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Case Report

Desmoplastic Fibroma of the Phalanx of the Hand: A Rare Case Report

Belzinder Pal Singh[a, b] Syurahbil Abdul Halim[a, b] Sahran Yahaya[a, b]
Nor Azman Mat Zin[a, b] Sharifah Emilia Tuan Sharif[b, c]
Wan Faisham Wan Ismail[a, b]

[a]Department of Orthopaedics, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia; [b]Hospital Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia; [c]Department of Pathology, School of Medical Sciences, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia

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Desmoplastic · Fibroma · Phalanges · Benign

Abstract
Desmoplastic fibroma is a rare primary benign bone tumour that typically affects the long bones, mandible, and pelvis. It has a similar local aggressiveness to soft tissue fibromatosis. It rarely involves the small bones of the hand. We describe an extremely rare case of desmoplastic fibroma of the proximal phalanx of the hand in a patient who presented with an aggressively enlarging but painless mass on the left ring finger. Radiological features suggested malignancy; however, an initial biopsy revealed fibrotic tissue. Trans-metacarpal amputation of the ring and little fingers and soft tissue reconstruction were performed using a local ulnar-based flap of the little finger. The final histopathological evaluation revealed desmoplastic fibroma. Aggressively growing masses in the hand should be treated according to a sarcoma management protocol, and desmoplastic fibroma should be included in the differential diagnosis.

Introduction

Desmoplastic fibroma (DF) is a rare primary benign bone tumour. Histologically, it is characterized by spindle-shaped fibroblasts/myofibroblasts within a densely rich collagenous matrix [1]. Although it can occur in any bone, it commonly affects long bones (56%), mandible (22%), and pelvis (14%), with a predilection for the metaphysis of the femur, humerus, radius,

Correspondence to:
Syurahbil Abdul Halim, syurhbil@usm.my

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and tibia [2–4]. It infrequently presents in the small bones of the hand [3, 5, 6]. To the best of our knowledge, DF of the phalanx has never been reported. Herein, we present such a case and our management of it. The CARE Checklist has been completed by the authors for this case report, attached as in online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000529895).

Case Presentation

A 61-year-old truck driver was referred for a painless mass on the left ring finger that lasted for 1 year; the lesion began to expand aggressively during the last 3 months. It started at the finger base and gradually expanded to involve the entire finger, which markedly limited hand function (Fig. 1a). There was no precipitating factor, swelling elsewhere, or history of malignancy. The patient reported an unintentional weight loss of 15 kg without other constitutional symptoms.

An examination revealed a 10 × 10 cm circumferential mass on the left ring finger extending from the distal interphalangeal joint to the fourth metacarpal head. It was firm to hard in consistency and was multilobulated with an irregular surface. The mass was warm, non-tender and attached to the overlying skin with dilated veins. The little and middle fingers were splayed due to the mass effect. Sensation over the ring finger was reduced, but intact circulation.

A plain radiograph showed a large soft tissue mass of the left ring finger that displaced neighbouring fingers (Fig. 1c). There was extensive destruction of the proximal and middle phalanges with soft tissue calcification. Local magnetic resonance imaging (MRI) showed an expansile mass arising from the proximal phalanx with a soft tissue component occupying the adjacent web spaces (Fig. 1d). The mass showed heterogeneous isointense in T1W and T2W and enhancement post-contrast. There was a non-enhancing cystic area, which may represent a cystic component or necrotic area. An area of persistent hypointensity was observed on T1W, T2W, and blooming on GRE and may represent calcification. It eroded the fourth metacarpal head and infiltrated the adjacent lumbricals and interossei muscles. Computed tomography scan of the lung showed no evidence of lung metastasis. The clinical and radiological differential diagnosis at that point of time was chondrosarcoma or osteosarcoma.

