

# Desmoid tumour Treatment Outcome: A single institution experience

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## Abstract

Desmoid tumour (DT) is a rare locally aggressive disease with tendency for local recurrence and the peak incidence age of 30-40 years.

DT prognostic factors are still conflicting like surgical margin, disease location, age and gender.

The various treatment options including surgery, radiotherapy and systemic agents are generally offered according to the disease presenting symptoms and progressiveness.

**Keywords:** desmoid tumour; surgical margin; radiotherapy

## Background

Desmoid tumour (DT) is a rare locally aggressive disease with tendency for local recurrence [1] and the peak incidence age of 30-40 years [2]

DT prognostic factors are still conflicting like surgical margin [3,4,5-10], disease location [3,5,9,11], age and gender [5,8,12,13].

The various treatment options including surgery, radiotherapy and systemic agents are generally offered according to the disease presenting symptoms and progressiveness [14-20].

Complete surgical resection without causing significant morbidity can be challenging. The administration of adjuvant radiotherapy may improve local control but can cause side effects like fibrosis and wound healing problems [3, 4, 21, 22], rarely there is concerns about development of radiation induced second malignancies.

The aim of this work is to present our experience using multidisciplinary team to reach the best treatment outcome for this locally aggressive disease and report the long-term outcomes of treatment.

## Patients and Methods

The medical records of total of 72 consecutive patients diagnosed with DT over a period of 10 years between January 2009 and December 2018 were reviewed at Radiation Oncology section, King Faisal Specialist

Hospital & Research Centre (KFSH&RC), Riyadh, Saudi Arabia. All the patients had biopsy proven DT at our institution in addition to baseline and periodic follow up CT and/or MRI scans.

Radiotherapy was delivered as definitive, preoperative or postoperative treatment. Radiation doses were 50 - 66 Gy in 2 Gy per fraction as preoperative, postoperative and definitive radiotherapy. Radiation energy was 4-6MeV delivered by either 3D-Conformal Radiation Therapy, Tomotherapy or Volumetric Modulated Arc Therapy techniques. Surgery was either wide local excision or intralesional debulking.

## Statistical analysis

Continuous data were checked for normality by using Shapiro Walk test. Kruskal Wallis H test was used to compare more than two groups of non-normally distributed data. Percent of categorical variables were compared using Pearson's Chi-square test or Fisher's exact test when was appropriate. Stratification of Recurrence Free Survival was done according intention to treat (ITT). These time-to-event distributions were estimated using the method of Kaplan-Meier plot, and compared using two-sided exact log-rank test. All tests were two sided. All statistics were performed using SPSS 22.0 for windows (IBM Inc., Chicago, IL, USA) and Med Calc 13 for windows (Med Calc Software bvba, Ostend, Belgium).

## Results

Parameters	All patients (N=69)		Parameters	All patients (N=69)	
	No.	%		No.	%
Sex			Radiotherapy (RT)		
Male	21	30.4%	No	15	21.7%
Female	48	69.6%	Yes	54	78.3%
Age (years)			Surgery (S)		
Median (Range)	27 (15 – 66)		No	24	34.8%
≤30 years	43	62.3%	Yes	45	65.2%
>30 years	26	37.7%	Surgical margin (N=45)		
Site of tumor			Negative	15	33.3%
			Positive	30	66.7%
Upper limb	15	21.7%	RT dose (N=54)		
			50Gy-25fraction	39	72.2%
Trunk	21	30.4%	60Gy-30fraction	12	22.2%
Pelvis	15	21.7%	66Gy-33fraction	3	5.6%
Lower limb	18	26.1%	RT timing (N=30)		
Size of tumor (cm)			Preoperative	12	40%
			Postoperative	18	60%
Median (Range)	7 (3 – 25)		Response to ttt (N=54)		
≤5 cm	20	29%	Complete Response	24	44.4%
>5 cm	49	71%	Partial Response	18	33.3%
Sequence			Stable Disease	12	22.2%
Surgery alone	15	21.7%	Recurrence (N=45)		
RT alone	24	34.8%	Negative	39	86.7%
RT then Surgery	12	17.4%	Positive	6	13.3%
Surgery then RT	18	26.1%			

**Table 1:** Basic characteristics and outcome of 69 patients with desmoid tumor.

### Patient characteristics

Two patients opted to undergo observation only and 1 patient opted to receive systemic treatment with Sorafenib who were excluded from the study. Therefore, total of 69 patients were evaluated (Table 1).

The median age of patient was 27 years (range 15-66) and approximately two-thirds were females. The majority were younger patients aged less than 30 years (n=43, 62%).

### Tumour characteristics

The median size was 7 cm (range, 3-25cm) and nearly two third of the patients had tumours larger than 5 cm. The most common site was trunk (n=21, 30%), followed by lower limbs (n=18, 26%), pelvis (n=15, 22%), and upper limbs (n=15, 22%).

