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ABSTRACTS – ORAL PAPERS

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Treatment of aggressive fibromatosis with pegylated liposomal doxorubicin (Caelyx): the Royal Marsden Hospital experience

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Background

Aggressive fibromatosis (AF), or desmoid tumours are monoclonal proliferations which are locally invasive but do not metastasise. Sporadic tumours are usually associated with mutations in the beta-catenin gene *CTNNB1*, whereas those occurring in the context of familial adenomatous polyposis usually have inactivating mutations in *APC*. Histologically, they are characterised by nuclear expression of beta-catenin. When surgery and radiotherapy are not applicable or fail to control the disease, systemic treatment in the form of anti-oestrogens, non steroidal anti-inflammatory drugs (NSAIDs) and chemotherapy can be used. A variety of regimens are reported to have activity including methotrexate/vinblastine and doxorubicin/dacarbazine. Recent reports indicate that single agent pegylated liposomal doxorubicin (Caelyx) is also effective.

Methods

10 patients with AF received Caelyx between June 2006 and December 2008. Caelyx was administered intravenously at a dose of 50 mg/m² over 1 hour every 4 weeks, with dose reductions or dose delay as indicated according to toxicity.

Results

The female/male ratio was 9:1 and the median age at presentation was 39.5 years (range 18-53). All patients had progressive fibromatosis the primary sites of which included: trunk/parietal (1), limb (3), head and neck (1), abdomen (2) perineum (1), chest wall (1), branchial plexus/axilla (1). Fifty percent had previously been treated with surgery/radiotherapy or both. All but one had previous systemic therapy which comprised tamoxifen/toremifene (6), NSAIDs (1), chemotherapy (1) and imatinib (1). Caelyx was well tolerated with mucositis (6/10), palmar-plantar erythema (4/10) and fatigue (2/10) being the main reported toxicities. A dose reduction to 40 mg/m² was required in 50% of cases, hence the optimal dose lies between 40 and 50 mg/m². One patient is currently receiving treatment and is too early to assess. For the nine patients who have completed treatment the median number of Caelyx cycles was 6 (range 4-6). Objective response according to RECIST was achieved in 4/10 patients and in 5 patients the best response was stable disease. Clinical benefit (pain relief, improved mobility) was observed in all patients. The duration of response ranged from 4 to 28 months.

Conclusion

This is the largest series of patients with AF receiving Caelyx presented to date. Caelyx as single agent therapy has acceptable toxicity and highly promising activity in unresectable AF and may provide long term clinical benefit in some patients.