Limb Salvage Surgery In A 17-year Old Boy With Osteosarcoma.

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Abstract

Osteosarcoma is a malignant mesenchymal tumour in which the cancerous cells produce bone matrix, hence the former name osteogenic sarcoma. It is the most common primary malignant tumor of bone exclusive of myeloma.
We present a case of a 17 yr old boy whose informed written consent was obtained for this study. The histology confirmed osteosarcoma of the left fibular head complicated by foot drop. He was managed by the Orthopaedic department, Wesley Guild Hospital, Ilesa unit of the Obafemi Awolowo University Teaching Hospital Complex. He had a limb salvage surgery with follow-up chemotherapy and radiotherapy.

Case Presentation

Master O O is a 17 yr old Nigerian student awaiting university admission. He was referred to the Orthopaedic department of Wesley Guild Hospital (WGH) Ilesa on the 24th August, 2006 from State Specialist hospital Ondo, Ondo State. An informed written consent was obtained from him and the father for this study including the use of his clinical photographs. He presented with a 10 week history of pain in the proximal third of the lateral aspect of the left leg and swelling of 8 weeks.
The pain was insidious in onset, deep seated, persistent, and radiated laterally downward associated with a tingling sensation in the foot. It was aggravated by activity especially walking long distances, but relieved by rest. Non steroidal anti-inflammatory drugs produced no sustained relief. Pain was sometimes severe enough to prevent him from performing his routine daily activities as well as sleeping when it occurred at night.
A swelling was noticed two weeks after the onset of the pain at the upper lateral aspect of his leg. The mass was initially about the patient's two apposed thumbs but had progressively increased to its size at presentation. There was no other swelling in the body. The appetite was good but he noticed weight loss. He was unable to lift the foot at the ankle and thus dragged while walking.
There was no history high grade fever or trauma to the left leg. He had no pre-existing bone disease nor was he exposed to significant radiation in the affected limb. There was no family history suggestive of Li-Fraumeni syndrome and he had no retinoblastoma in childhood.

He is the 5th child of the six children in a polygamous family. No history of Juvenile Diabetic mellitus. No history of drug allergy and he neither smokes cigarette nor drinks significant alcohol. The mother is the first wife and trades in clothing. The father works in Kaduna as a tractor technician.

Examination revealed a depressed -looking young man with a high stepping gait. The general clinical condition was satisfactory. The musculoskeletal system revealed an almost circumferential mass (20cm X 14cm) over the upper left leg, more prominent on the posterolateral aspect. The swelling had a bosselated smooth shiny skin surface, with distended veins coursing over it with differential warmth. The mass was fixed, immobile, and was not attached to overlying skin. It was stony hard and tender.

The popliteal artery could not be palpated because of the tumor bulk .The left dorsalis pedis artery was palpable and of equal volume with the contralateral side. The popliteal lymph nodes were not felt. There was no significantly enlarged inguinal lymph node.
The extension at the left knee joint was full but flexion

limited to 30 degrees. There was a left foot drop.

Investigations revealed an elevated erythrocyte sedimentation rate of 128mm/hr (Westergreen method). All other hematological and biochemical evaluations were normal.

Plain radiographs of the left leg showed the characteristic destructive lesion of the metaphysis (cortical and cancellous bone) of the left fibula head, extra osseous soft tissue sun ray appearance and a wide zone of transition between tumour and normal host bone. The chest X-ray was normal macroscopically. He could not afford CT of the lung/ left fibula and MRI. Bone scintigraphy was not available at the study centre.

Histology of biopsy tissue confirmed osteoblastic osteosarcoma of the proximal third of the left fibula put as clinical stage II. The pre-treatment and post operative radiographs are shown in Figures 1, 2, and 4. The post surgical excision of the left fibular osteosarcoma clinical photograph is illustrated in figure 3 and 5.

The surgical treatment given included a surgical wide excision of the tumour with investing capsule and 5-Fluorouracil local infiltration of the tumour bed .He was then placed on chemotherapy and radiotherapy at the University College Hospital (UCH), Ibadan.

The limb was salvaged from amputation. He is been followed up at the orthopaedic clinic at the Wesley Guild Hospital Ilesa.

DISCUSSION.

Classic high-grade osteosarcoma is a highly malignant spindle-cell sarcoma of bone in which the malignant cells produce osteoid[1]. It is the most common primary malignant tumour of bone, excluding myeloma, and is the third most common malignant disease in adolescence after leukemia and lymphoma. However, it is still a rare tumour with only 1000 new cases per year in the United States. There appears to be no racial or ethnic influence associated with its incidence [2]. However, an increase increased prevalence has been noted in families affected with the Li-Fraumeni syndrome [3] and in patients who have had retinoblastoma [4] (a 40% prevalence in those with bilateral disease), have undergone radiation therapy [5], or have Paget disease [6].

The patient presented in his second decade of life with classic high-grade osteosarcoma.The usual peak prevalence is between the ages of ten and twenty-five years [1].The male to female ratio is approximately 1.5:1. Osteosarcoma can occur in any bone but is most common in the metaphyses of long bones as seen in our patient, about 80-90% of the tumours occurring in those locations, and it often extends into the epiphysis. Pain is the most prevalent presenting symptom in osteosarcoma, occurring in 85% of patients [7]. The pain could be due to micro fractures through the involved areas of the bone or, in severe cases, to compression or stretching of adjacent anatomic structures well illustrated in the presented patient. The pain usually is exacerbated by activity, and only 21% of patients have pain at night. Almost 50% of the patients relate the onset of symptoms to an episode of minor trauma. A palpable or visible mass is noted in about 40% of patients . Painless masses may be obscured when they arise in the pelvis or proximal part of the thigh. Less common findings include a limp, weakness, and a decreased range of motion in associated joints, venous engorgement, oedema, and striae.