### Treatment characteristics

Twenty four patients were treated with definitive radiotherapy, 15 patients by surgical resection alone and 30 patients by combination of both surgery and radiotherapy (12 cases had preoperative and 18 cases had postoperative irradiation).

The surgical margin was positive in 30 patients out of total of 45 cases who had surgery (67%). Radiation was given to total of 54 cases with doses of 50 Gy in 25 fractions delivered in 39 cases (72%) and 60- 66 Gy in 15 cases (28%) (Table 2).

### Response to treatment

The median follow up was 5.5 years (range 1- 10yrs). The local control in patients who had radiotherapy (n=54 pts) as part of their treatment was 89%. The patients who had surgery (n=45 pts) as a part of the treatment had local control of 87% (Table 2).

### Surgery alone group

The patients who had surgery (n=15) alone had no recurrence.

### Radiotherapy alone group

The local control in patients who had definite radiotherapy (n=24) was 100% with partial response in 18 pts (75%) and stable disease in 6 pts (25%). All the patients who had definite radiotherapy had size greater than 5cm.

### Combined Surgery and radiotherapy group

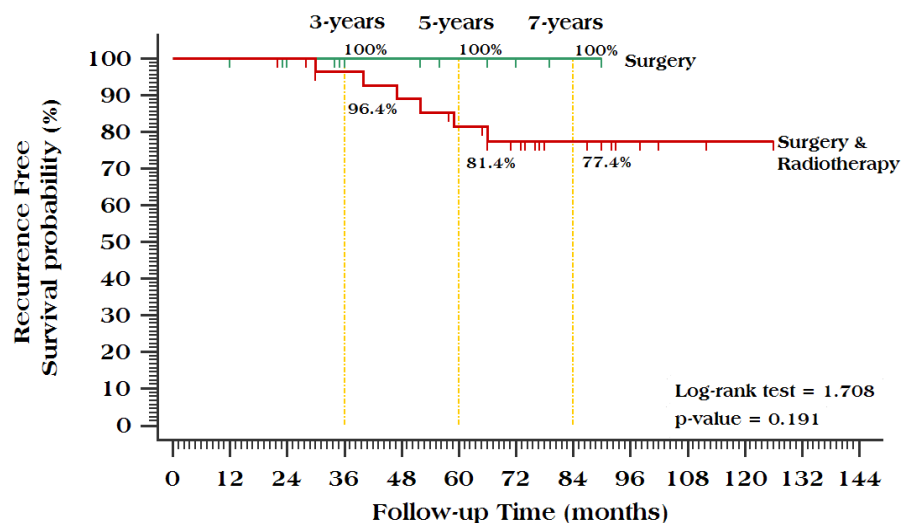
All the patients who had combined surgery and radiotherapy (either pre or post-operative) had positive margins. The local control in patients with positive surgical margin (n=30 pts) and radiotherapy was 80% (n=6 patients developed recurrence). All the recurrences happened in the lower radiotherapy dose of 50 Gy and those who had post operative radiotherapy. There was no recurrence in the higher radiotherapy dose of 60 Gy (p= 0.05) and those who had pre operative radiotherapy (p=0.05). The patients group treated with surgery plus radiotherapy most of the recurrence was in patients with tumour size less than 5 cm (5 out of total of 6 recurrent patients). There was no recurrence in the patients with trunk site and all the recurrences happened in the extremities group. All the recurrences were in the less than 30 years group (Table 2).

### Survival and long term side effects

The 3yrs, 5yrs and 7yrs recurrence free survival in patients who had both surgery and radiotherapy was 96.4%, 81.4% and 77.4% respectively. There was no grade 3 or 4 acute or late toxicity in all the groups of patients. There was no incidence of second malignancy in this cohort of patients at median follow up of 5.5 years (Figure 1).

Parameters	Surgery (N=15)			Radiotherapy (N=24)			Surgery + RT (N=30)		p-value
	No.	%		No.	%		No.	%	
Sex									
Male	3	20%		12	50%		6	20%	0.036‡
Female	12	80%		12	50%		24	80%	
Age (years)									
Median (Range)	33 (19 – 42)			31 (15 – 66)			25 (16 – 42)		0.091•
≤30 years	7		46.7%			12	50%		
>30 years	6	53.3%		12	50%		24	80%	0.029‡
Site of tumor							6	20%	
Upper limb	0	0%		6	25%				<0.001‡
Trunk	9	60%		0	0%		9	30%	
Pelvis	3	20%		9	37.5%		12	40%	
Lower limb	3	20%		9	37.5%		3	10%	
Size of tumor (cm)							6	20%	
Median (Range)	6 (4 – 10)			9 (6 – 25)			6 (3 – 11)		
≤5 cm	6		40%			0	0%		
>5 cm	9		60%			24	100%		
							14	46.7%	<0.001‡
Response to ttt							16	53.3%	
CR				0	0%				
PR				18	75%				<0.001‡
SD				6	25%		24	80%	
PD				0	0%		0	0%	
Recurrence							0	0%	<0.001‡
Absent	15	100%					6	20%	
Present	0	0%							
RFS							24	80%	0.157‡
3-year RFS	100%				96.4%		6	20%	
5-year RFS	100%				81.4%	0.191†			
7-year RFS	100%				77.4%	0.191†			0.191†
									0.191†

