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Research Paper

Desmoplastic fibroma of bone: A rare bone tumour

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ABSTRACT

We identified thirteen patients with desmoplastic fibroma of bone treated at our institute over a 30 year period. The patients had a mean age of 25.9 years; eight were female. The incidence of desmoplastic fibroma of bone in all patients with benign bone tumours in our population is 0.003%. Surgical treatment ranged from primary amputation to intra-lesional curettage. The incidence of local recurrence was 15.4%. All cases of local recurrence after curettage or marginal excision demonstrated soft-tissue extension of their tumours on initial presentation suggesting that extra-osseous extension requires more radical surgery to control the disease.

This study presents the largest single centre series of desmoplastic fibroma of bone with a mean follow-up time of 8 years. We recommend wide surgical excision, particularly if the lesion can be resected without significant loss of function, as the treatment modality of choice with the lowest rate of recurrence. Patients undergoing intra-lesional or marginal resection need to be advised of the possibility of local recurrence and the need for long-term surveillance.

1. Introduction

Desmoplastic fibroma (DF) is a rare, locally aggressive benign bone tumour with a reported incidence of 0.11% of all primary bone tumours [1,2]. It was initially described by Jaffe [3] in 1958 who highlighted the histological resemblance to aggressive fibromatosis (desmoid tumour). Numerous authors have subsequently attempted to clarify the histological criteria and, in 2013, the WHO described the microscopic appearance of desmoplastic fibroma as being composed of slender, spindle to stellate cells with minimal cytological atypia and abundant collagenous matrix [4]. Because of the infiltrative pattern of growth, desmoplastic fibroma was considered by some to be the osseous counterpart of extra-abdominal desmoid tumour [5], however, a recent study revealed that there were no mutations in exon 3 of CTNNB1, encoding for B-catenin, thus genetically distinguishing desmoplastic fibromas of bone from desmoid-type fibromatosis [6]. Local recurrence rates are between 37% and 72% [7–9] following operative intervention.

Patients present typically with pain and swelling of the affected area. Plain radiographs reveal a trabeculated, lucent, expansile lesion often with lobulated margins [10]. Cortical thinning with breakthrough and a soft-tissue mass may also be seen [7]. Establishing a diagnosis is difficult by imaging studies alone [2] as many tumours resemble desmoplastic fibroma. The differential diagnosis includes benign lesions, such as fibrous dysplasia, simple bone cyst, aneurysmal bone cyst, non-ossifying fibroma, eosinophilic granuloma and chondromyxofibroma [2,7]. If cortical destruction and a soft tissue mass is noted, desmoplastic fibroma of bone may resemble more sinister pathology, such as fibrosarcoma, intra-osseous osteosarcoma and metastases [2,7].

To date, small numbers of case reports and case series have been published regarding desmoplastic fibroma. The aim of this paper is to present the largest single-centre series of desmoplastic fibroma of bone with a view to outline a management strategy and to discuss the criteria associated with local recurrence.

2. Methods

We conducted a retrospective search of a prospective tumour database to identify all patients treated at our unit with a diagnosis of desmoplastic fibroma of bone. The diagnosis was confirmed following a multidisciplinary review of the patient's clinical history, pertinent radiological imaging and histological findings. Patient demographics were recorded along with the site of their disease and management, with local recurrence and the need for revision surgery being defined as end-points.

2.1. Statistical analysis

Simple summary statistics were collated using SPSS v 18.0 software (IBM Corp., Armonk, New York). Categorical data was

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compared using the chi-squared test or Fisher's exact test, with an alpha value of 0.05 considered statistically significant.

3. Results

A search of our database, which holds prospectively gathered data on over 30,000 patients including 4692 benign bone tumours, identified 13 patients with desmoplastic fibroma of bone, giving an incidence of 0.003% in our population. There were 8 females (61.5%) and 5 males (38.5%). Mean age at presentation with symptoms was 25.9 years (range 5–50 years). Skeletal distribution of the lesions is shown in Fig. 1.

All patients had been referred to our unit with suspicious plain radiographs following a history of pain/swelling of the affected body area and subsequently underwent magnetic resonance imaging and needle biopsy to confirm the diagnosis (Table 1).

