THIGH TUMOURS AT KENYATTA NATIONAL HOSPITAL

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ABSTRACT

***Background*: The thigh is a common seat for tumours. Many histopathological types can arise there. Management challenges begin from the clinical diagnosis, to the surgery and ultimately the oncological therapy be it medical or radiation based.**

***Objective*: To review the thigh tumours prospectively managed by the authors over a 12 year period.**

***Design*: A 12 year prospective observational study.**

***Setting*: Ward 6D of the Kenyatta National Hospital, Nairobi, Kenya. An Orthopaedic Unit.**

***Patients and Methods:* Forty one patients admitted to Ward 6D with thigh tumours between 1st October 1999 and 30th September 2011 and treated by the authors in consecutive and prospective sequence are analysed.**

***Results*: Forty one patients with thigh tumours treated prospectively over the 12 year period.**

***Conclusions:* Twenty three histopathological varieties of tumours occured in the thigh and their diagnosis and management encompased incisional biopsy, excisional biopsy, curretage and bone grafting, amputation and hind quarter disarticlation. Adjuvant chemotherapy and radiotherapy were administered. Distant metastases necesitated specialised surgical procedures.**

INTRODUCTION

Thigh tumours are of numerous varieties and arise in all age groups. They originate from any of the anatomical tissues in the thigh. They may also be benign or of all grades of malignancy. Diagnosis rests on the utilization of thorough physical examination coupled with several imaging modalities and finaly histopathological examination of biopsied tissue.

Definitive treatment is primarily surgical with chemotherapy and radiotherapy as adjunctive therapies.

The thigh consists of a cylinder of compact bone, the femoral shaft, which is completely enclosed by muscle groups through which important neurovascular pathways run (1). It is considered the most common seat of musculoskeletal tumours (2,3).The region comprises of skin, soft tissues mainly muscles, the femur, nerves and the vasculature. Each of these tissues is a known source of neoplastic growths. The femur has various benign neoplastic tumours known to originate there. Malignant neoplasms also arise from the femur. The predominant malignant lesions from the femur are mainly osteogenic sarcoma, Ewings sarcoma and chondrosarcoma (4). The 'other bone sarcomas' arising from the femur are rare lesions and include fibrosarcoma, malignant fibrous histiocytoma, leiomyosarcoma and others (4). Other lesions such as myositis ossificans reported to mimick osteosarcoma are also on record arising in the thigh (5).

The soft tissues of the thigh give rise to tumours more commonly than the femur bone .The bulk of these are benign tumours (6), though a number of malignant tumours dubbed soft tissue sarcomas also do occur.The variety of soft tissue tumours that can arise in the thigh is wide, forming a heterogenous group containing more than 200 clinico-pathological sub-types (7).

Liposarcoma is considered as one of the commonest soft tissue sarcoma (8,9), followed by synovial sarcoma, malignant fibrous hystiocytoma, fibrosarcoma, epithelioid sarcoma and clear-cell sarcoma (2,3,7,8). Other lesions such as lipomas and

aneurysmal bone cysts of soft tissue have also been documented (8,10). The thigh is observed to be the site of occurence of one third of all soft tissue sarcomas (2,3). Despite the wide variety of tumour types occuring in the thigh, these are rare tumours, comprising only 0.5-1 % of all malignancies (3,7,11,12).

Plain radiographs are important in determining whether the lesion is primarily in soft tissue or in bone (11) while CT scan and MRI have become the gold standard for evaluating the size and extent of a soft tissue sarcoma (11). The primary treatment for these thigh tumours is surgical excision (9, 13,17,18), with limb-sparing surgery being given foremost consideration (14,15). One form of biopsy is mandatory to precede excision. The acknowledged biopsy forms are fine needle aspiration, core needle biopsy, excisional biopsy and incisional biopsy (11,13,19).

Radiation therapy is the most advocated adjuvant modality of treatment (15,16) and chemotherapy administered in a minority of these lesions, as most of them are considered to be scarcely chemosensitive (18). Radiation alone as treatment for these tumours is advocated in some very rare instances of those patients who are technically or medically inoperable and whose lesions are relatively small (16). Because of the relative infrequency of the different histopathological subtypes, no single institution examines an adequate number of cases of each subtype within a short period to allow treatment comparisons (12). Also, due to its rarity, no large prospective randomized trial has been conducted on treatment of extremity soft tissue sarcoma (12).The other consequence of this is that the general pathologist and orthopaedic surgeon may not meet these types of tumours for years (7). Ultimately, clinicians have been left with single institutional experiences gathered either in a retrospective or prospective fashion. Ward 6D of Kenyatta National Hospital is one such Institution. It is here that we have undertaken a twelve years prospective observational study on thigh tumours.

