ORIGINAL RESEARCH

A retrospective study describing desmoid tumour characteristics and management at a private, nonprofit physical rehabilitation facility in Kampala, Uganda

Miriam Nyeko-Lacek[1], Andrew Hodges[2]

[1]School of Medicine, Cardiff University, United Kingdom
[2]CoRSU Rehabilitation Hospital, Kampala, Uganda

Correspondence: Dr Miriam Nyeko-Lacek (mnyekolacek@gmail.com)

East Cent Afr J Surg. 2021;26(1):36-40
https://doi.org/10.4314/ecajs.v26i1.6

Abstract

Background
Desmoid tumours are rare fibroblastic tumours. They have a high rate of recurrence and can be locally destructive. Comprehensive Rehabilitation Services in Uganda (CoRSU) runs CoRSU Rehabilitation Hospital in Kampala, Uganda, where the encountered desmoid tumours are often managed surgically. This study aimed to describe the demographics and management of patients presenting to CoRSU with desmoid tumours.

Methods
The CoRSU database was searched for any patients with diagnoses of "desmoid tumour" or "fibromatosis". Histopathology reports were assessed to confirm diagnoses and patient eligibility. Descriptive statistics were calculated using the demographic and clinical data of the eligible patients.

Results
The initial database query returned 19 records of patients diagnosed with desmoid tumours. Two patients were excluded due to a lack of histopathologic data confirming their diagnoses, and a further 3 records had histopathologic findings that contradicted a desmoid tumour diagnosis. Fourteen patients with suitable histopathologic reports were included in the analysis: 6 males and 8 females, with a median age of 12 years (range, 5-35 years) at diagnosis. The most common tumour sites were the extremities, limb girdles, and chest wall. Three patients had at least 1 recurrence, each of whom had a primary tumour in the extremities or limb girdles.

Conclusions
In our small retrospective series, we observed that the demographic characteristics of desmoid tumour patients managed at CoRSU Rehabilitation Hospital were similar to those reported for desmoid tumour patients elsewhere in the world. Younger patients more commonly developed extra-abdominal tumours, and abdominal tumours only occurred among patients aged 15 years or older. Most patients were managed surgically, in line with current recommendations.

Keywords: desmoid tumour, plastic surgery, fibromatosis, tumour recurrence, Uganda

Introduction

Desmoid tumours are rare benign fibroblastic tumours. They comprise about 0.03% of all malignancies, with an incidence of about 3 to 4 cases per 1 million people in the United States.[1] They do not metastasize but can be locally aggressive. Desmoid tumours can recur, making management difficult, especially when they recur in a functionally important site. They are often managed surgically, but current evidence suggests that they may be managed using radiotherapy and chemotherapy, either alone or as adjuncts to surgery.[2]-[4] They tend to occur in younger patients, with a peak incidence around 30 years of age.[5],[6] There is a relatively high desmoid tumour incidence among women of childbearing age, suggesting a possible association with oestrogen.[7] Desmoid tumours have been linked to genetic syndromes, such as familial adenomatous polyposis syndrome and Gardner syndrome.[7],[8] They can occur anywhere in the body, although the most common sites among

Table 1. Demographic and clinical characteristics of patients with desmoid tumours managed at a private rehabilitation hospital in Kampala, Uganda (N=14)

Characteristic	n (%)
Age, years	
≤10	5 (36)
10≤15	5 (36)
15≤20	1 (7)
20≤25	1 (7)
25≤30	1 (7)
≤30	1 (7)
Management	
Surgery only	10 (71)
Surgery and chemotherapy	2 (14)
Tamoxifen and chemotherapy	1 (7)
Palliative care	1 (7)
Number of operations	
0	3 (21)
1	8 (57)
2	2 (14)
3	0 (0)
4	1 (7)
Tumour site	
Abdominal wall	2 (14)
Retroperitoneal/pelvic	0 (0)
Extremities/girdles	11 (79)
Head & neck/intrathoracic	1 (7)
Intra-abdominal	0 (0)

children tend to be the limbs and limb girdles, while among adults, the tumours are more frequently in the abdominal region.[8] They can occur at sites of previous trauma, including surgical sites.

