



Chemotherapy in Clear Cell Sarcoma

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BACKGROUND

Clear cell sarcoma (CCS) is a rare soft tissue sarcoma subtype characterised by the chromosomal translocation t(12;22)(q13;q12)/ *EWSR1-ATF1* fusion. The mainstay of treatment is surgery. CCS is a chemo-resistant disease, but few studies have directly recorded the response and progression-free survival (PFS) of patients treated with palliative chemotherapy. The aim of this study was to document the response and PFS in a series of patients treated at a single referral centre.

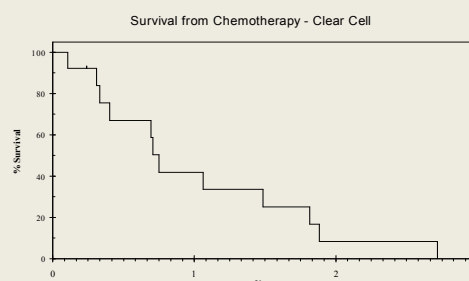
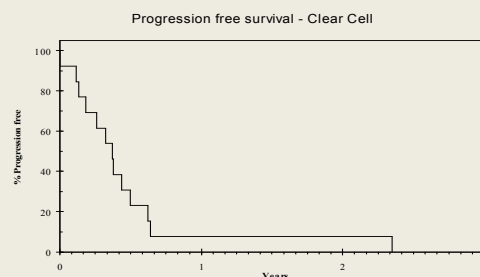
PATIENTS & METHODS

A retrospective search of the prospectively maintained Royal Marsden Hospital Sarcoma Unit database was made to identify patients treated with palliative chemotherapy between 1990 and 2009. Response to treatment was assessed using RECIST and WHO criteria.

RESULTS

Thirteen CCS patients were treated with palliative first line chemotherapy. The median age at presentation was 38 years (15-65). The response rate was: Partial Response 8% (1), Stable Disease 46% (6), Progressive Disease 46% (6). The median PFS following palliative first-line chemotherapy was 4 months (95%CI : 3 - 6 months). One patient stopped first-line chemotherapy due to ifosfamide induced encephalopathy. Two patients experienced febrile neutropenia and one developed severe stomatitis. At the time of analysis 12 patients had died and one patient was lost to follow-up. The median overall survival from start of chemotherapy was 9 months (95%CI : 8 - 10 months).

Patients (gender)	Primary site	Previous surgery	Previous radiotherapy	Systemic treatment (Response)
1 (M)	Right buttock	1 for local disease 0 for metastatic disease	40 Gray (local recurrence)	Doxorubicin/ cisplatin – ifosfamide (PD) Ifosfamide (PD) Doxorubicin (PD)
2 (M)	Pelvis	1 for local disease 0 for metastatic disease	Adjuvant	Doxorubicin/ cisplatin (SD)
3 (M)	Right arm	2 for local disease	Adjuvant	Epirubicin (SD) Phase I (SD) Ifosfamide (PD)
4 (M)	Right inguinal	2 for local disease 1 for metastatic disease	None	Ifosfamide/ carboplatin/ epirubicin/ etoposide (PD) Ifosfamide/ dacarbazine (PD) Doxorubicin (PD)
5 (M)	Right foot	2 for local disease 0 for metastatic disease	None	Doxorubicin/ ifosfamide (SD)
6 (F)	Left hand	10 for local disease 1 for metastatic disease	61 Gray in 30# (arm) 35 Gray in 15# (axilla)	Doxorubicin/ ifosfamide (PR) Ifosfamide (SD)
7 (M)	Right shoulder	2 for local disease 3 for metastatic disease	Adjuvant 18 Gray in 3# (lung)	Doxorubicin (SD) Ifosfamide (PD)
8 (F)	Left thigh	2 for local disease 1 for metastatic disease	35 Gray in 15# (spine)	Doxorubicin (PD)
9 (F)	Vulva	1 for local disease 1 for metastatic disease	60 Gray in 30# (inguinal)	Doxorubicin (SD)
10 (M)	Left wrist	2 for local disease 0 for metastatic disease	Adjuvant 35 Gray in 15# (thorax)	Doxorubicin (PD)
11 (M)	Left achilles tendon	2 for local disease 0 for metastatic disease	None	Doxorubicin (SD) Phase I (PD)
12 (M)	Right foot	1 for local disease 4 for metastatic disease	None	Doxorubicin (PD)
13 (F)	Left thumb	2 for local disease 0 for metastatic disease	Adjuvant	Ifosfamide (PD) Doxorubicin (PD)



CONCLUSION

Conventional chemotherapy consisting of doxorubicin and/ or ifosfamide has minimal efficacy in clear cell sarcoma.