A tissue biopsy showed a low-grade spindle cell lesion. Subsequently, the patient underwent rays amputation of the ring and small fingers and wound closure by means of a local ulnar-based flap originating from the small finger (Fig. 1b). Histopathological examination revealed an ill-circumscribed and destructive bone lesion composed of mostly uniform plump spindled cells that have an indistinct moderate eosinophilic to amphophilic cytoplasm. The intervening stroma was highly collagenized. No osteoid was seen. Thick fibrous septae were present in between the tumour cells. No significant nuclear atypia or mitosis was observed (Fig. 2). Tumour cells were focally positive for SMA but negative for desmin, EMA, BCL2, TLE-1, SATB2, STAT6, CK7, CD34, and B-catenin. The overall morphology was in favour of DF of bone. However, the surgical resected margin was involved with tumour cells. At 6 months of follow-up, there was no local recurrence and he had good overall hand function with acceptable hand grip strength (Fig. 3). Nevertheless, a long-term follow-up was necessary to evaluate for local recurrence.

Discussion

DF accounts for 0.1–0.13% of all primary bone tumours [1, 2]. It shows similar local aggressiveness to soft tissue fibromatosis [1]. The majority occurs in those below 30 years of age [7]. The cause is still undetermined. DF of small bones of the hand is extremely rare and

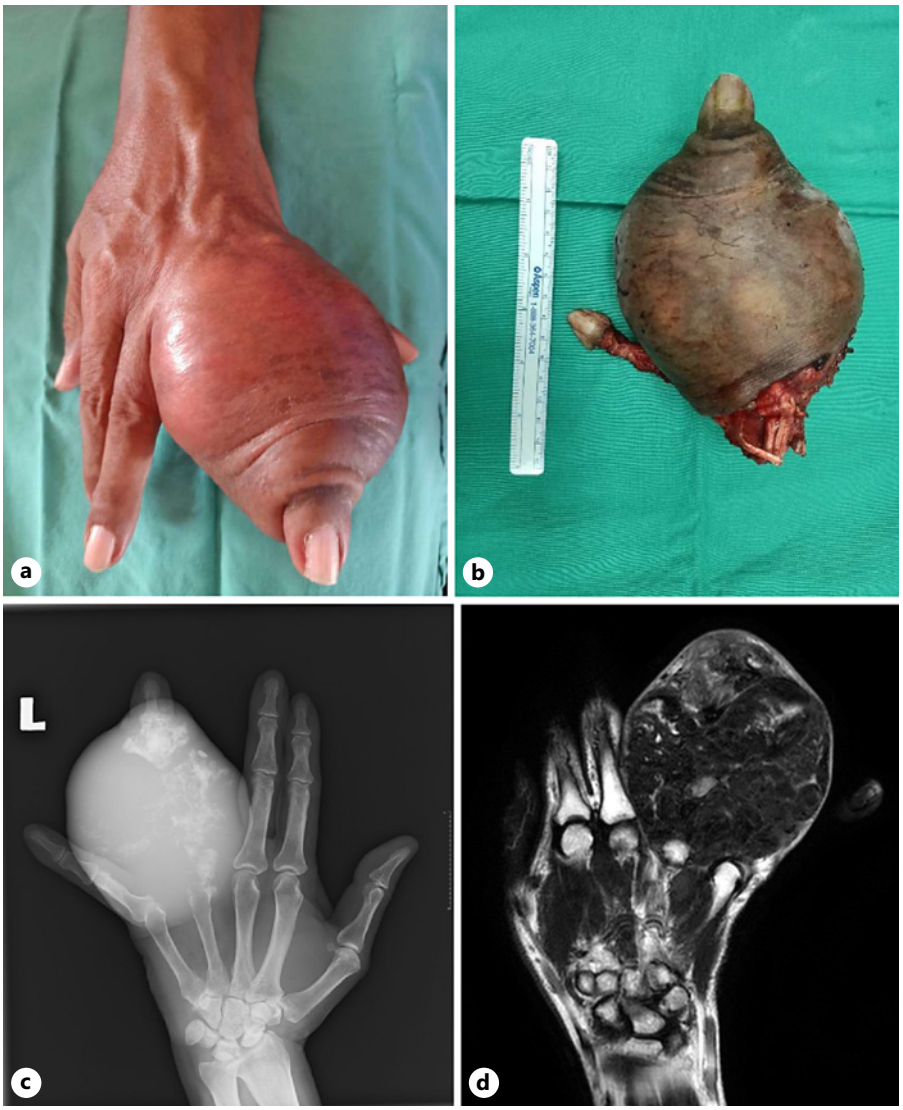

Fig. 1. a Clinical presentation of mass over left ring finger. **b** Resected specimen. **c** Radiograph left hand showing extensive destruction of phalanges of left ring finger. **d** MRI T2-weighted image showing heterogenous signal intensity within the mass.

has only been reported involving the metacarpus [3, 5, 6]. To our knowledge, DF in the phalanges of the hand has never been described, especially with extensive surrounding soft tissue involvement.

