Clinical Oncology xxx (2018) 1–4

Contents lists available at ScienceDirect

Clinical Oncology

journal homepage: www.clinicaloncologyonline.net

Original Article

Radiation Therapy as an Alternative Treatment for Desmoid Fibromatosis

A.M. Hong *†, D. Jones *, R. Boyle ‡, P. Stalley ‡

* Central Clinical School, Faculty of Medicine and Health, The University of Sydney, Sydney, Australia
† Department of Radiation Oncology, Chris O'Brien Lifehouse, Camperdown, Australia
‡ Department of Orthopaedic Surgery, Royal Prince Alfred Hospital, Camperdown, Australia

Received 17 February 2018; received in revised form 11 April 2018; accepted 9 May 2018

Abstract

Aim: To determine the outcome after radiation therapy for desmoid fibromatosis.
Materials and methods: A retrospective review of 50 patients treated between 1988 and 2016 in a specialised bone and soft tissue tumour clinic.
Results: The median age at the time of radiation therapy was 36.8 years (range 15.1–69.0) and the median follow-up time was 51 months. Forty-three patients underwent radiation therapy as the definitive treatment with a median dose of 56 Gy (range 30–58.8 Gy). The median dose for the seven patients treated with postoperative radiation therapy was 50.4 Gy (range 48–56 Gy). Eleven patients (22%) developed progressive disease after radiation therapy at a median time of 41 months (range 12–113 months). The recurrences were within the radiation therapy field in four patients and outside the field in seven patients. One patient developed a radiation-induced malignancy 20 years after treatment.
Conclusions: Radiation therapy is an alternative treatment in the management of desmoid fibromatosis. It should be considered in patients for whom surgical resection is not feasible, or as adjuvant therapy after surgery with involved margins where any further recurrences would cause significant morbidity.

Keywords: Desmoid tumour; fibromatosis; radiation therapy; soft tissue tumour

Introduction

The optimal treatment of desmoid fibromatosis is still evolving, with National Comprehensive Cancer Network guidelines recommending a careful 'wait and see' approach if the tumour is asymptomatic and not located in an area that could lead to functional limitations if the tumour increases in size [1]. Spontaneous regression has been reported in 4–28% of patients, with the highest rate reported in abdominal wall locations [2,3]. If progression occurs, options for treatment include surgery, radiation therapy and systemic therapy [1]. Radiation therapy is a treatment option in progressing desmoid fibromatosis that is not amenable to surgery or when surgery might lead to an unacceptable functional outcome [4,5].

The aims of this study were to describe the characteristics of a large series of patients with desmoid fibromatosis treated with radiation therapy and to report the long-term outcomes.

Materials and Methods

Patients

All consecutive patients with desmoid fibromatosis treated with radiation therapy between 1988 and 2016 were included in this study. The study was approved by the ethics committees of the Sydney Local Area Health Services. Clinicopathological details and follow-up details were retrieved from the medical records up to December 2017. The baseline and follow-up magnetic resonance images (MRI) of the region were reviewed to assess the response. Patients were in general followed clinically with a MRI every 6 months for 3 years, then yearly thereafter.

Author for correspondence: A.M. Hong, Room 255, RMC Gunn Building, Central Clinical School, The University of Sydney, Sydney, NSW 2006, Australia.
E-mail address: angela.hong@sydney.edu.au (A.M. Hong).

https://doi.org/10.1016/j.clon.2018.05.002

Outcome Analysis

Follow-up time was calculated from the date of completion of radiation therapy to the date of last follow-up. The local control rate was calculated from the date of completion of radiation therapy until documented local recurrence or last follow-up. Local control was defined as stable disease or any regression of tumour mass in patients where disease was present at the time of radiation therapy, and as no recurrence in patients who received postoperative radiation therapy where gross disease was not present at the time of radiation therapy. Regression of disease was determined by an objective decrease in size on follow-up imaging and/or by clinical examination during follow-up.