MATERIALS AND METHODS

Between 1st October 1999 and 30th September 2011, we as orthopaedic surgeons based in Ward 6D of the Kenyatta National Hospital, Nairobi, Kenya, operatively treated 41 patients with thigh tumours. The complete medical records of these patients have been retrieved and analysed with the approval of the Kenyatta NationalHospital/University of Nairobi Ethical and Research Committee . The accompanying radiographs, laboratory reports and charts have also been analysed. The histopathological reports of the examined tumour tissues have been reproduced as reported. No review of the histopathological examination of the tumour tissues was sort. All patients treated for thigh tumours during the duration of this study whose complete records were traced are included in the analysis. Data pertaining to the age of the patients, gender, site of tumour, side affected, previous surgeries on tumour, imaging studies conducted on tumour, clinical size of tumour, radiological size of tumour, pathological size of tumour, surgical and diagnostic procedures performed, histopathological studies reported, adjuvant modalities of treatment received and length of hospital stay and outpatient post operative follow-up were extracted. Data analysis was carried out manually with the computation of means, ranges and percentages being done with the aid of a basic electronic calculator.

RESULTS

Over the study period spanning 1st October 1999 to 30th September 2011 we prospectively treated 41 patients with thigh tumours. The complete records of 40 patients were traced and are included in this analysis. The one patient whose complete records were not traced is ommitted from analysis in this study. The yearly number of patients with thigh tumours is broken down in Table 1. The age distribution of the patients is captured in Table 2, with peak incidence being in the second decade of life and a smaller peak in the fourth decade. The youngest patient was 3 years while the oldest was 70 years old. The mean age was 31.4 years. Out of the 40 patients analysed, 23 (57.5%) were males, while 17 (42.5%) were females. Twenty two patients (55%) had left thigh tumours, while 18 (45%) had right thigh tumours. Tumours in the distal thigh comprised the bulk in 24 patients (60%), followed by 15 (37.5%) proximal thigh tumours, and only 1(2.5%) middle thigh tumour. Thirty two patients(80%) had had no prioir surgical intervention, while 4 (10%) patients had one previous surgery, 2 (5%) patients having had two previous surgeries and 2 (5%) other patients having had three previous surgeries. The ultimate summary was that 38 (95%) patients had primary thigh tumours while 2 (5%) patients had metastatic thigh tumours. Further on, 25 (62.5%) patients had thigh tumours of soft tissue origin whereas 15 (37.5%) patients had tumours of bony origin.

The most utilized and most informative imaging modality was the plain X-ray(Table 3). Thirty eight patients underwent plain X-ray with the most common finding being soft tissue swelling(22 patients), followed by bone destruction (10 patients). Findings from CT-Scan(2 patients), MRI(6 patients) and MRI angiography(1 patient), did not appear to contribute much to the diagnosis nor to the operative management planning. Most patients were managed by a single procedure that was either a diagnostic incisional biopsy or an excisional biopsy(Table 4). Additional surgery was required for a minority of patients who either had inconsistent histopathological reports, or had local recurrences or had distant metastases requiring other specialists intervention (Table 5).

A total of 23 histopathological varieties of thigh tumours were encountered with the most common and most consistent tumour being the osteogenic sarcoma (Table 6, Figure 1). Next in frequency were the lipomas, inflammatory and non-specific tumours.

Length of hospital stay was very varied. Lipoma patients recorded the shortest stay, averaging 3.5 days while osteogenic sarcoma recorded the longest stay, averaging 99 days but with a range of 8 to 263 days . Radiotherapy was the most administered mode of adjuvant therapy (7 patients), with 6 patients receiving chemotherapy (Table 7). It is notable that only one patient received neo-adjuvant chemotherapy. Four tumour types recorded metastases to distant sites (Table 8), each creating a need for specialized surgery or radiotherapy. Over the study period three mortalities were recorded, one from metastatic adenocarcinoma at 6 weeks, another from osteogenic (pleomorphic) sarcoma at 8 months and lastly from osteogenic (chondroblastic) sarcoma at 2 years.