Patients with desmoid tumours most commonly present with a palpable mass or a painful mass, and tumours are often described as hard, fibrous lumps. Associated local lymphadenopathy is more characteristic of malignant sarcomas, such as dermatofibrosarcoma protuberans.[7] Magnetic resonance imaging (MRI) is the most effective imaging modality for diagnosing desmoid tumours.[3] A definitive diagnosis can be made via biopsy, such as core or excisional biopsy. Histologically, desmoid tumours can grossly be described as "fibroblastic spindle cell proliferation with wavy nuclei"[9] and can be similar to fibrosarcomas or myxofibrosarcomas, which also have an infiltrative pattern.[9] This highlights the importance of sending large, high-quality samples for histopathologic examination (as a small tumour specimen may be indistinguishable from other tumour types), as well as the importance of full, detailed histopathology reports.

Methods

Data were collected from the patient database at a specialized surgical facility in Kampala, Uganda, run by Comprehensive Rehabilitation Services in Uganda (CoRSU). We searched the database at CoRSU Rehabilitation Hospital using the queries "desmoid tumour" and "fibromatosis". Histopathology reports were then assessed to confirm diagnoses and patient eligibility. Biopsies for the patients included in this study were processed and evaluated by a private histopathology laboratory (Surgpath Medical Consultants, Kampala, Uganda). For the majority of patients, there were 2 reports: the first reporting the results of an initial smaller (core) biopsy examination and the second reporting an excisional biopsy result. The second report was weighted more strongly when determining patient eligibility, as desmoid tumours are known to be confused with other tumour types when only a small sample is assessed.

Descriptive statistics were calculated using the demographic and clinical data of the eligible patients. Results are presented as frequencies and percentages.

Results

The initial database search returned 19 patient records reporting desmoid tumour diagnoses. Three histopathology reports indicated diagnoses (fibromyxoid sarcoma, neurofibroma, and keloid) other than desmoid tumours, and these records were excluded from the analysis. Two patients were excluded due to a lack of histopathologic data confirming their desmoid tumour diagnoses.

The 14 patients included in the study were all treated at CoRSU Rehabilitation Hospital between 2010 and 2019. There were 6 males and 8 females, with a median age of 12 years (range, 5-35 years; mean, 14 years). Most of the patients were managed surgically, with *surgery only* (n=10, 71%) being the most common treatment (Table 1). Two patients (14%) underwent surgery and chemotherapy, 1 (7%) was treated with tamoxifen and chemotherapy, and another patient was managed palliatively. Tumour resections were carried out with wide surgical margins. Most patients had a single operation, but 2 patients each had 2 operations, and 1 underwent 4 operations because of tumour recurrences.

The most common tumour sites were the extremities and limb girdles. Specifically, there were 3 extremity tumours and 8 limb-girdle tumours. There were 2 abdominal wall tumours, and there was 1 tumour of the posterior neck.

Tumour location by age

There was a higher prevalence of extra-abdominal tumours than abdominal tumours among patients under 15 years of age (Table 2). Among patients older than 15 years, abdominal and extra-abdominal tumours were equally common. Tumours defined as "abdominal" included both abdominal

Table 2. Frequency of extra-abdominal and abdominal tumours by sex and recurrence status among patients with desmoid tumours managed at a private rehabilitation hospital in Kampala, Uganda (N=14)

Variable	n (%)	
	Extra-abdominal	Abdominal
Males		
<15 years old	4 (29)	0 (0)
≥15 years old	2 (14)	0 (0)
Females		
<15 years old	6 (43)	0 (0)
≥15 years old	0 (0)	2 (14)
Recurrence		
Yes	3 (21)	0 (0)
No	9 (64)	2 (14)