Results

Patient Characteristics

In total, 50 patients treated with radiation therapy between 1988 and 2016 were included in the study (Table 1). All patients were managed through a multidisciplinary team (MDT) clinic with input from surgical oncology, radiation oncology and medical oncology. In general, the MDT would recommend observation for asymptomatic lesions. For patients with symptomatic disease, surgery is the treatment of choice followed by medical therapy. Radiation therapy is recommended for those lesions that are inoperable. Pathology and radiology were reviewed by expert pathologists and radiologists.

Table 1
Patient demographics and treatment details before radiation therapy

Total number	50
Median age at diagnosis (range)	35.2 years (13.6–68.7)
Gender	
Male	26 (52.0%)
Female	24 (48.0%)
Median duration of symptoms before initial diagnosis (range)	12 months (1–120 months)
Number of surgeries before radiation therapy	
0	25 (50.0%)
1	20 (40.0%)
2	4 (8.0%)
3	1 (2.0%)
Medical therapy before radiation therapy	
Yes	7 (14.0%)
No	43 (86.0%)
Total number of therapies before radiation therapy	
0	22 (44.0%)
1	18 (36.0%)
2	9 (18.0%)
3	1 (2.0%)
Timing of radiation therapy	
Definitive	43 (86.0%)
Postoperative	7 (14.0%)
Mean radiation therapy dose (range)	56 Gy (30–58.8 Gy)
Progression after radiation therapy (any time during follow-up period)	
Yes	11 (22.0%)
No	39 (78.0%)

The median age at diagnosis of desmoid fibromatosis was 35.2 years (range 13.6–68.7) and the median age at the time of radiation therapy was 36.8 years (range 15.1–69.0). Twenty-six patients (52%) were men. The median duration of symptoms was 12 months (range 1–120 months) before diagnosis. The most common locations were lower neck/shoulder region in 17 patients (34%) followed by lower limb in 14 patients (28%), chest wall in seven patients (14%), abdominal wall in four patients (8%), hip/buttock region in four patients (8%) and forearm in four patients (8%). One patient with a chest wall desmoid was diagnosed during pregnancy and she was initially observed. The mass continued to grow for 12 months after the pregnancy.

Treatment Details

After histological confirmation of the diagnosis of desmoid fibromatosis, the general treatment approach was observation with MRI monitoring for asymptomatic lesions. For those where treatment was recommended, wide local resection was the preferred option if clear margins could be achieved without significant impact on functional outcome. For those where wide resection was not recommended, the MDT would decide on medical therapy or radiation therapy. Twenty-two patients (44%) had radiation therapy as their only treatment. Eighteen patients (36%) had one line of therapy before radiation therapy, nine patients (18%) had two lines of therapy and one patient had three lines of therapy before receiving radiation therapy. Twenty-five patients (50%) received radiation therapy for recurrences after previous surgery. In these 25 patients, 20 had one resection, four patients had two resections and one patient had three resections before radiation therapy. Seven patients (14%) had medical therapy (six had hormonal therapy and one had both hormonal therapy and adriamycin) before radiation therapy. The hormonal therapy consisted of goserelin injection in three patients and tamoxifen in four patients.

All patients were treated with megavoltage photon and 10 patients were treated with volumetric modulated arc therapy (VMAT). The median clinical target volume was 264.4 cm^3 (range 76.94–1702.41). Forty-three patients (86%) underwent radiation therapy as the definitive treatment; the median radiation therapy dose was 56 Gy (range 30–58.8 Gy). The median dose for the seven patients who received postoperative radiation therapy was 50.4 Gy (range 48–56 Gy).

Outcomes

The medium follow-up time was 51 months (range 7–330 months) from the time of diagnosis and 43 months (range 1–308 months) from the time of completion of radiation therapy. Eleven patients (22%) developed

Please cite this article in press as: Hong AM, et al., Radiation Therapy as an Alternative Treatment for Desmoid Fibromatosis, Clinical Oncology (2018), https://doi.org/10.1016/j.clon.2018.05.002