Table 1
Thigh tumours ward 6D 1999-2011

Year	No. of patients
2000	4
2001	2
2002	6
2003	1
2004	4
2005	1
2006	2
2007	4
2008	4
2009	1
2010	5
2011	7
Total	*41*

Table 2
Age distribution

Age group (years)	No. of patients	Age (%)
1-10	5	12.5
11-20	11	27.5
21-30	5	12.5
31-40	8	20
41-50	3	7.5
51-60	2	5
61-70	6	15
Total	40	100

Table 3
Imaging modalities

A) Plain X-rays: 38 patients (2 patients: MRI, CT Scan)

X-ray feature	No. of patients
Soft tissues swelling	22
Bone destruction	10
Calcification	5
New bone formation (Codman's Triangles)	2
Periosteal reaction	5
Osteoarthritis both knees	1
Osteoarthritis lumbar spine	1
Pathological fracture	5
Osteoporosis	1
Bone cyst	4
K-nail *in-situ*	*1*
Bone dysplasia	*1*

B)CT Scans—2 patients:

a) Tumour infiltrating Gastrocnemius

b) Non specific bony tumour proximal left thigh

C) MRI—6 patients---5 Proximal, 1 Distal

1. Recurrent neurofibrosarcoma of left femoral stum
2. Mixed intensity mass in left gluteus medius/ maximus suggesting a soft tissue sarcoma.
3. Right buttock soft tissue mass
4. There is an ill-defined soft-tissue mass with necrotic areas involving right proximal anterolateral thigh
5. Expansile multiseptated right intertrochanteric bone lesion

6. Bone cyst on posterior distal femur and periosteal reaction

D) MRI angiography—1 patient: Moderately vascular tumour of left thigh.

Table 4

Diagnostic procedures performed

Diagnostic procedures	No. of patients
Fine needle aspiration	2
--Pleomorphic sarcoma	
--ongoing chronic inflammation	
(Metastatic thyroid adenoma)	
Incisional biopsy × 1	13
Incisional biopsy × 2	3
Excision biopsy	19
Excision biopsy × 2	1
Excision biopsy × 3	1
Curretage & bone graft	3

Table 5

Surgery performed (additional to diagnostic procedure)

Surgical procedure	No. of patients
Above knee amputation	6
Debulking	1
Underwater seal drainage	1
Ventriculo-Peritoneal Shunting (VP)	1
Excision of paraspinal mass	1
Disarticulation at right hip	1
Skin graft	1

Table 6

Histological types of thigh tumours in ward 6D KNH 1999-2011

Histopathological type	No. of patients	Age %
Osteogenic sarcoma	6	15
Lipoma	4	10
Inflammatory (Osteomyelitis)	4	10
Non specific	4	10
Neurofibroma	3	7.5
Fibrosarcoma	2	5.0
Fibrous cyst	1	2.5
Malignant peripheral nerve sheath	1	2.5
Dermatofibrosarcoma	1	2.5
Dermatofibrosarcoma protuberans	1	2.5
Myxoid fibroma	1	2.5
Aneurysmal bone cyst	1	2.5
Metastatic adenocarcinoma	1	2.5
Metastatic thyroid adenoma	1	2.5
Haematoma	1	2.5
Villonodular synovitis (GCT synovium)	1	2.5
Malignant fibrous hystiocytoma	1	2.5
Desmoid tumour	1	2.5
Synovial osteochondromatosis	1	2.5
Benign synovioma	1	2.5
Fibrous dyplasia	1	2.5
Non Hodgkins lymphoma	1	2.5
Giant cell tumour of bone	1	2.5
Total	40	100