Two patients (cases 1 and 7) declined any treatment following confirmation of their diagnosis and were discharged with advice to return if concerned.

6 patients underwent intra-lesional curettage. Of those patients, 2 (cases 8 and 13) developed rapid local recurrence at 9 and 7 months post-curettage respectively. Both cases had extra-osseous soft-tissue components to their tumours at initial curettage. Case 8 had intractable pain associated with an extensive local recurrence that necessitated a below knee amputation to control their disease 24 months after initial curettage. Case 13 presented with a distal radial desmoplastic fibroma that was initially treated with curettage alone because of the difficulty of resection and reconstruction. However, rapid local recurrence necessitated further curettage with marginal excision of the soft-tissue component. 12 months post-revision surgery, he has normal upper limb function with no symptoms of recurrence.

3 patients underwent a marginal excision. There have been no recurrent tumours reported with a median time since operation of 12.1 years (range 9.7–15.3).

One patient (case 6) underwent wide excision of their lesion, with endo-prosthetic replacement (EPR) of their proximal tibia. No local recurrence has been detected 15.1 years following their index operation, although revision EPR was required for aseptic component loosening of the tibial component after 11.3 years.

1 patient (case 4) elected to undergo a below knee amputation as limb salvage was not deemed possible due to the extensive intra and extra-osseous nature of their distal tibial lesion.

In total, local recurrence occurred in 2 (15.4%) patients in our series. There was a statistically significant association between local recurrence and the presence of an extra-osseous soft-tissue component ($p < 0.023$).

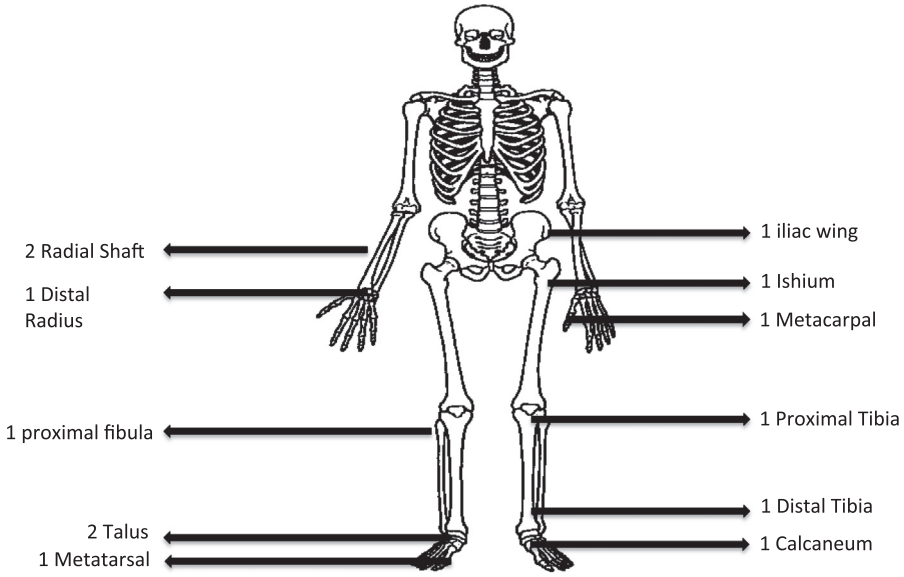

Fig. 1. Skeletal distribution of disease.

Table 1
Patient demographics, treatment and survival.

Case no.	Sex	Age	Site	Clinical presentation	Treatment 1	Margins	Time to recurrence (months)	Symptom free period (months)
1	F	5	Ischium	Pain	Nil	–	–	–
2	M	44	Radial diaphysis	Pain, swelling	Excision	Marginal	–	116
3	F	35	Proximal fibula	Pain, swelling	Excision	Marginal	–	145
4	F	22	Extensive lesion distal tibia	Pain	BKA	Amputation	–	158
5	M	15	Radius	Swelling	Excision + fibula graft	Marginal	–	183
6	M	24	Extensive lesion proximal tibia	Pain	EPR proximal tibia	Wide/ complete	–	181
7	F	29	Iliac wing	Pain	Nil	–	–	–
8	F	12	Calcaneum	Pain, swelling	Curettage	Intralesional	9	9
9	F	18	1st metacarpal	Swelling	Curettage	Intralesional	–	66
10	F	50	Talus	Pain, swelling	Curettage	Intralesional	–	59
11	F	48	2nd metatarsal	Pain	Curettage	Intralesional	–	23
12	M	25	Talus	Pain	Curettage	Intralesional	–	14
13	M	10	Distal radius	Pain, swelling	Curettage	Intralesional	7	7