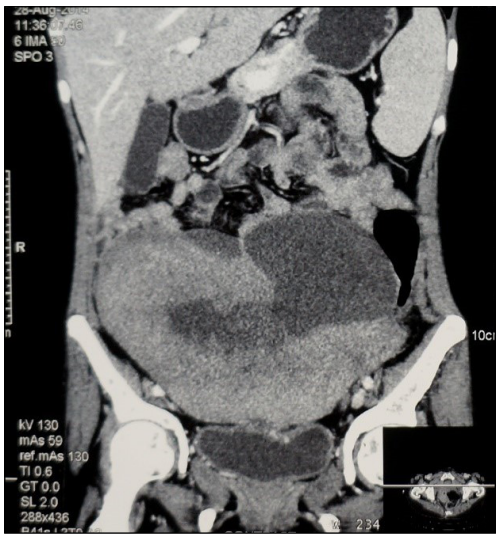

Figure 1. Computed tomography image of desmoid tumour on the abdominal wall of a female patient

This tumour was later successfully resected.

wall tumours and intra-abdominal tumours. Extra-abdominal tumours include those involving the extremities, the limb girdles, head, neck, and thoracic wall.

Clinical appearance

Clinically, the tumours were firm, smooth, and often adherent to local structures. In some cases, they were large enough to restrict or alter movement in areas such as the gluteal region or shoulder girdle. They did not always present with pain; however, in certain cases, nerves had clearly been affected. Some tumours were associated with altered sensation, including tingling or numbness. One patient developed a foot drop resulting from compression of the sciatic nerve.

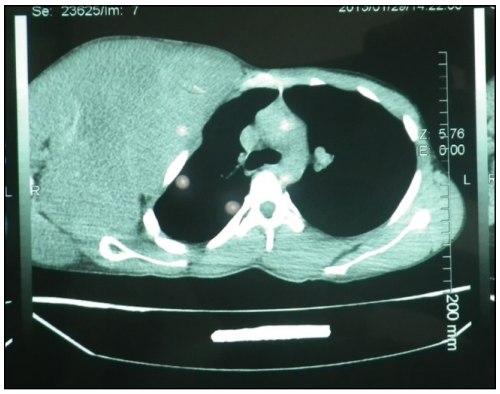

Figure 2. Computed tomography image of a shoulder-girdle desmoid tumour of a young male patient

This tumour was considered inoperable because it had invaded the pleural cavity.

Imaging

Most of the patients (n=12, 86%) underwent computed tomography (CT) imaging (Figure 1, Figure 2). The 1 patient (7%) who underwent MRI was a young female with a right forearm tumour that was managed surgically.

Follow-up and recurrences

Among the patients who underwent operative management, 3 (21%) developed recurrences within a mean follow-up duration of 255 days (range, 0-615 days) from their initial consultations at CoRSU Rehabilitation Hospital. The median age of patients who experienced tumour recurrence was 12 years, and the median interval until a first recurrence was 567 postoperative days (range, 176-1461 days), with index operations performed either at CoRSU Rehabilitation Hospital or elsewhere before patients presented to CoRSU.

Discussion

In this retrospective study, the majority of patients were managed surgically. This is in line with the European Organisation for Research and Treatment of Cancer/Soft Tissue and Bone Sarcoma Group initiative, which recommends that symptomatic patients with operable tumour masses should be offered surgery.[3] Radical resection achieving negative margins offers the best chance of complete remission. However, research suggests that adjuvant radiotherapy, given after surgery in cases of recurrent tumours, can reduce the chance of further recurrence.

A similar retrospective review was conducted at Groote Schuur Hospital in Cape Town, South Africa, including all desmoid tumours (N=70) encountered at the hospital between 2003 and 2016.[10] That study, conducted at a large tertiary facility, had a larger sample size than ours, as well as an older overall median age (36.5 years at diagnosis vs 12 years in our study) and an older mean age (25 years vs 11 years in our study) among patients who experienced tumour recurrence.

