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Poster Abstracts

Clear Cell Sarcoma. The Royal Marsden Experience

Authors: Matthew A Clark, Maria B Johnson, Khin Thway, Cyril Fisher, J Meirion Thomas, Andrew J Hayes

Contact: mariajohnson13@hotmail.com

Sarcoma and Melanoma Unit, Royal Marsden Hospital, London

Aim:

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

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 nine months (2–79 months) after resection of the primary, and nodal or distant metastatic disease was seen in 63% after 14 months (0–177 months). Five and ten year survival was 52%, and 25% retrospectively.

Conclusions:

CCS has a number of similarities to 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.