Desmoplastic fibroma of the ulna bone

Samuel U. Eyesan, Tolulope G. Kehinde, Adesope S. Adesina, Christopher S. Ayeni, Blaise F. Abdulkareem

Department of Orthopaedic Surgery, Bowen University Teaching Hospital, Ogbomoso, *Department of Morbid Anatomy and Histopathology, College of Medicine, University of Lagos – Idi Araba, Nigeria

ABSTRACT

A fibroma is a benign tumour composed of fibrous connective tissue and they can grow in all organs. They can be classified based on consistency into hard or soft fibroma, based on histological characteristics into desmoplastic, chondromyxoid, ossifying, non-ossifying fibroma. They can also be classified based on tissue of origin or location in the body, it can also be classified into superficial or deep fibroma. This is a report of a 15-year-old Nigerian boy with a recurrent left ulnar tumour which was rapidly growing and has been excised three times. Preoperative investigations, i.e., plain radiograph, full blood count and fine needle aspiration cytology were done. Patient could not afford CT scan which was requested for. First excision was in 2009. Recurred within 1 year and had a repeat excision in 2011. He had a second recurrence 9 months after and had to have another excision. The last tumour excision left only the part of the left ulna that forms the proximal radioulnar joint and elbow joint. Patient has been followed up after the last excision for 18 months with no recurrence. 

Key words: Bone, desmoplastic, fibroma, ulna

INTRODUCTION

A desmoplastic fibroma is an extremely rare tumour with less than 200 cases in the published literature. It is a solitary tumour, microscopically composed of well-differentiated myofibroblasts with abundant collagen production.

It is rare, accounting for 0.1% of all primary bone tumours. It is asymptomatic in the initial stages but symptomatic cases present with pain and swelling, may present with effusion if near a joint and pathological fracture has been reported in only 12% of cases. Plain radiograph features include osteolytic, expansile medullary lesion with well-defined sclerotic margins.

Diagnosis is made by plain radiographs, bone scans, CT scan, MRI and histopathology.

Treatment option for this tumour is marginal or wide surgical excision.

Genetically, it is associated with trisomies 8 and 20. Chondromyxoid fibroma is a close differential of this tumour.

CASE REPORT

A 15-year-old boy first presented at National Orthopaedic Hospital, Igbobi (NOHI) with a left ulnar tumour about 4 years ago (2009) and subsequently had excision of the tumour. Plain radiograph of the left forearm before excision is as shown in Figure 1. Histology revealed a firm mass with a cut section showing grayish yellow surface macroscopically and microscopy showed plump spindle nuclei arranged in stacks and disposed in haphazard interlacing pattern with fragments of cartilage and bone as shown in Figures 4 and 5, a diagnosis of fibroma was confirmed.

Patient presented at Bowen University Teaching Hospital (BUTH) about 1 year after the first excision with a recurrence at the same location of 6-month duration. Plain radiograph after recurrence, before repeat excision is as shown in Figure 2. He subsequently had a repeat excision. He represented at BUTH, Ogbomoso 9 months after the second excision with a recurrence at the same location for which he had another excision leaving only the part of the left ulna that forms the proximal radioulnar joint and the elbow joint as shown in Figure 3. The histological
Eyesan, et al.: Desmoplastic fibroma of the ulna bone

Figure 1: Plain radiograph of the patient before the first excision. The moth eaten end of the distal ulna and the bone tumour in the soft tissue eroding the media cortex of the radius.

Figure 2: Plain radiograph of the first recurrence. Again observe the moth eaten end of the ulna.

Figure 3: Plain radiograph after the last excision.

Figure 4: Photomicrograph of the tumour after excision.

Figure 5: Same photomicrograph at ×40 magnification.

A fibroma is a benign tumour composed of fibrous connective tissue and they can grow in all organs of the body. Histologically, they can be classified into certain specific types including chondromyxoid fibroma, desmoplastic fibroma, ossifying fibroma, non-ossifying fibroma, cemento-ossifying fibroma and cystic fibroma. Fibroma can be superficial or deep, soft or hard or could belong to a wide spectrum of fibroblastic and myofibroblastic neoplasm with similar pathological appearances and variable clinical behavior collectively known as Musculoaponeurotic Fibromatoses.

A desmoplastic fibroma is an extremely rare tumour with less than 200 cases in the published literature. It is characterized by aggressive local infiltration and the most common site is the mandible followed by the femur and pelvis. It is a solitary...

features of the excised specimen have remained the same with each excision.

Post-operative treatment included analgesic and wound dressing. He is still been followed up. There has been no recurrence in the last 20 months.