The diagnosis of DF is challenging because the clinical presentation can mimic a wide range of benign and malignant pathologies such as lipoma, schwannoma, giant cell tumours, intraosseous osteosarcoma, or low-grade fibrosarcoma [3, 8, 9]. Thus, differentiating DF from sarcoma, particularly low-grade fibrosarcoma, is difficult [10].

The clinical presentation is nonspecific, ranging from incidental findings to painful swelling/deformity and progressive loss of function [1]. Unlike our case, most of the DF in the hand presents as gradual swelling, discomfort, and a small mass [5, 6, 8]. DF rarely presents as a large mass mimicking sarcoma [9]. Radiologically, DF has no pathognomonic characteristics. Common features include radiolucency with varying degrees of margin, a soap bubble appearance, cortical thinning or destruction, and rarely, matrix mineralization [11, 12]. As 50% of DF extends into the

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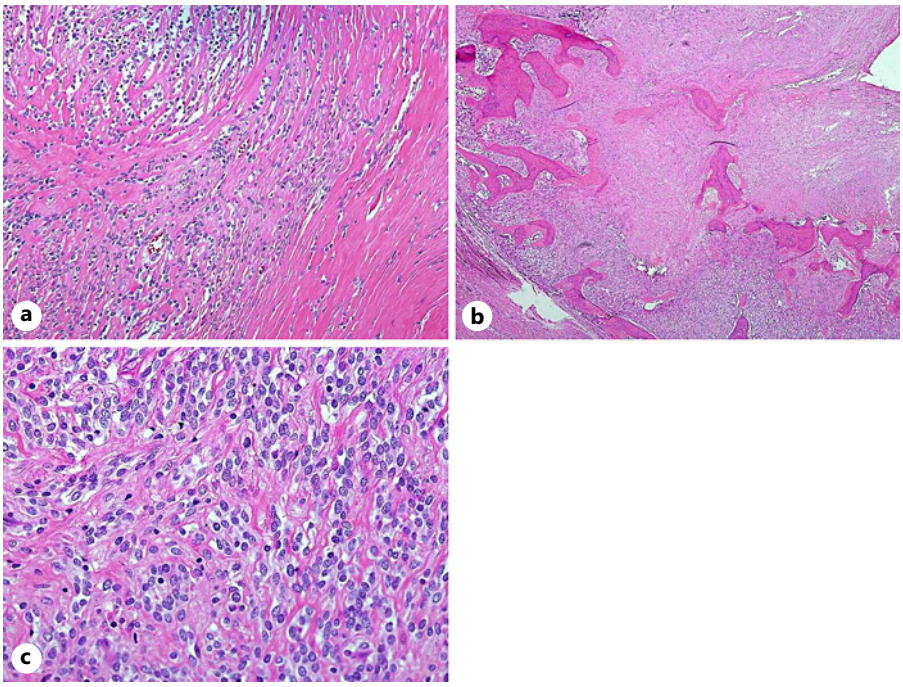

Fig. 2. Haematoxylin and eosin (H&E) stain (**a**, **b** ×40) exhibits spindle cells arranged in infiltrating the bone with highly collagenized stroma. **c** At high magnification (×400), tumour cells show uniform bland spindled to oval cells with vesicular nuclei and indistinct cytoplasm. Note the pinkish collagen fibres in between the tumour cells.

