Sporadic extra-abdominal desmoid-type fibromatosis: high relapse rates after surgical treatment

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Background
Desmoid-type fibromatosis (DF) continues to pose a significant challenge to surgeons, radiotherapists and oncologists. Although DF does not metastasize, its infiltrative growth and frequent recurrences commonly lead to significant morbidity. DF lesions may occur throughout the entire body (both intra- and extra-abdominally), in a sporadic form or associated with familial adenomatous polyposis. For many years, surgery has been the mainstay of treatment for DF. Several authors however reported dissatisfying results of surgical treatment, while others demonstrated that spontaneous regression may occur. Therefore, various centers have encouraged a more conservative treatment strategy[1]. Recently, a consensus treatment algorithm for DF was published[2]. For any type and localization of DF, a watch and wait policy was advocated. In case of progression of lesions affecting the extremities, girdles or chest wall, surgical (if morbidity is limited) or medical therapy was advocated. However, there is little evidence to support these recommendations.

Questions / Purposes
We therefore evaluated our single-center experience in the treatment of sporadic extra-abdominal desmoid-type fibromatosis, with the aims to compare relapse rates after different types of treatment and to assess whether our experiences are in agreement with recent treatment proposals.

Patients and methods
We retrospectively identified all consecutive patients who were treated for symptomatic sporadic extra-abdominal DF in our tertiary referral center, between 1978 and 2014. Seventy-four patients (46 female, 62%) with a median age of 40 years (8-78) at the time of diagnosis were identified. Fifty-six patients (76%) presented in our clinic with a primary lesion, 18 (24%) were referred for a residual or recurrent lesion following treatment elsewhere. Sixty-nine patients (93%) had unifocal disease. Lesions predominantly affected the thorax and trunk (n=31, 42%), thigh (n=11, 15%), head/neck region (n=7, 10%) and shoulder girdle (n=6, 8%). Of 56 patients presenting with a symptomatic primary lesion, 29 (52%) underwent surgery alone, 19 (34%) surgery and radiotherapy and eight (14%), radiotherapy alone (usually 56 Gy in 2 Gy fractions). Relapse-free survival was estimated with Kaplan-Meier curves and compared between groups using log-rank tests.

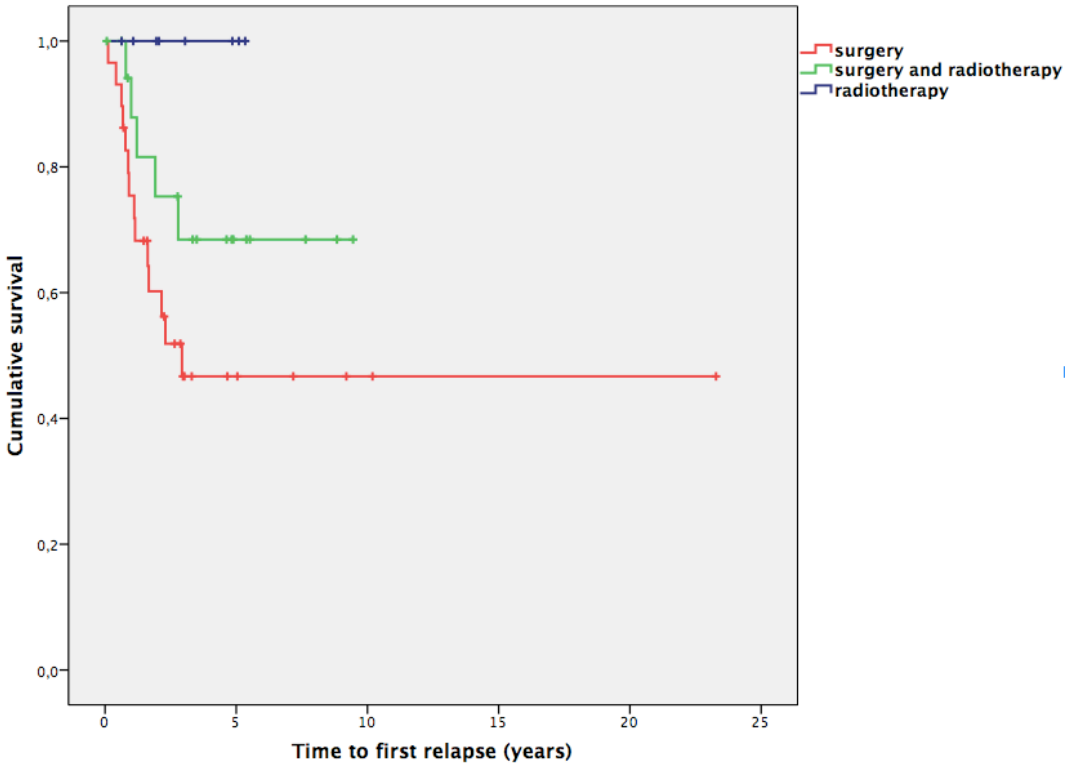
Results
Mean follow-up for the patients that presented with a primary lesion was 6.6 years (0.4-27.6). Of these patients, 19 (34%) had a relapse after treatment; relapse rates were 0% (0/8) after radiotherapy alone, 26% (5/19) after surgical treatment with adjuvant radiotherapy and 48% (14/29) after surgery alone (p=0.05, figure 1). First relapses all occurred within three years after primary treatment. One patient who was treated with surgery and radiotherapy (46 Gy in 2 Gy fractions, plus a boost of 14 Gy) developed a radiation-induced sarcoma 6.2 years after treatment and died of disease.

Conclusions
Progression of disease was frequent after treatment of sporadic extra-abdominal desmoid-type fibromatosis, especially in patients who underwent surgical treatment only. Local control was most often

achieved with radiotherapy alone or a limited resection followed by radiotherapy. While the patients who were offered radiotherapy as an adjuvant after surgery were presumably considered to be at high risk for relapse, the risk of progression was lower in these patients than in patients receiving surgery alone; indicating the beneficial effect of added radiotherapy. Other than the recent consensus treatment algorithm for patients with progression sporadic extra-abdominal DF, given the excellent results obtained with irradiation in our series, we currently consider radiotherapy alone or a limited resection with adjuvant radiotherapy the treatment of choice. In asymptomatic lesions, we recommend a wait and see policy.

Level of evidence: therapeutic studies, level IV

Figure 1: Kaplan-Meier curve demonstrating survival to disease progression, stratified according to primary treatment (log rank test, p=0.05).



References

1. Colombo, C.*, et al.* Sporadic extra abdominal wall desmoid-type fibromatosis: surgical resection can be safely limited to a minority of patients. *European journal of cancer (Oxford, England : 1990)* 51, 186-192 (2015).
2. Kasper, B.*, et al.* Management of sporadic desmoid-type fibromatosis: a European consensus approach based on patients' and professionals' expertise - a sarcoma patients EuroNet and European Organisation for Research and Treatment of Cancer/Soft Tissue and Bone Sarcoma Group initiative. *European journal of cancer (Oxford, England : 1990)* 51, 127-136 (2015).