DISCUSSION

A fibroma is a benign tumour composed of fibrous connective tissue and they can grow in all organs of the body. Histologically, they can be classified into certain specific types including chondromyxoid fibroma, desmoplastic fibroma, ossifying fibroma, non-ossifying fibroma, cemento-ossifying fibroma and cystic fibroma. Fibroma can be superficial or deep, soft or hard or could belong to a wide spectrum of fibroblastic and myofibroblastic neoplasm with similar pathological appearances and variable clinical behavior collectively known as Musculoaponeurotic Fibromatoses.

A desmoplastic fibroma is an extremely rare tumour with less than 200 cases in the published literature. It is characterized by aggressive local infiltration and the most common site is the mandible followed by the femur and pelvis. It is a solitary...
Eyesan, et al.: Desmoplastic fibroma of the ulnar bone

It occurs most often in the adolescent and young adults (first three decades of life) and there is no sex predilection.\textsuperscript{1-5} It is rare, accounting for 0.1% of all primary bone tumors.\textsuperscript{1-4} It is asymptomatic in the initial stages but symptomatic cases present with pain and swelling, may present with an effusion if near a joint and pathological fracture has been reported in only 12% of cases.\textsuperscript{1}

However, pathological fracture or deformity of the affected bone can be a presenting symptom.\textsuperscript{5}

Plain radiograph features include osteolytic, expansile medullary lesion with well-defined sclerotic margins.\textsuperscript{1} The tumor is often found in the metaphysis aligned with the long axis of the bone, cortex is usually thinned and fine intralesional trabeculae gives a lobulated appearance describes as ‘soap-bubble’.\textsuperscript{1} ‘Honeycomb’ or ‘moth-eaten’ patterns have also been described.\textsuperscript{5} CT scan enhances the cortical breakthrough. These features are corroborated by the findings in our patient’s radiograph in Figures 1 and 2. MRI demonstrates the separation of the interosseous tumour from the bone\textsuperscript{7} but has a low signal intensity on both T1 and T2-weighted images.\textsuperscript{5}

Histopathology shows grayish to yellowish white colour and a rubbery consistency with irregular edges, round and blunt. The tumour has occasional cystic foci with clear fluid. Microscopically, the tumour has interlacing bundles of dense collagen and low cellularity. The fusiform cells that are present have no atypia and the nuclei are ovoid and elongated.\textsuperscript{1} Both the macroscopic and microscopic picture of the resected specimen of our index case also fit well into this pattern.

Immunohistochemistry shows myofibroblastic and more primitive component which are positive for vimentin and smooth muscle actin, while the myofibroblastic component is more strongly positive for pan-actin HHF – 35.\textsuperscript{5} Due to lack of facilities we could not assay for vimentin and pan-actin HHF-35.

Treatment option for this tumor is marginal or wide surgical excision. Without resection, recurrence rate is 55-72% and with resection, recurrence is 17%. One study has recommended aggressive curettage as surgical option.\textsuperscript{1-4} Local relapse has been reported as late as 8 years following primary surgery.\textsuperscript{5} In the absence of frozen section facilities our margins in the first two resections has appeared less than adequate. There has, however, been no recurrence in our index case after the third resection in almost 2 years. This may be because we tried to be more generous with the level of bone section during the last surgery in the absence of frozen section facility. He is still been followed up.

Due to risk of local recurrence\textsuperscript{1-4} propensity to pathological fracture, curettage and intralesional treatment are not recommended for this lesion and wide resection is associated with a low rate of recurrence.\textsuperscript{1,4} We believe that our first two resections may not have been wide enough.

CONCLUSIONS

Desmoplastic fibroma of the bone is an aggressive tumour with a high rate of recurrence, however, with a chance of a cure without significant morbidity if properly managed.

REFERENCES

1. Böhm P, Kröber S, Greschner A, Laniado M, Kaiserling E. Desmoplastic fibroma of bone. Cancer 1996;78:1011-23.
2. Nishida J, Tajima K, Abe M, Honda M, Inomata Y, Shimamura T, et al. Desmoplastic fibroma Clin Orthop Relat Res 1995;320:142-8.
3. Bullough, Peter. Orthopaedic Pathology, 3rd ed. London: Times Mirror International Publishers Limited; 1997.
4. Hurous, Andrew. Bone Tumors: Diagnosis, Treatment and Prognosis. Philadelphia: W.B. Saunders Co.; 1991.
5. Benign Bone Tumors – Paediatric Orthopaedic Pathology. Available from: http://www.pedorthopath.com/about.html [Last accessed on 2013 Mar 15].
6. Available from: http://en.m.wikipedia.org/wiki/fibroma. [Last accessed on 2013 Apr 20].
7. D’Souza H, Kulkarni DV, Patel B, Shivraman A. Musculoaponeurotic fibromatosis of both bones of the forearm. J Postgrad Med 1996;42:57-9.