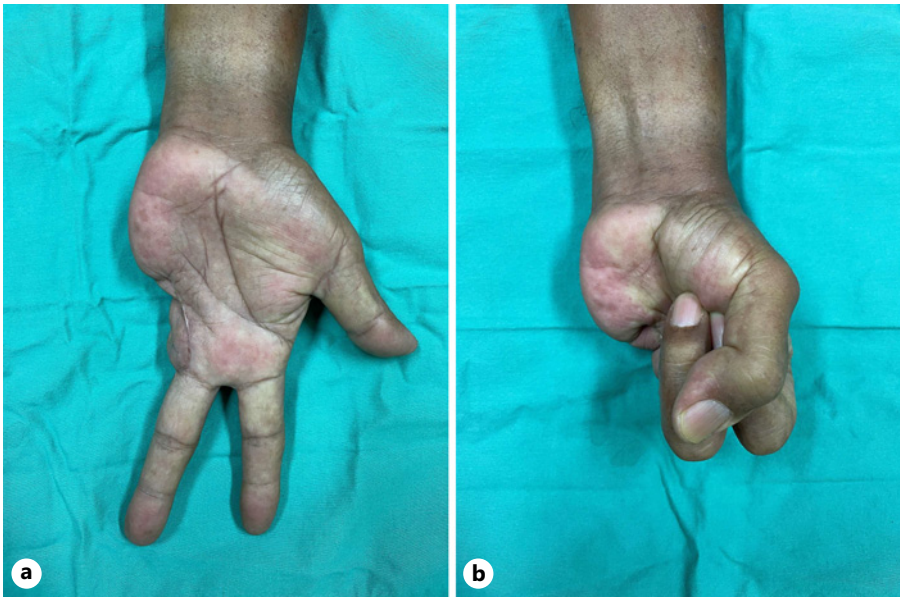

Fig. 3. a, **b** Left-hand appearance and function at 6-month follow-up.

adjacent soft tissue, the imaging modality of choice is MRI to delineate medullary and soft tissue expansion [2, 12]. T2-weighted MRI showing low to intermediate signal intensities may differentiate DF from others [11]. Our imaging features strongly suggested malignancy such as heterogeneity, huge destructive lesion, and necrotic areas.

The histology of DF is similar to desmoid-fibromatosis of soft tissue. It is an infiltrative, unencapsulated tumour composed of uniform spindle fibroblastic/myofibroblastic cells, with low to variable cellularity in the highly collagenized stroma [1]. DF characteristically lacks nuclear atypia and mitosis. However, it may confuse low-grade (central) osteosarcoma and fibrous dysplasia with a more cellular lesion. Therefore, correlation with radiology is essential. Although histology is similar to that of soft tissue, positive beta-catenin, which has been linked to desmoid-type fibromatosis is usually inconclusive in DF [13]. B-catenin nuclear positivity was not detected in a few DF series [13], which was also in ours.

The purpose of treatment is to completely excise the lesion and prevent local recurrence. Depending on the size, location, and expected functional morbidity surgical treatment ranges from simple intralesional curettage to wide resection [3, 6, 12]. Recurrence is high in intralesional excision, up to 47% [7]. Thus, wide-margin excision is preferable [6, 12]. If a wide margin is impossible, postoperative irradiation may be appropriate [14]. We approach our case as per sarcoma due to advancing age, increasing mass with extensive destruction of phalanges and large soft tissue infiltration, and inconclusive biopsy. Our dilemma was to obtain a clear resection margin and maintain good hand function. Evans et al. [6] showed that infiltration into adjacent soft tissue is associated with a poor prognosis and requires more extensive surgical intervention.

Conclusion

DF can occur in the phalanges of the hand, and the differentials for an aggressively growing mass in the hand must include DF, in addition to sarcoma. Treatment aim should be to achieve a wide-margin excision while preserving the patient's residual good hand function. Follow-up should be regular and over a long duration to detect local recurrence.

Statement of Ethics

A written informed consent was obtained from the patient for the publication of the details of the medical case and any accompanying images. This case report did not require ethical approval in accordance with local guidelines.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Data acquisition and manuscript drafting: Belzinder Pal Singh, Syurahbil Abdul Halim, and Sharifah Emilia Tuan Sharif. Data analysis, critical revision, and final manuscript: Syurahbil Abdul Halim, Sahran Yahaya, Nor Azman Mat Zin, Wan Faisham Wan Ismail, and Sharifah Emilia Tuan Sharif.

Data Availability Statement

All data generated or analysed in this case are included in this article and its online supplementary material files. Further enquiries can be directed to the corresponding author.

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