Table 7

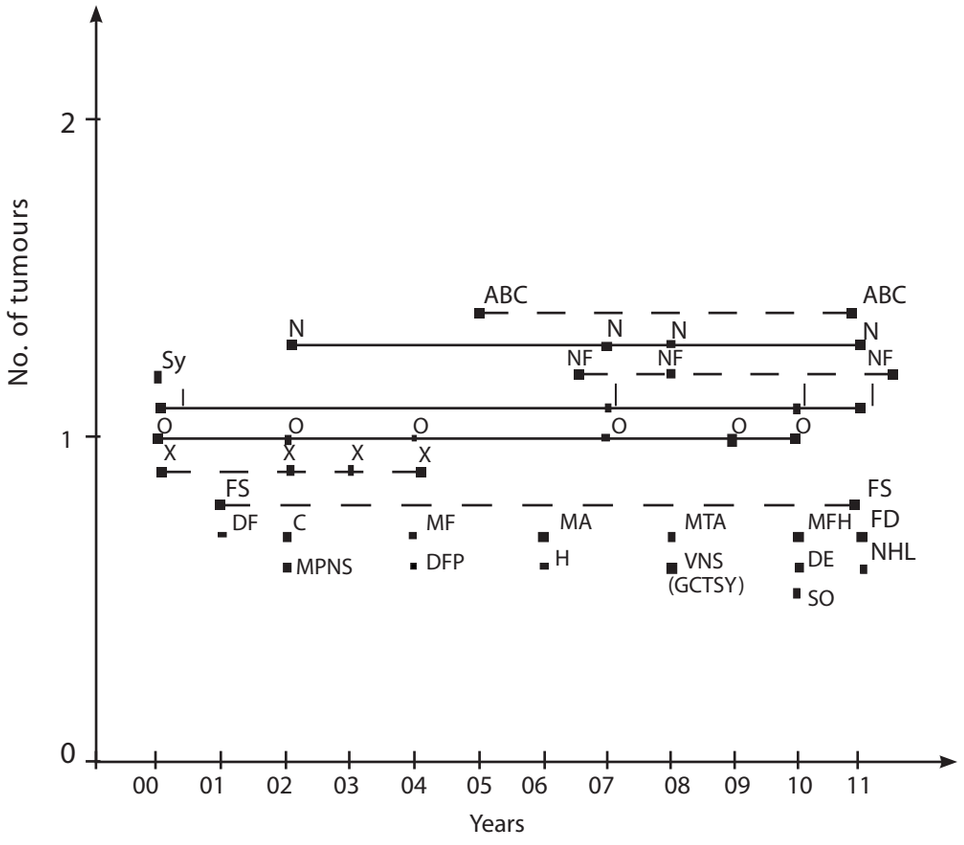
Adjuvant therapy

Modality of therapy	Regime	No. of patients
Neoadjuvant chemotherapy	VAC Pleomorphic (Osteogenic)Sarcoma	1
Chemotherapy	VAC (Osteosarcoma)	4
	Doxorubicin/Cisplatin (Dermatofibrosarcoma Protuberans)	1
Radiotherapy	Malignant peripheral nerve sheath tumour dermatofibrosarcoma protuberans	7
	Pleomorphic (Osteogenic) sarcoma	
	Osteogenic sarcoma-left thigh metastases	
	Metastatic thyroid adenoma	
	Desmoid tumour	
	Osteogenic sarcoma	

Table 8
Metastatic patterns

Histological tumour type (4)	Metastatic site(s)
Malignant Peripheral Nerve Sheath Tumour	Brain, liver
Pleomorphic (Osteogenic) sarcoma	Chest (pleural effussion)
	Posterior cranial fossa (hydrocephalus)
Myxoid fibror	Thoracic Paraspinal (Myxoid Liposarcoma
Osteogenic sa ma (RT)	Left thigh

Figure 1
Thigh tumours ward, KNH 6D 1999-2011



KEY:

ABC -Aneurysmal Bone Cyst
DF - Dermatofibroma
F - Fibroma
MPNS - Malignant Peripheral Protuberas Sheath Tumour
DFP -Dermatofibrosarcoma Protuberans
MA - Matastatic Adenocarcinoma
H - Haematoma
NF - Neurofibroma
MTA - Matastatic thyroid Adenoma
VNS - Villonodulla Synovitis
Sy - Synovioma
I - Inflamatory
O - Osteosarcoma
X - Lipoma
N - Non specific tissues
C - Cyst
NHL - Non - Hodgkin Lymphoma
MF - Malignant Fibroma
MFH - Malignant Fibrous Hystiocytoma
DE - Desimoid
SO - Synovial Osteochondromatosis
FS - Fibrosarcoma
FD - Fibrous Dysplasia

Figure 2
Distal left thigh tumour in an 11 year old boy. Osteogenic (Pleomorphic) sarcoma

Figure 3
X-ray of distal thigh soft tissue tumour. A haematoma.

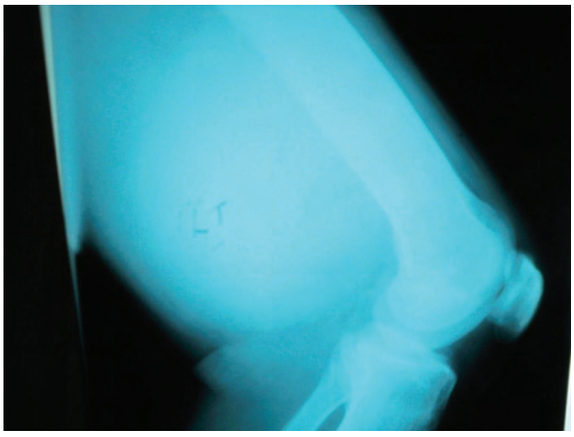


Figure 4
X-ray of distal bony left thigh tumour in a 22 year old male. Non specific tumour tissue

