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CASE REPORT

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Unusual Presentation of Ewing Sarcoma in a Nigerian Adolescent: A Case Report and Literature Review

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Abstract

Ewing sarcoma is a malignant, aggressive small, round, blue cell tumour, likely of neural crest origin. It is the second most common bone cancer in children, usually affecting the long bones and central axis. The involvement of flat and irregular bones is rare. We present a Nigerian adolescent male with a three-year history of left foot pain following a minor trauma, a one-year history of foot swelling, and progressive swelling in the left lower limb, scalp, and jaw over seven months. A foot ulcer developed a few days after biopsy. A duplex ultrasound scan revealed multiple enlarged inguinal and popliteal lymph nodes. A head CT scan showed a solitary lytic lesion in the right posterior parietal bone. An MRI of the left foot demonstrated a large soft tissue mass on the first metatarsal with cortical destruction. An initial histopathology diagnosis with open biopsy was Bacillary angiomatosis. Histology of a second open biopsy specimen from the affected foot showed mainly granulation tissues, but a subsequent USS-guided core-needle biopsy of the scalp and jaw masses done one month later revealed a small, round, blue cell tumour. Immunohistochemistry confirmed *Ewing sarcoma*. The patient responded remarkably to multi-agent chemotherapy, with significant tumour regression and symptom relief. Flat and irregular bone involvement in Ewing sarcoma is rare but possible. This highlights the importance of maintaining a high index of suspicion, utilising appropriate imaging, and ensuring adequate tissue sampling for accurate diagnosis and effective treatment.

Keywords: Bone biopsy, Blue Cell Tumour, Ewing sarcoma, Flat bones, Magnetic Resonance Imaging.

Introduction

Ewing sarcoma family of tumours refers to a group of malignant small, round, blue cell tumours, likely of neural crest origin, that typically affect bones and soft tissues.[1] The anatomic sites of primary tumours are the long

bones and the central axis. Involvement of the flat bones of the skull and irregular bones of the jaw is rare.[2] Metastasis commonly occurs in the lungs, bones, and bone marrow. [3] It parades as the second most common bone cancer in children and adolescents. [4] It is a rare tumour and even rarer in people of African descent, with an annual incidence of about 1.6 cases per 1million people in the white population and about 0.2 cases per 1 million people in the African Americans. [4,5] This case is reported to create awareness about this rare condition which was initially misdiagnosed. This is to highlight the need for a high index of suspicion and adequate tissue sampling for histological diagnosis.

Case Presentation

A 16-year-old Nigerian adolescent male with a three-year history of left foot pain following a minor trauma, a one-year history of left foot swelling, and progressive swelling in the left lower limb, scalp, and jaw over seven months, with high-grade fever presented at the University of Nigeria Teaching Hospital, Ituku-Ozalla, Enugu, Nigeria.

Serial full blood count showed anaemia and leucocytosis, for which the patient received blood transfusions and antibiotics. Examination revealed significant, multiple, firm, and tender matted left inguinal and femoral lymphadenopathy, measuring 4-8cm. An aggressive foot ulcer developed a few days after an open biopsy was taken from the left foot swelling. He was initially managed in another centre, where a histopathologic diagnosis of Bacillary angiomatosis was made. But on presentation at our facility, a wedge biopsy was taken from the site of the foot ulcer for second-opinion histology. This showed granulation tissue with no evidence of malignancy. This tissue may not have been representative enough as it was taken from an ulcer base. One month into admission, a repeat biopsy was performed under ultrasound (USS) guidance (as recommended by the multidisciplinary oncology group). The USS-guided core needle biopsy retrieved strands of tissue from both the scalp and the jaw masses, and histology reported a small, round, blue cell tumour. Immunohistochemistry confirmed Ewing sarcoma. The patient responded remarkably after two cycles of multi-agent chemotherapy (VAC+IE) regimen [Vincristine, Adriamycin, Cyclophosphamide, Ifosfamide and Etoposide], with significant tumour regression and symptom relief, as jaw, scalp, lower limb, and lymph node swelling began resolving dramatically, pain subsided, and the ulcer started contracting. The VAC + IE regimen was used intravenously every three weeks, alternately, at the doses of Vincristine 1.4mg/m^2, Adriamycin 75mg/m^2, Cyclophosphamide 1000mg/m^2, Ifosphamide 1.8g/m^2, and Etoposide 100mg/m^2. He was subsequently discharged home and is currently being followed up every three weeks.

