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Extraabdominal desmoid tumors – From watchful waiting to extensive surgery: results of different treatment modalities in Switzerland

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Introduction: Extraabdominal desmoid tumors (DT) are benign and rare with a persistent treatment dilemma due to its high recurrence rate and its heterogenous behaviour. In the past wide local excision was the usual treatment, however the risk of local recurrence was high (> 40%). The goal of the retrospective study is to evaluate the mid- and longterm results of extraabdominal desmoid tumors with respect to the different treatment modalities in Switzerland.

Material and Methods: From January 1970 through December 2013 93 patients with DT were initially treated with surgery (n = 41) or surgery with radiation (n = 14). Further alternative initial treatment options were radiation alone (n = 7), systemic therapy (n = 12) or waitful watching (n = 19) in the three sarcoma centers. Patient demographics, tumor site and histology, history of previous recurrences, and follow-up status with recurrence rates of the different treatment modalities were recorded retrospectively.

Results: There were 59 females and 34 males with a mean follow-up of 6 years in the current study. All patients presented with primary tumors located at the extremities in 51%, pelvis in 6%, trunk in 17% and at the abdominal (15%) or thoracic (11%) wall. By immunohistochemistry, 86% of patients in this study were positive for beta-catenin. Overall 45 % (n = 25) of DT patients developed a recurrence after primary surgical treatment (n = 55). The overall recurrence rate in the surgery group was 50%, depending on the resection margins with wide margins 36 %, 50 % with marginal resection margins, and 80% with intralesional resection. In the surgical plus radiation group 36% presented a recurrent DT. The DT patients who received radiation alone, all patients presented stable or regredient disease. The patients with systemic therapy showed in 33% progression and required further treatment. In the watchful waiting group we observed 67% stable disease, 21 % spontaneous regression and in 12% progression.

Conclusion: The results of our observation group could encourage us not initially resect DT and to wait with careful observation. Besides surgery, radiation is an additional treatment option for better local control also in our study. Extraabdominal DT have been also treated with systemic therapy. The goals of the medical therapies are the stabilisation of the disease, which has achieved in a similiar rate than the therapy with surgery plus radiation in our study. The result of this study should be the basis for clear guidelines for a therapeutical strategy for these patients. In a second stage, a prospective study should be carried out in order to evaluate the adequacy of the strategy.