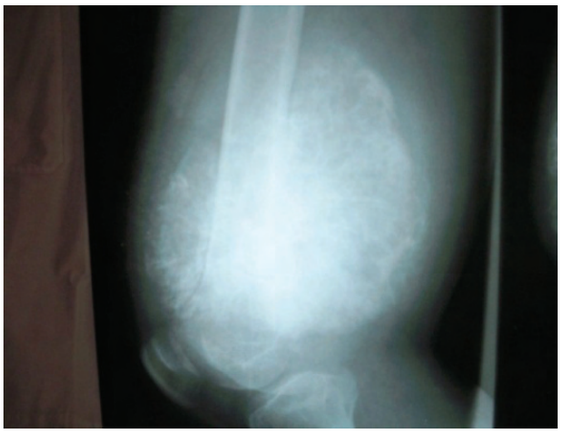


Figure 5A
Distal right thigh tumour

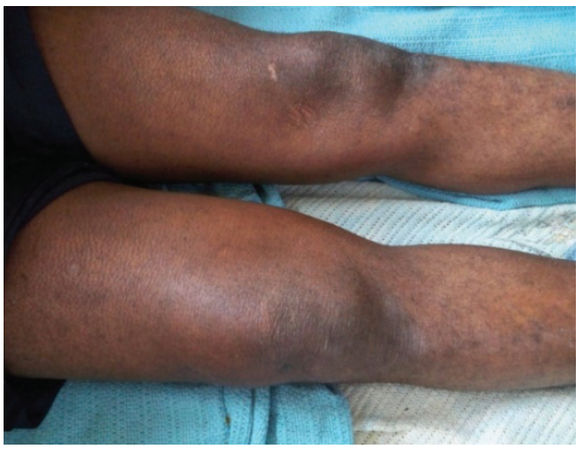


Figure5B
X-ray showing cystic lesion in Right distal femur. A giant cell tumour of bone