**Table 2:** Comparison between the studied groups regarding basic characteristics and outcome. RFS recurrence free survival, ‡ Chi-square test; • Kruskal Wallis H test; † Log-rank test; p-value<0.05 is significant.



**Figure 1:** Kaplan Meier plot shows comparison between the studied groups regarding Recurrence Free Survival.

## Discussion

Desmoid tumour is benign tumour of connective tissue but with high chances of local recurrence. Surgery is the main treatment modality of therapy if it can be done with acceptable morbidity. Radiotherapy is given in locally advanced inoperable tumours as definitive therapy or in combination with surgery in high risk tumours. The radiation dose used is 50-66 Gy depending on the tolerance dose to the surrounding organs of risk. The clinical course can be variable and rarely may undergo spontaneous regression. Therefore, close observation to monitor the rate of growth is a reasonable option if patient is minimally symptomatic or even when the surgical margins are positive when resection is possible without significant morbidity.

In our cohort regarding to gender and age, females have aggressive disease similar to studies reported by [5, 8, 12 & 13] while age 30 years or less has poor prognosis as in series by [5, 8 & 12] while aggressive in age more than 30 years in series by Rock et al [11] and less than 32 years by Sorensen et al [13].

In our study regarding to site, the extremities have a worse disease while the trunk has a more favorable outcome similar to Ballo et al [3], Broekhoven et al [8], Huang [9] and Rock et al [11] while non-significant in Merchant et al [5].

Our cohort showed recurrence free survival (RFS) of 96.4%, 81.4% and 77.4% at 3, 5 and 7 years of the group treated by surgery plus radiotherapy while Merchant et al [5] noticed the 2-year and 5-year local RFS rates of 80% and 75%, respectively, Zeng et al [10] had the estimated 5- and 10-year RFS of 74.2 % and 70.7 % respectively while Sorensen [13] showed the 5 year local RFS rate of 73%.

local control in our cohort received irradiation as a part of treatment was 89% and 87% if surgery was a part of treatment, while Goy et al [4] noted the 6-year actuarial local control 78% (+/-14%) in cases managed by surgery and irradiation.

In our cohort, patients (n=30) with perioperative irradiation had positive margin with local control of 80% and local recurrence in 20% (n=6), while Ballo et al [3] reported the 10-year actuarial relapse rate of 25%.

Our study showed that patients who had surgery alone with negative surgical margin did not have local recurrence with median 5.5 years follow up similar to results by Goy et al [4], Zeng et al [10] and Rock et al [11], contrary to Merchant et al [5], Broekhoven et al [8], Huang et al [9] and Shido et al [12].

Our cohort showed that the rate of disease recurrence is significant in lesion less than 5cm, extremities site, radiation dose 50Gy, age 30 years

or less & patients received postoperative not preoperative irradiation, Contrary to Merchant et al [5] as all prognostic factors were insignificant for predicting local recurrence even positive margin, while Broekhoven et al [8] noted the risk of recurrence increased for extremity lesions, however insignificant of age, resection margins or adjuvant radiotherapy. Huang et al [9] noted that tumor site, size, and resection margin had significant impacts on event-free survival. Zeng et al [10] reported worse RFS in tumor size larger than 5 cm, extra-abdominal tumor sites and resection margin status. Rock et al [11] reported recurrence was evident in females, in patients more than thirty years old, in lower extremity locations and most importantly after treatment by intralesional or marginal excision. Shido et al [12] found that tumor size, surgical margin and previous surgical history were not associated with local recurrence, however younger age <30 years and females had local recurrence risk, suggesting the tumor effect biological behavior.

## Conclusions

Radiotherapy plays an important role in local control of DT. The factors associated with poor response in the cohort of patients are younger age group, lower dose of radiation and post-operative radiotherapy compared to preoperative radiotherapy. The majority of the recurrent tumours were small indicating that biology of the disease is more important.

## Declarations

1. Ethics approval and consent to participate: For retrospective review of data with less than minimal risk to the patients, no consent was required by the ethics committee.
2. Consent for publication: Not applicable.
3. Availability of data and materials: The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.
4. Competing interests: All authors declare that there is no conflict of interest.
5. Funding: Not applicable.
6. Authors' contributions: First author collect the data, write the manuscript while second author analysed the data and shared in writing the manuscript.
7. Acknowledgements: Not applicable.

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