Details of the Radiological Investigations

Duplex ultrasound scan: Multiple enlarged left inguinal and popliteal lymph nodes and marked compression of the femoral and popliteal vessels.
Chest X-ray: Suspected right scapular sclerotic lesion. No obvious lung metastasis.
Head CT scan: A solitary lytic mass centred on the right posterior parietal bone. This lesion shows extracranial and intracranial extra-axial soft tissue components. No intra-axial lesion seen. These features suggest metastasis.
MRI scan of the left foot: Large soft tissue mass of the left foot centred around the first metatarsal (8.4cmx5.7cmx5.5cm) with solid and cystic components. The lesion shows rim enhancement. The mass encases the first metatarsal bone and shows cortical destruction and altered marrow intensity.

Assent and consent were obtained from the child and the parents with the permission to use his data and images in this research. The parents felt the child got the best care available following

appropriate diagnosis. The family was satisfied with the care received, especially with the obvious progressive improvements in the child's clinical condition.

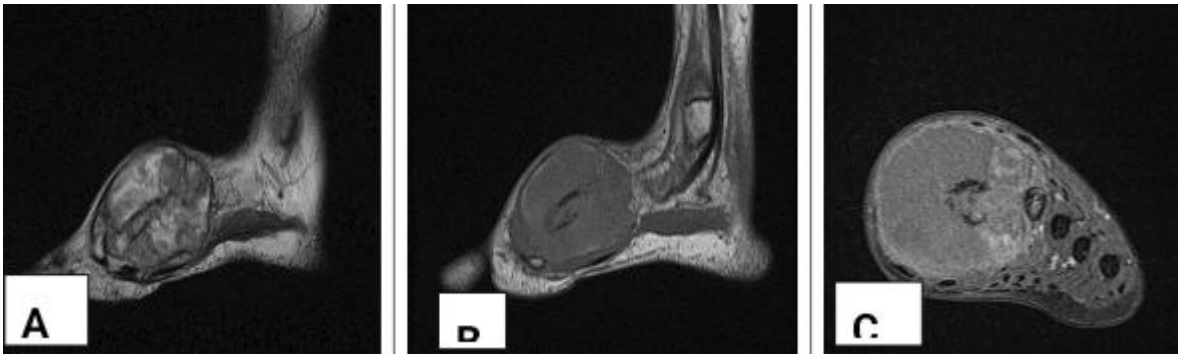

Figure 1: MRI of the left foot.
(A) T2 Sagittal FSE image of the left foot showing the mass lesion encompassing the first metatarsal bone.
(B) T1 non-contrast FSE Sagittal MR image of the left foot showing the mass lesion encompassing the first metatarsal bone.
(C) Coronal T1 fat-sat with contrast MR image of the left foot showing the mass lesion encompassing the first metatarsal bone.
MRI - Magnetic Resonance Imaging; FSE - Fast Spin Echo; Fat-sat - Fat saturation.

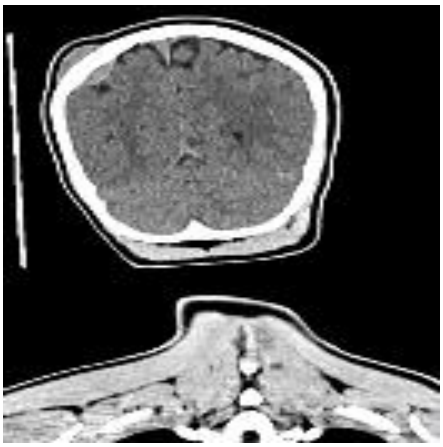

Figure 2: Head CT scan.
Coronal head CT image in standard window showing extracranial and intracranial extradural right parietal soft tissue mass caused by the underlying bone-destructive lesion.
CT - Computed Tomography.

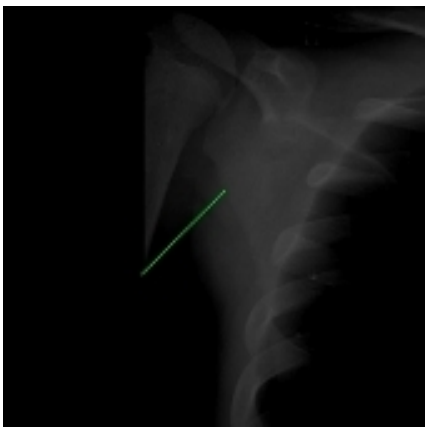

Figure 3: Chest plain radiograph
PA Chest radiograph (close-up view) showing a sclerotic focus on the lateral aspect of the right scapula (arrowed).
PA – Posteroanterior.