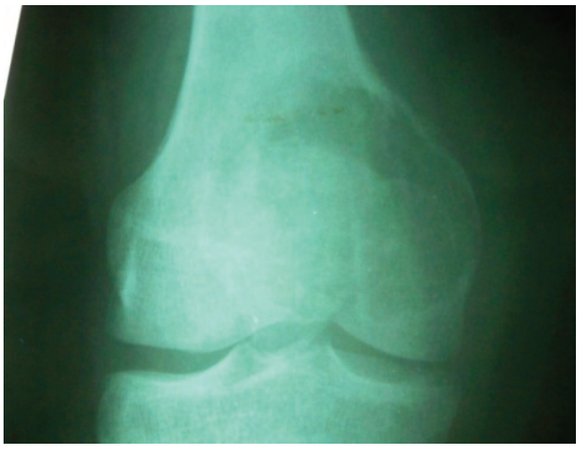


Figure 6A
Fungating proximal soft tissue thigh tumour

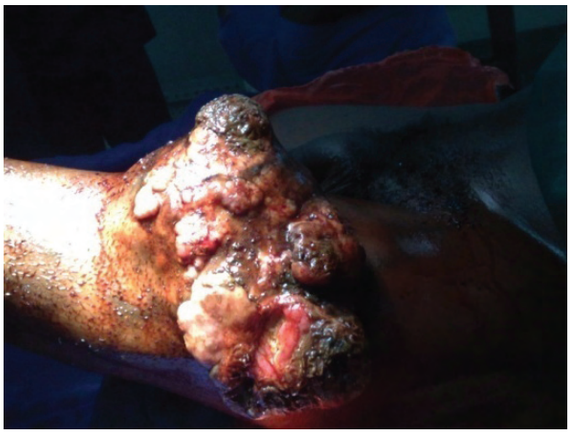


Figure 6B

X-ray of fungating soft tissue thigh tumour. A malignant fibrous histiocytoma

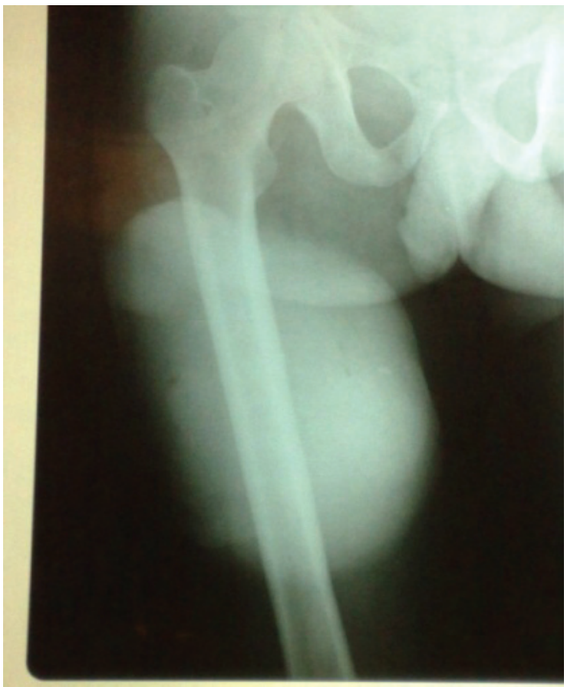


Figure 7

Extensive involvement of the proximal femur with a permeative destructive process and pathological fracture. A Non-Hodgkins Lymphoma.

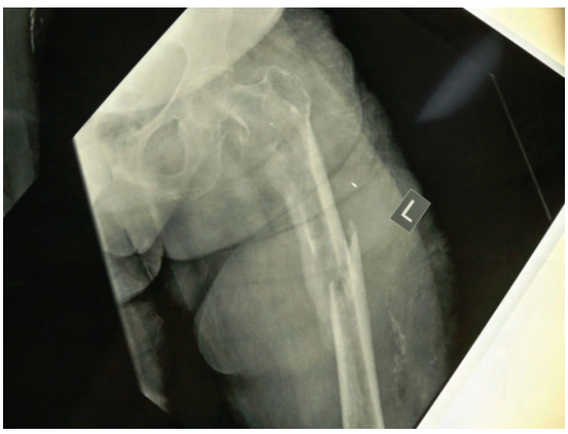


Figure 8

Dysplasia of proximal femur, old healed fracture, fresh pathological fracture and soft tissue tumour of proximal thigh (DESIMOID)

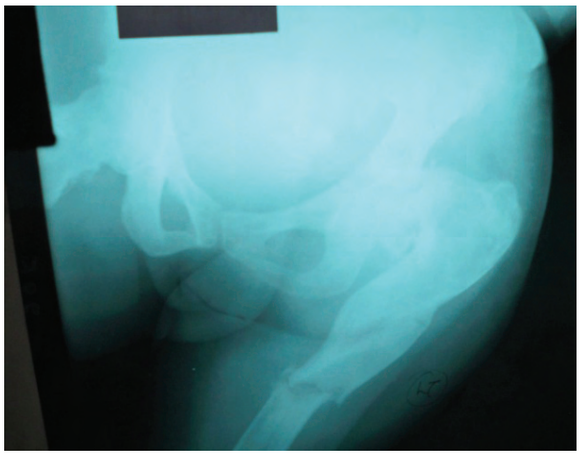


Figure 9

A previously operated distal left thigh tumour

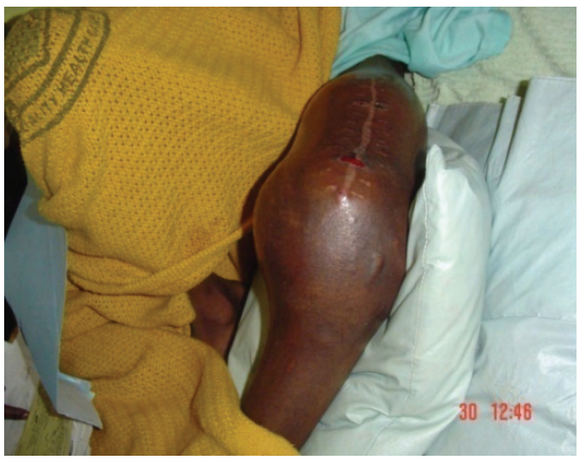


Figure 10A

A bony proximal left thigh tumour

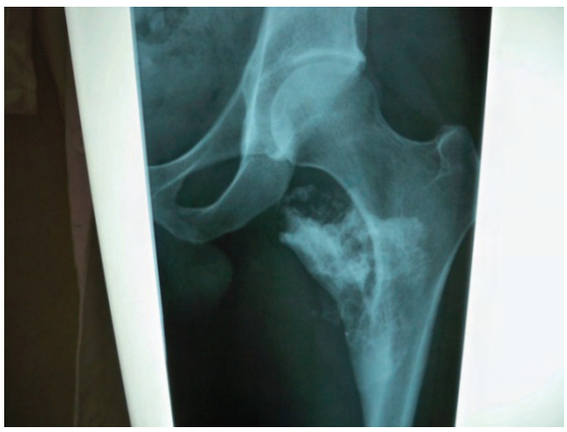


Figure 10B
CT scan of proximal bony left thigh tumour. See clear demarcation of bony tumour from proximal femur

