



Chemotherapy in Epithelioid Sarcoma

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Aim:

Epithelioid sarcoma is a rare soft tissue sarcoma subtype, which is prevalent in young adults. It typically affects the extremities and multiple local recurrences are common. A more aggressive proximal-type has also been described. The aim of this study was to document the efficacy and toxicity of palliative first-line chemotherapy in metastatic epithelioid sarcoma.

Methods:

A retrospective search of the prospectively maintained Royal Marsden Hospital Sarcoma Unit database was made to identify epithelioid sarcoma patients treated with chemotherapy between 1990 and 2009. The histopathology was confirmed in all cases by experienced soft tissue pathologists. RECIST and WHO criteria were used to assess response.

Results:

Fifty-five epithelioid sarcoma patients were identified, 20 of these were treated with chemotherapy (18 males). The median age was 33 years (range 17-64). The primary tumour site was the upper limb in 8 (40%), lower limb in 3 (15%) and other in 9 (45%). Nine (45%) had the proximal-type and 11 (55%) did not. Nineteen patients were evaluable for response: partial response 5 (26.3%), stable disease 9 (47.4%) and progressive disease 5 (26.3%). The median progression-free survival was 7 months (95%CI, 4-9). The median overall survival was 11 months (95%CI, 5-21). An analysis of the relative benefit of chemotherapy in the proximal-type and classical epithelioid sarcoma will be presented.

Seven patients received second-line chemotherapy; two achieved stable disease and 4 progressive disease. One patient is currently receiving second-line therapy. Three received third-line therapy; 2 achieved stable disease and 1 progressed.

Chemotherapy was well tolerated. One patient received one cycle of adjuvant chemotherapy but had to stop due to ifosfamide-induced encephalopathy. Two patients treated with first-line chemotherapy were admitted with neutropenic sepsis. One treated with second-line chemotherapy was admitted with neutropenic sepsis. No grade 3 or 4 treatment related toxicity was observed in those treated with third-line chemotherapy.

Conclusion:

Epithelioid sarcoma is a chemosensitive soft tissue sarcoma histological subtype.