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High local recurrence rate after surgical treatment of extra-abdominal desmoid-type fibromatosis

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Introduction: Desmoid-type fibromatosis (DF) is a rare monoclonal, fibroblastic proliferation that usually arises in deep soft tissues. Although histologically benign they are characterized by infiltrative growth. Surgical treatment was often the first choice except if mutilating and associated with considerable function loss. However, choosing the right treatment for DF is challenging because of the unpredictable behaviour, including long periods of stable disease or even spontaneous regression. This study is meant to quantify the results of different treatment modalities, in order to change our strategies in the future, if indicated.

Methods: Eighty-two patients who were treated for extra-abdominal DF (52 female, 63%) from 1978-2014 were retrospectively studied. Mean age was 41 years (8-79). Sixty-four (78%) presented with a primary lesion; 18 (22%) were referred for a residual or recurrent lesion following treatment elsewhere. Seventy-six patients (93%) had unifocal disease. Lesions predominantly affected the thorax and trunk (n=33, 40%), thigh (n=11, 13%), head/neck (n=8, 10%) and shoulder (n=8, 10%).

Results: Of 64 patients presenting with a primary lesion, 28 (44%) underwent surgery alone (2 wide margins, 10 marginal, 17 intralesional or questionable), 19 (30%) surgery and radiotherapy, eight (13%) radiotherapy alone, four (6%) received systemic treatment (tamoxifen) and four (6%), watchful waiting (three had symptomatic treatment with NSAIDs). Twenty-two (34%) had progression after treatment; progression rates were 0% after radiotherapy alone (0/8), 26% after surgical treatment with adjuvant radiotherapy (5/19), 50% after surgery alone (14/28) and 50% after systemic treatment (2/4). First relapses all occurred within three years (range). Of 18 patients who presented with residual or recurrent lesions in our center, nine experienced a further relapse after treatment (50%).

Conclusions: Progression of disease was frequent, especially in patients presenting with a residual or recurrent lesion. Local control of primary lesions was most often achieved with radiotherapy or a limited resection combined with radiotherapy. Based on the high recurrence rate in this study we recommended a more watchful waiting policy, such as the recent European consensus.