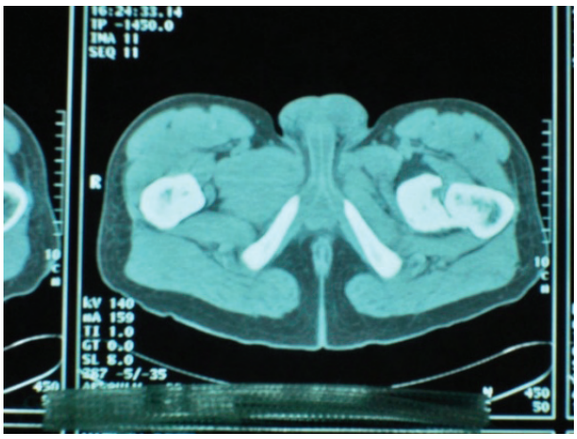


Figure 10C
MRI of bony proximal left thigh tumour

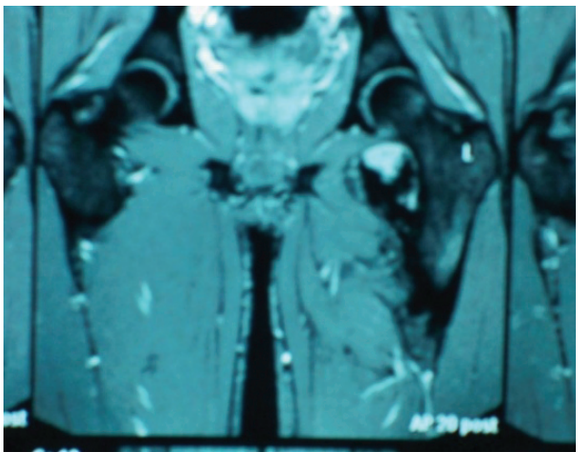


Figure 10D
MRI Angiography of bony proximal left thigh tumour. Moderate vascularity of tumour reported

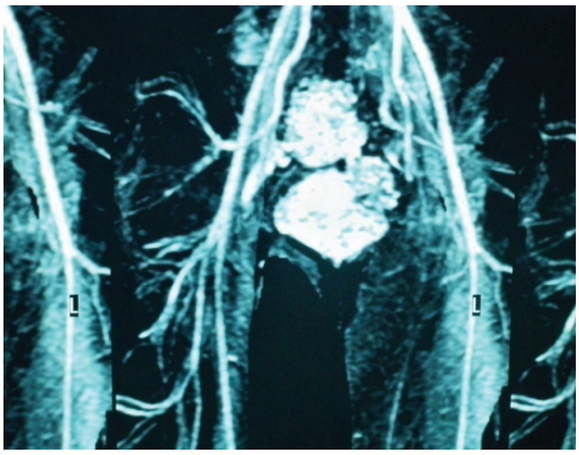
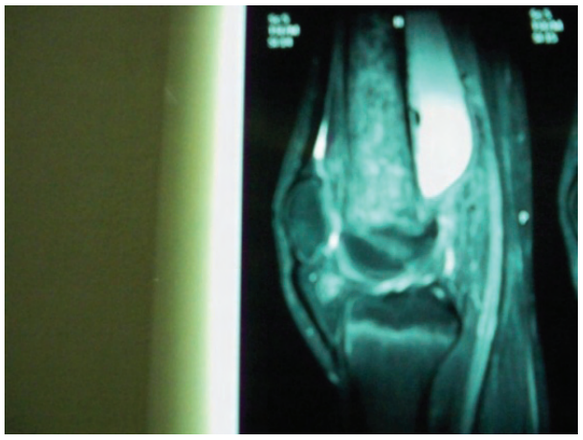


Figure 11
MRI of distal thigh showing bone cyst on posterior distal femur and periosteal reaction. Osteomyelitis in a 13 year old boy