Laboratory tests are generally not helpful in the diagnosis of osteosarcoma, although elevated serum lactate dehydogenase and alkaline phosphatase levels have been associated with a poorer prognosis [8,9].

The plain radiographic features illustrated destructive lesion of the metaphysis of the fibula. It typically exhibits a mixture of lytic and blastic areas. Its overall appearance is that of an aggressive process characterized by destruction of both cortical and cancellous bone and a wide (permeative) zone of transition between tumour and normal host bone.Osteosarcoma usually has an extra osseous soft-tissue mass with fluffy irregular densities indicative of neoplastic bone formation. Codman triangle is not specific to osteosarcoma but indicate reaction to an aggressive process.

The staging of the patient tumour was limited without MRI or CT scan due to patient's financial constraint. The chest radiograph was screened for pulmonary metastases.MRI is the current standard technique for determining the local extent of the disease in the involved bone. It defines the marrow extent of the tumour, and is therefore useful for determining the appropriate resection level at surgery [10]. The sagittal or coronal images of the entire bone visualize skip metastases. Axial imaging provides accurate depiction of soft-tissue mass extent, relationship to neurovascular structures, as well as adjacent joint involvement. Bone scintigraphy plays a major role in staging by defining areas of primary involvement, skip metastases, and sites of synchronous metastases to other parts of the skeleton. Lymphatic spread is so uncommon that routine examination of lymphatics is not done.

Successful surgical management is predicated on attaining wide surgical margins. This was accomplished by limb-sparing resection and not the usual 100% limb amputation .There does not appear to be any significant difference in long-term survival between patients who undergo amputation and those who have a limb sparing procedure provided that wide margins are obtained [11]. Limb-salvage surgery is indicated for patients in whom wide margins can be obtained without sacrificing so much tissue that the remaining limb is nonfunctional. The major nerves and major vessels were preserved in the patient. The leg reconstruction was done, retaining the function to walk

unaided during the post operative period. He was treated with neoadjuvant chemotherapy (methotrexate) and a post operative course of adjuvant combination chemotherapy (doxorubicin, methotrexate and cisplatin).A loco-regional radiotherapy is still being given to the affected limb.

Current management protocols provide long-term survival rates of between 60% and 80% for patients without clinically apparent metastatic disease at presentation[12, 13] .The survival rate for patients who have metastases at the time of presentation is between 10% and 20% [14, 15.When] pulmonary metastases develop after completion of therapy and the metastases can be resected, a five-year survival rate of 20% to 40% can be expected [16]. The single most important predictive factor in osteosarcoma is the presence or absence of detectable metastatic disease at presentation. The presented patient's prognosis is expected to be good because of absence of detectable metastases, resectability of the patient's tumour, none elevated level of alkaline phospatase and serum lactate dehydrogenase . Also, the location of the tumor away from the pelvis, proximal part of the femur, and proximal part of the humerus confers a favourable prognosis [17,18].

CONCLUSION.

Limb salvage surgery in suitable patients with osteosarcoma is more favored in the present Bone and Joint Decade. The prognosis of osteosarcoma is very good when the patient presents early without metastases allowing limb-sparing surgery combined with adequate chemotherapy and radiotherapy-an especially poignant issue in a country without customized endoprostheses.

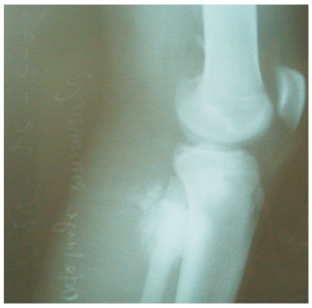

Fig. 1: X-Ray Showing Left Fibula Osteosarcoma

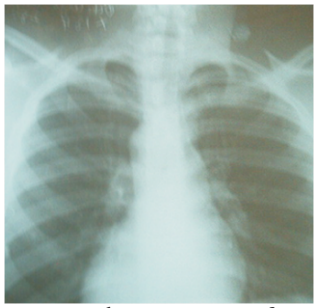

Fig.2: Chest X-Ray of patient at presentation

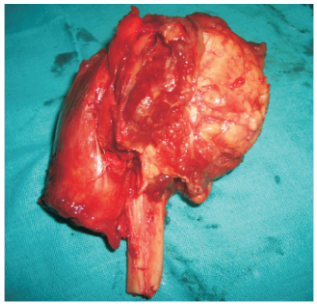

Fig.3: Post Surgical Excision of the Left fibula Osteosarcoma

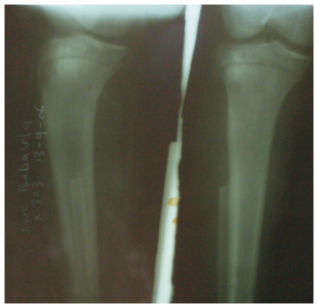

Fig.4: Post operative X-Ray of left leg.

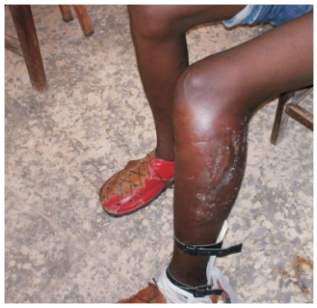

Fig.5: 10day Post operative state of left leg with anti foot drop splint

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