As with most soft tissue tumours, CT and MRI are recommended to provide appropriate preoperative assessments for patients with desmoid tumours. MRI can give surgeons a more detailed picture of the surgical prognosis. On CT, desmoid tumours can appear sharply defined, as is usually the case with abdominal wall tumours, or with less well-defined margins, as is often seen in association with extra-abdominal tumours. On contrast-enhanced scans, desmoid tumours are usually isodense or hyperdense relative to adjacent muscle. Where MRI is available, different signal intensities can indicate different phases of the tumour lifecycle.[11] However, for most of the patients presenting to CoRSU, MRI is prohibitively expensive.

As is best practice, tumour resections for the patients at CoRSU were carried out with wide surgical margins. This can be challenging due to the locations of adjacent structures, particularly with extra-abdominal tumours in areas such as the limb girdles. Here, removing the tumour has to be balanced against the possibility of inducing disability, for example, by resecting neurovascular bundles which have been invaded by the tumour. The literature suggests that abdominal wall tumours may sometimes be more straightforward to remove[8] and, thus, are less likely to recur than limb tumours.

High-dose tamoxifen (in combination with sulindac) has been associated with limited toxicity; however, 1 study found that desmoid tumours responded minimally to the combination and that tamoxifen plus sulindac had little effect on progression-free survival.[12] One of the patients seen at CoRSU for assessment was previously given tamoxifen at another centre alongside 4 cycles of chemotherapy for an inoperable tumour. Some success was achieved in shrinking the tumour, but the patient never reached full remission with this treatment. Some authors have suggested nonsteroidal anti-inflammatory drugs as a treatment for desmoid tumours, although there is debate about their effectiveness.[9],[13] Four of the patients at CoRSU were given nonsteroidal anti-inflammatory medication—either meloxicam or lornoxicam—with limited effect, and 3 of these 4 patients experienced tumour recurrence.

Despite guidelines largely recommending surgical management of resectable tumours, there is some limited evidence supporting the use of chemotherapy.[3],[5] One published case report describes a young Rwandan child who received chemotherapy before surgery, which helped reduce the size of a large and aggressive facial tumour to allow for resection.[14] This patient appears to have done remarkably well, although it is not clear from the report whether he developed a recurrence later down the line. Neither of the patients in our study who received chemotherapy before surgery developed recurrences.

This study was limited by its small sample size as well as the loss of patients to follow-up. However, there are still lessons to be learned from this review. The practice of sending the entire tumour specimen for histopathologic examination after resection helps rule out malignancy. Surgical management for growing but resectable tumours has been beneficial when wide surgical margins are feasible.

Conclusions

Desmoid tumours, although benign, can cause significant morbidity and mortality due to their infiltrative nature. Extra-abdominal tumours, particularly limb tumours, are more likely to recur and are more likely to occur in children. Abdominal tumours, on the other hand, are more likely to occur in women of childbearing age and are less likely to recur. It is important to send high-quality, adequately sized specimens for histopathologic examination and to send the fully excised tumour for analysis, as desmoid tumours can be misdiagnosed as other tumours with similar appearances and vice versa. Surgical management is the first-line treatment if the tumour is resectable and can be done for debulking purposes as an adjunct to other treatments. Radiotherapy has been described as a good adjunct to surgery for recurrent tumours, and it can be used to treat inoperable tumours.

The management of these tumours at CoRSU Rehabilitation Hospital has followed best-practice guidelines as much as possible, given some limitations, such as resources for assessment, most notably MRI. In the future, more patients will likely be encouraged to undergo adjuvant treatments after surgery, such as radiotherapy, as studies have shown that this may reduce the number of patients undergoing multiple operations for these rare tumours.

Acknowledgements: Thanks to Surgpath Medical Consultants, Kampala, Uganda.