DISCUSSION

In this study, spanning twelve years, 41 thigh tumours were operatively treated. Over the same period the in-patients admitted to ward 6D of the Kenyatta National Hospital totalled 16,430. Of these, 13465 were adults while 2965 were children. The thigh tumours thus comprised a mere 0.25% of the admissions over this period. This clearly demonstrates the rarity of thigh tumours.This study managed to fully analyze 40 patients (Table 2, Figure 1).The age range was wide with a group mean of 31.4 years, though various histopathological entities had variable mean ages. Not much meaning can be derived from the differences in gender distribution and the side affected. The thigh tumours were mainly distributed to the proximal thigh (37.5%) or the distal thigh (60%) (Figure 2), with only one mid-thigh tumour reported. Plain radiographs were the most utilized imaging modality and also the most informative (Table 3, Figure 3, 4A,5A, 6B, 7, 8, 10A). The additional information obtained from the CT scans , MRI and MRI Angiography (Figure10B,C,D) did not radically change the picture painted by the plain radiographs. That as many as 20% of the patients had had prior surgery was an outright indication of the locally recurrent nature of a number of the thigh tumours. Some of the diagnostic procedures we performed were curative as we in several instances performed outright excisional biopsies in thigh tumours that were even larger than the recommended 2-3cm diameter (Figure 6A,6B). Multiple excisional biopsies were undertaken for those tumours that recurred in close proximity to vital structures like

the sciatic nerve. Classicaly one was a case of malignant peripheral nerve sheath tumour. Additional surgery was mainly above knee amputation and this was undertaken in five cases of osteogenic sarcoma and one case of multiply recurrent malignant peripheral nerve sheath tumour. It is notable in this study that one case of osteogenic sarcoma declined any form of surgery or adjuvant therapy after the histological report and instead opted for "ALTERNATIVE" treatment elsewhere. The specialized surgery for metastatic lesions was dictated by the site of the metastases (Table 5).

It is impressive that in a population of 40 patients fully analysed, 23 histological tumour types are recorded. Osteogenic sarcoma was leading at 15%, followed by lipoma, inflammtory (osteomyelitis) and non specific tumour tissue each recording 10%.The rest of the thigh tumours recorded, 19 varieties ,all seem extremely rare(Table 6, Figure 1). A classic rarity is the case of fibrosarcoma, one case appearing in 2001 and the next one appearing in 2011, a clear ten years apart. Only a few of these tumours could be definitively diagnosed on clinical and radiological features and their definitive treatment planned out before the incisional biopsy. Such tumours are osteogenic (pleomorphic) sarcoma (Figure 2,3), nonspecific tumour tissue (Figure 4), giant cell tumour of bone (Figure 5A,5B) and fibrous dysplasia. Some of these thigh tumours had such perplexing radiological appearances that only the histopathological report of the incisional biopsy brought the diagnosis to consideration. This is exemplified by the haematoma (Figure 3) and the Non Hodgkins Lymphoma (Figure 7). In another instance the presence of dysplasia in the proximal femur coupled with a pathological fracture obscured for months the presence of a proximal soft tissue thigh tumour that eventually turned out to be a Desimoid tumour(Figure 8). Particular challenges were posed by recurrent thigh tumours that had had prior surgery elsewhere . Some of these were clear outright local recurrences of osteogenic sarcoma (Figure 9), but had to be taken through the ritual of incisional biopsy before above knee amputation , radiotherapy and chemotherapy. The major diagnostic challenges were between osteogenic sarcoma, inflammatory (osteomyelitis) and non specific tumour.They have nearly similar mean ages, with osteosarcoma averaging 11.5 years, (range 3- 20 years), while inflammtory (osteomyelitis) averaged 11 years. Non specific tumour tissues averaged 31 years. Translated to patient care, this means that distal thigh tumours in patients below 20 years must be carefully evaluated to distinguish between osteogenic sarcoma and osteomyelitis .The difference between patients who received adjuvant chemotherapy (6 patients) and those who received radiotherapy (7 patients) is not significant as some of the patients received both modalities of adjuvant therapy.

Followup of these patients revealed some very interesting trends. The shortest follow-up in this series was the lipoma patients who did not record a single return visit after the surgery.The most consistent follow-up was recorded in the osteogenic sarcoma patients while the longest recorded follow-up of 9 years was the patient of malignant peripheral nerve sheath tumour. The total dissappearance of the lipoma patients from follow-up after surgery is a reaffirmation of the benign nature of lipomas. Further , with the last lipoma having been excised in 2004, it may be safely concluded from this study that thigh lipomas are now extinct tumours (Figure 1).

The presence of four thigh tumours of non specific nature is mind boggling. It is our postulate that these could be variants of myositis ossificans(5), or advanced osteochondromas. We propose a future study to address these unique thigh tumour lesions.

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