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Original Article

Treatment of Aggressive Fibromatosis: the Experience of a Single Institution

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ABSTRACT:
Aims: **Aggressive fibromatosis is a locally aggressive infiltrative low-grade tumour, although pathologically benign, and it does not metastasise, yet it can cause serious local distressing symptoms by virtue of local destruction and impairment of local function. The aim of this study was to emphasise the role of radiotherapy and adequate surgery in the treatment of fibromatosis in patients presenting with newly diagnosed or recurrent disease and to analyse our treatment results over 15 years for this rare tumour type.**
Materials and methods: **Fifty-four patients with confirmed diagnosis of aggressive fibromatosis treated at King Faisal Specialist Hospital between 1990 and 2006 were identified from our local cancer registry. Forty-seven patients had surgery: complete resection (R0) in 20 patients, incomplete surgery (R1/2) in 27 patients, and seven patients had biopsy only. Forty-five patients were treated with radiotherapy: 38 patients were treated with postoperative radiotherapy, three patients were treated with preoperative radiotherapy and four patients had radiotherapy as the only treatment. The radiotherapy dose ranged between 45 and 60 Gy (median 50.4 Gy). Three patients did not receive any form of treatment apart from biopsy, but were still included in the final analysis.**
Results: **Fifty-two per cent (28/54 patients) of our patient population had tumour recurrence when first presented to King Faisal Specialist Hospital. The median age was 29.5 years (range 2–63 years). The most common site of involvement was the extremities (28 patients). Among the 54 patients (with primary and recurrent presentation) there were 10 local recurrences, all of which were within the original primary site. The 5-year progression-free survival and overall survival rates for the whole group were 75 and 95%, respectively. Univariate and multivariate Cox regression analysis showed that the depth of invasion significantly affected progression-free survival.**
Conclusion: **Aggressive fibromatosis is effectively treated with surgery and postoperative radiotherapy. Patients first presenting with tumour recurrence may still have local tumour control comparable with newly diagnosed patients.**
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Key words: Fibromatosis, local tumour control, radiotherapy, surgery

Introduction

Aggressive fibromatosis is a locally aggressive infiltrative low-grade tumour. It virtually never metastasises, but can cause local destruction and organ dysfunction through infiltration of blood vessels and/or nerves. Radical surgery is paramount in local control, but because of the tumour's infiltrative nature, positive surgical margins and, hence, tumour recurrence are common findings [1,2]. Almost 100 years ago, James Ewing proposed treating inoperable tumours with radiation [3]. At that time, the precise role of radiotherapy was still not clearly defined [4]. Radiotherapy may be given as adjuvant treatment to decrease local recurrence, especially when the surgical margin is positive, or as a single treatment in cases where surgery will result in severe morbidity or disruption of organ function [5]. Nuyttens *et al*. [6], in a comparative review between surgery and radiotherapy, found that local control can approach 83% for primary radiotherapy.

The aim of the present study was to analyse our treatment results in this rare tumour type and to identify the important prognostic factors that could affect treatment outcome.

Materials and Methods

This was a retrospective analysis of patients with primary and recurrent aggressive fibromatosis treated at King Faisal Specialist Hospital (KFSH) between 1990 and 2006.

This study was approved by the research accreditation and ethical committee at KFSH in 2007.

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Patient Data Sets

Fifty-four patients with confirmed diagnosis of aggressive fibromatosis treated at KFSH between 1990 and 2006 were identified from our local cancer registry. All pathological specimens referred from outside hospitals were reviewed. Information taken from each patient's chart included: gender, birthday, tumour location, operations before referral to KFSH, tumour size, surgery date, microscopic marginal status, radiation dose and fractionation, tumour control status, date of recurrence if any, date of last follow-up, and radiation complications if any. Late radiation complications were reported according to the Radiotherapy Oncology Group and European Organization for Research and Treatment of Cancer common toxicity criteria in a retrospective manner.

Patient Characteristics

Fifty-two per cent (28/54 patients) of our patient population had tumour recurrence when first presented to KFSH. There were 37 women and 17 men, with a female to male ratio of 2.l: 1. The most common presenting symptom was a palpable mass in 29 patients (53.7%), 17 (31.5%) patients presented with both mass and pain. The median age of the studied population was 29.5 years (range 2–63 years). The duration of symptoms ranged between 1 and 120 months, with a median of 18 months. There were equal numbers of patients with either superficial (27 patients; 50%) or deeply seated tumours (27 patients; 50%). Superficial tumours were defined as lack of any involvement of the superficial fascia in extremity or trunk lesions; deep lesions were defined as: (a) all intraperitoneal, retroperitoneal, and most head and neck lesions; (b) lesions deep to or which involved the superficial fascia, in agreement with the American Joint Committee on Cancer staging 2002 [7]. The most common involved site at presentation was the extremities (28 patients; 51.8%), followed by the trunk (21 patients; 38.8%). The remaining five (9.2%) patients presented with tumours in the head and neck region. Twenty-eight patients had tumours measuring more than 10 cm. Patient characteristics are shown in Table 1.

Table 1 – Patient characteristics

Characteristic	All patients ($n = 54$)
Gender	
Male	17 (31%)
Female	37 (69%)
Median age (range)	29.5 (2–63)
Presentation	
Primary	26 (48%)
Recurrent	28 (52%)
Tumour size	
>10 cm	28 (52%)
≤ 10 cm	26 (48%)
Resection status	
R0	20 (37%)
R1/R2	27 (50%)
Biopsy only	7 (13%)
Site of tumour	
Extremity	28 (52%)
Trunk	21 (38%)
Head and neck	5 (10%)
Depth of infiltration	
Superficial	27 (50%)
Deep	27 (50%)
Type of radiotherapy ($n = 45$)	
Preoperative	3 (6%)
Postoperative	38 (84%)
Single modality treatment	4 (7.4%)
Median dose (range)	50.4 Gy (45–60)
Symptoms	
Mass	29 (54%)
Pain	3 (6%)
Both	17 (31%)
Others	5 (9%)

R0, complete resection; R1/2, microscopic or gross residual disease.

Surgery

Forty-seven patients had surgery: complete resection (R0) in 20 patients (13 patients were considered by our pathologists to have an adequate margin [≥ 1 cm], seven patients were considered to have a close margin [<1 cm]), incomplete (microscopic or gross residual disease; R1/2) surgery in 27 patients, and seven patients had biopsy only.

Radiotherapy

Forty-five (83.4%) patients were treated at KFSH with radiotherapy: 38 (84%) patients received postoperative radiotherapy, three (11%) patients were treated with preoperative radiotherapy and four (8.8%) patients were treated with radiotherapy alone. The radiotherapy dose ranged between 45 and 60 Gy, depending on the tumour volume, with an overall median dose of 50.4 Gy. Most patients (35 patients; 77.7%) were planned conventionally on the simulator using two-dimensional techniques, as most patients presented early in a period where computed tomography planning was not yet implemented as routine practice in our department. All treatment was carried out on linear accelerators, energy ranged between 6 and 18 MV, and/or electron beam, depending on tumour depth and location.

The margin around the tumour ranged from 3 to 5 cm in all directions; joints were spared whenever appropriate. Irradiation of the entire circumference of an extremity was avoided, leaving a strip of normal tissue to avoid lymphoedema. Radiotherapy treatment compliance was good, with $\geq 90\%$ of patients completing their treatment without interruption.

Three patients did not receive any form of treatment apart from biopsy; they had a very short follow-up, but were still included in the final analysis.

Follow-up Schedule

All patients were seen 2 months after the end of their treatment, then every 3 months for the first 2 years, every 6