References

1. Kasper B, Ströbel P, Hohenberger P. Desmoid tumors: clinical features and treatment options for advanced disease. *Oncologist*. 2011;16(5):682-693. doi:10.1634/theoncologist.2010-0281 [View Article] [PubMed]
2. Constantinidou A, Jones RL, Scurr M, Al-Muderis O, Judson I. Pegylated liposomal doxorubicin, an effective, well-tolerated treatment for refractory aggressive fibromatosis. *Eur J Cancer*. 2009;45(17):2930-2934. doi:10.1016/j.ejca.2009.08.016 [View Article] [PubMed]
3. Kasper B, Baumgarten C, Bonvalot S, et al; Desmoid Working Group. 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. *Eur J Cancer*. 2015;51(2):127-136. doi:10.1016/j.ejca.2014.11.005 [View Article] [PubMed]
4. Seinen JM, Niebling MG, Bastiaannet E, Pras B, Hoekstra HJ. Four different treatment strategies in aggressive fibromatosis: A systematic review. *Clin Transl Radiat Oncol*. 2018;12:1-7. doi:10.1016/j.ctro.2018.03.001 [View Article] [PubMed]
5. El-Haddad M, El-Sebaie M, Ahmad R, et al. Treatment of aggressive fibromatosis: the experience of a single institution. *Clin Oncol (R Coll Radiol)*. 2009;21(10):775-780. doi:10.1016/j.clon.2009.08.012 [View Article] [PubMed]
6. Wang YF, Guo W, Sun KK, et al. Postoperative recurrence of desmoid tumors: clinical and pathological perspectives. *World J Surg Oncol*. 2015;13(26):26. doi:10.1186/s12957-015-0450-8 [View Article] [PubMed]
7. Shields CJ, Winter DC, Kirwan WO, Redmond HP. Desmoid tumours. *Eur J Surg Oncol*. 2001;27(8):701-706. doi:10.1053/ejso.2001.1169 [View Article] [PubMed]

8. Otero S, Moskovic EC, Strauss DC, et al. Desmoid-type fibromatosis. *Clin Radiol*. 2015;70(9):1038-1045. doi:10.1016/j.crad.2015.04.015 [View Article] [PubMed]
9. Papagelopoulos PJ, Mavrogenis AF, Mitsiokapa EA, Papaparaskeva KT, Galanis EC, Soucacos PN. Current trends in the management of extra-abdominal desmoid tumours. *World J Surg Oncol*. 2006;4(21):21. doi:10.1186/1477-7819-4-21 [View Article] [PubMed]
10. Pickard HDP, Jacob N, Malherbe F, Panieri E, Naiker T, Cairncross L. The management of desmoid tumours at Groote Schuur Hospital: A retrospective review of current practice. *S Afr J Oncol*. 2019;3. doi:10.4102/sajo.v3i0.68 [View Article]
11. Braschi-Amirfarzan M, Keraliya AR, Krajewski KM, et al. Role of Imaging in Management of Desmoid-type Fibromatosis: A Primer for Radiologists. *Radiographics*. 2016;36(3):767-782. doi:10.1148/rg.2016150153 [View Article] [PubMed]
12. Skapek SX, Anderson JR, Hill DA, et al. Safety and efficacy of high-dose tamoxifen and sulindac for desmoid tumor in children: results of a Children's Oncology Group (COG) phase II study. *Pediatr Blood Cancer*. 2013;60(7):1108-1112. doi:10.1002/pbc.24457 [View Article] [PubMed]
13. Cho JY, Gupta S, Cho HS, et al. Role of nonsteroidal anti-inflammatory drug in treatment of extra-abdominal desmoid tumors. *Clin Orthop Surg*. 2018;10(2):225-233. doi:10.4055/cios.2018.10.2.225 [View Article] [PubMed]
14. Kanyamuhunga A, McCall N, Tuyisenge L, Mumena C, Stefan DC. Aggressive desmoid fibromatosis: first case in a Rwandan child. *SAJCH*. 2013;7(3):117-118. doi:10.7196/sajch.593 [View Article]

Peer Reviewed

Competing Interests: None declared

Received: 28 Jun 2019 • **Revised:** 9 Sep 2019, 17 Oct 2020

Accepted: 4 Dec 2020 • **Published Online**: 7 Jan 2021

Cite this article as: Nyeko-Lacek M, Hodges A. A retrospective study describing desmoid tumour characteristics and management at a private, nonprofit physical rehabilitation facility in Kampala, Uganda. *East Cent Afr J Surg*. 2021;26(1):36-40. doi:10.4314/ecajs.v26i1.6