4. Discussion

The aim of this study was to use a large database of patients with benign orthopaedic tumours in order to outline a management strategy for patients diagnosed with desmoplastic fibroma of bone and to discuss the criteria associated with local recurrence. Of the 4692 patients treated at our institute with a benign bone tumour over 30 years we found 13 had a diagnosis of desmoplastic fibroma of bone. Our unit has treated 1440 patients with osteosarcoma during the same period as we treated these patients with desmoplastic fibroma and it is known that the incidence of osteosarcoma is 2.5/million population per year [11]. Thus, the incidence of desmoplastic fibroma of bone is approximately 13/1440 of osteosarcoma i.e. a figure of 0.009/million population per year or approximately 1% of the reported incidence of osteosarcoma. This converts to an expected 2.5 cases of desmoplastic fibroma of bone for every 100 million population.

Local recurrence occurred in 15.4% of patients in our series and was statistically significantly linked to desmoplastic fibromas with associated soft-tissue components. This suggests that desmoplastic fibromas with extra-osseous extension are at the extreme end of the locally aggressive spectrum and, therefore, require more radical surgery than curettage or marginal excision.

The cross-sectional imaging features of desmoplastic fibroma have been described [2,9,12] as an osteolytic lesion with destruction of cortical bone, marginal sclerosis and pseudotrabeculation. The tumour shows low signal intensity on T1-weighted images with signal enhancement after the administration of contrast [2,9]. Cortical breakthrough was present in 23.1% of patients in our series, which is comparable to the study by Crim [13] who, after evaluating the radiographical features of desmoplastic fibroma in 83 published case reports, demonstrated cortical breakthrough in 29% of patients. We recommend magnetic resonance imaging (MRI) of the affected area to accurately delineate the extent of the tumour and it's relation to surrounding anatomical structures. Computed topography (CT) can supplement MRI and provide a more precise picture of the boney architecture so as to guide surgical management.

The diagnosis is very difficult on radiographical imaging alone as many tumours have similar radiological appearances to desmoplastic fibroma [2,7]. Biopsy is the gold standard method of determining the exact nature of any bony lesion as it provides a tissue diagnosis. The biopsy ideally should be performed at a reference bone tumour centre under the auspices of a fully accredited multi-disciplinary team. A unit familiar with the diagnosis of such rare pathology can, therefore, evaluate the clinical, radiological and histological features of this difficult disease so as to plan management.

Radiotherapy has been reported to be a therapeutic option in the treatment of desmoplastic fibroma. Sanfilippo et al. [14] used 60 Gy in 30 fractions to treat desmoplastic fibroma of the ilium, while Nag et al. [15] treated one patient with 45 Gy in 25 fractions for desmoplastic fibroma to the distal femur. We have no experience in the use of adjuvant radiotherapy in the management of desmoplastic fibroma of bone at our institute.

In conclusion, desmoplastic fibroma is a rare, locally aggressive, benign neoplasm of bone. The diagnosis and management is often difficult and requires a combined specialty approach. Our study highlights that extra-osseous tumour growth appears to be a poor prognostic sign. Wide resection is the ideal treatment of choice particularly if the lesion is located in a region where limb salvage surgery can provide restoration of function. Curettage or marginal resection can be used if en bloc resection would result in significant post-operative morbidity and loss of function. However, patients should be advised about the need for serial clinical and radiological follow-up to detect any local recurrence as early as possible, particularly if their tumours have soft-tissue components, with amputation always being available in the armamentarium for recalcitrant disease.

Conflict of interest statement

The authors declare that there are no conflicts of interest.

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