell sarcoma (CCS) is a rare tumour with a propensity for local recurrence and nodal metastasis. About 300 cases have been reported, thus further clarification regarding the course and outcome of the disease is required.
Methods: Patients with a histopathologic diagnosis of CCS were identified from prospective histopathology and sarcoma databases and supplemented with a retrospective analysis of the patients' hospital records.
Results: Between 1990 and 2005, a total of 72 patients with a diagnosis of CCS were identified, 35 having been referred for management and 37 having been referred for histopathologic opinion. The median age was 39 years (range 5–90 years). Of the 35 patients referred to the Royal Marsden Hospital for management, 23% developed local recurrence or in-transit metastases at a median of 9 months (2–79 months) after resection of the primary, and nodal or distant metastatic disease was seen in 63% after 14 months (range 0–177 months). Five- and 10-year survival were 52% and 25%, respectively.
Conclusions: CCS has a number of similarities with melanoma, particularly in its peripheral distribution and propensity for nodal disease. Wide excision with clear margins offers the best chance of cure. Local recurrence and regional metastases are common, and are almost always followed by distant metastases and death.

Keywords: Clear cell sarcoma; Melanoma of soft parts; Soft-tissue sarcoma

Introduction

Clear cell sarcoma (CCS) of soft tissue,[1] previously known as clear cell sarcoma of tendons and aponeuroses, is a malignant soft-tissue tumour with melanocytic differentiation. It typically involves the peripheral tendons and aponeuroses of young adults. It is a rare tumour with around 300 cases described. CCS is primarily treated by wide surgical resection (Fig. 1). Although originally categorised by Enzinger as a soft-tissue sarcoma (STS),[2] it has characteristics of both STS (deep soft-tissue primary location, propensity for pulmonary metastasis) and melanoma (distal limb distribution, tendency for local recurrence, in-transit disease and regional nodal spread). The alternative nomenclature of melanoma of soft parts (MSP) has also been used in the past.[3] Recent discoveries include the finding of a characteristic chromosomal translocation, which is unique to CCS, fuelling conflicting interpretations of its classification and histogenesis.[4,5] What is beyond doubt is the aggressive nature of the disease, with overall 5-year survival of between 40 and 67%.[6–9]

The Royal Marsden Hospital (RMH) is Europe's largest tertiary referral centre for the treatment of STS and other rare soft-tissue tumours. Further patients are referred for specialist pathological review after clinical management has been performed at other institutions. We report our experience with CCS, with a particular focus on the clinical behaviour.

Patients and methods

Patients referred for clinical management were identified from the RMH sarcoma database. Details of treatment, histopathology and outcome were obtained from the prospective

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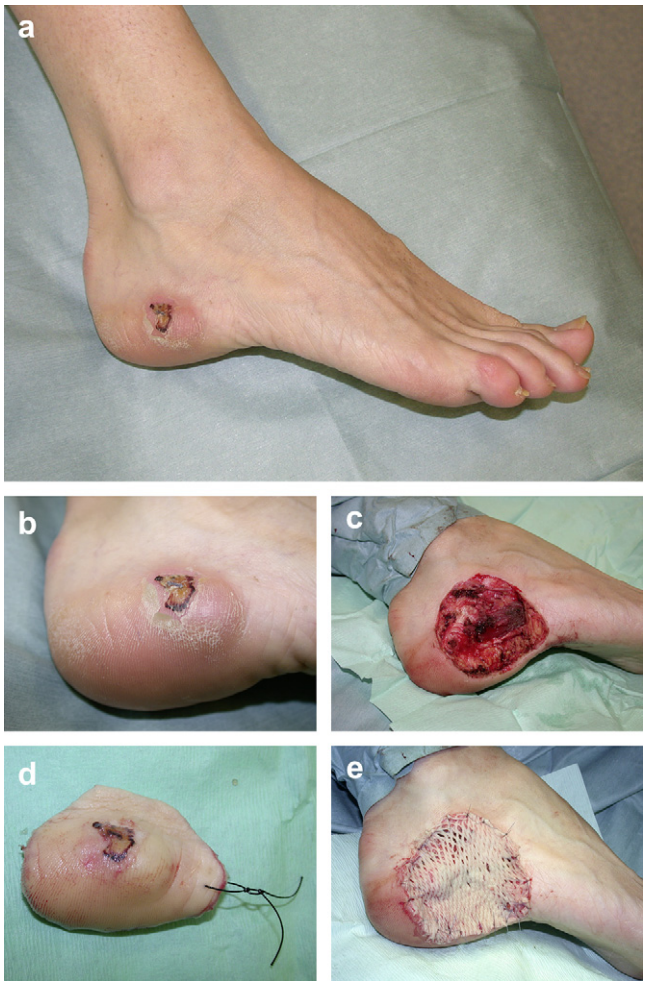

Figure 1. (a) Clear cell sarcoma of the heel. (b) Close-up. (c) Bed of resection. (d) Resected and orientated specimen. (e) Site grafted with meshed skin.

database, supplemented with hospital and General Practitioner records.

Patients referred only for pathological review were identified from the Pathology department database. Demographic information and site details were usually obtainable on these patients, but clinical information was not available.

In all patients, the pathological specimens were diagnosed and reported by a single Pathologist with experience in soft-tissue tumours (CF). The diagnosis was made according to standard criteria.[1,10]

Survival curves were constructed according to Kaplan–Meier method.[11] A Cox proportional hazard model[12] was used to identify variables influencing survival.

Results

Seventy-two patients with a histopathologic diagnosis of clear cell sarcoma were identified. Of these, 35 patients were referred to RMH for management and 37 patients were referred for histopathologic diagnosis or confirmation.

Tumour demographics

Demographic data for all patients are shown in Table 1. Median age was 39 years (range 5–90 years) with an M:F ratio of 44:28. Limb sites predominated, and over half of the limb tumours arose in hand or foot. Only five tumours arose from central sites and chest wall.

There was no difference in tumour demographics between those patients referred for pathological review and those referred for clinical management. Comprehensive clinical information was available only for patients treated at RMH, and all subsequent analyses refer to this group of patients.

A median follow-up of 41 months (range 1–321 months, average 62 months) was recorded, with three patients lost to follow-up.

Treatment

In 94% of patients the initial excision was performed before referral to RMH because the diagnosis was not suspected. Initial resections were intra-capsular (with positive margins) in 14, marginal (with narrow but clear margins) in four, and wide in five patients; margins were unable to be determined in 12. Patients were usually referred to RMH once the histologic diagnosis was determined, after a median period of 12 weeks.

Twelve patients subsequently had wider excisions; in 10 patients residual tumour was identified at this operation.

Surgery was also performed where necessary for true local recurrence, for in-transit metastases, for clinical nodal metastases, and for isolated distant metastases (including pulmonary metastasectomy in one patient). No further recurrences occurred in the nodal basin after seven therapeutic lymphadenectomies performed at RMH.

Adjuvant radiotherapy was delivered to the site of the excised primary tumour in 15 patients.

Adjuvant chemotherapy was used in a number of patients, and as palliative treatment for non-resectable pulmonary metastases.

Few tumours demonstrated chemosensitivity—partial responses were only seen occasionally, and no complete responses were noted (Table 2).

Table 1
Demographics for all patients and for subset referred for clinical management

	All patients	Patients referred for clinical management	Patients referred for histological opinion
Number of patients	72	35	37
Sex M:F (% male)	44:28	24:11	20:17
Median age (range)	39 Years (range 5–90)	38 Years (range 12–76)	39 Years (range 5–90)
Site			
Lower limb	35	14	21
Upper limb	26	16	10
Other	11	5	6