Discussion

Ewing sarcoma is an uncommon bone cancer which affects males more than females. Children and adolescents, aged 5 years to 20 years, are predominantly affected, with the median age of diagnosis being 15 years. [6] The incidence of Ewing sarcoma is significantly higher in people of European ancestry compared to those of African ancestry. The reason for this disparity is not apparent. Still, it may be due to genetic differences, such as a specific polymorphism linked to an increased risk of Ewing sarcoma, which is found more frequently in whites than in

blacks. [7] It involves long bones and rarely affects flat and irregular bones. The involvement of the skull bones is particularly uncommon, accounting for approximately 1% of all Ewing's sarcomas, and the temporal bone is most commonly affected, followed by the frontal and parietal bones. [8] Unusual presentation of Ewing sarcoma usually leads to misdiagnosis with its attendant treatment delay, which negatively impacts the outcome.

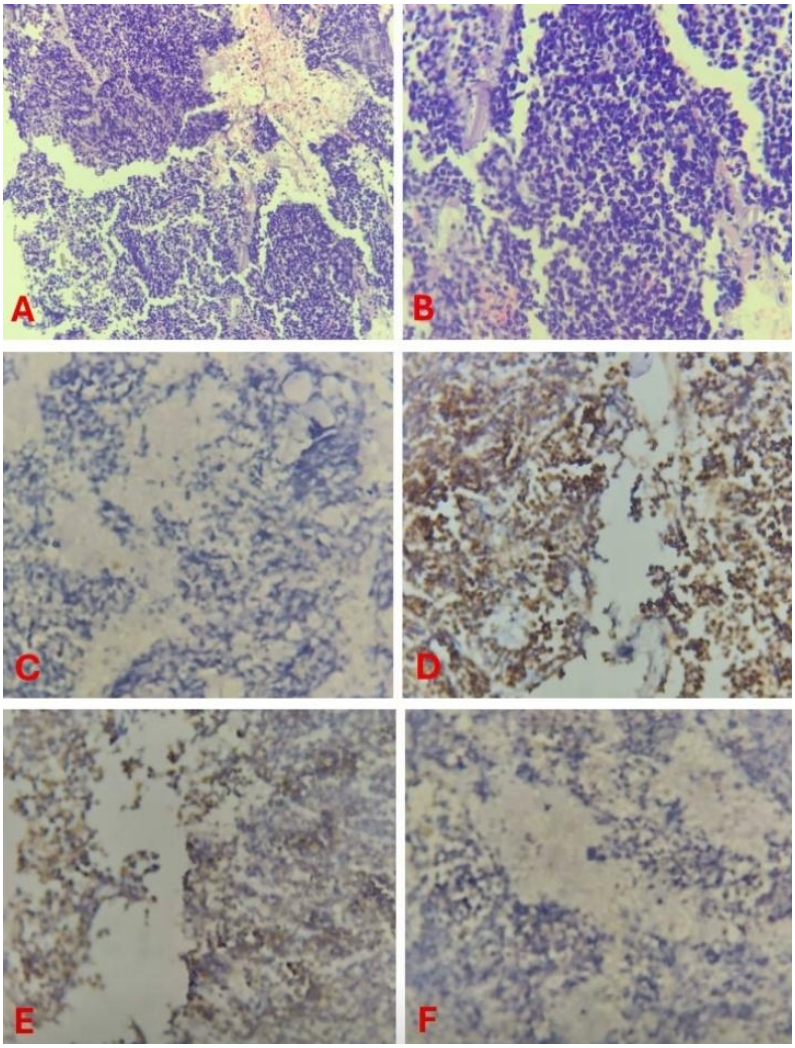

Figure 4: Photomicrographs of Ewing sarcoma in a 16-year-old male
(A) Note sheets of small, round, uniform cells divided into irregular lobules by thin fibrous strands (H&E x200); (B) High power view showing lesional cells with round, hyperchromatic nuclei with scanty cytoplasm (H&E x400); (C) Negative Pan-Cytokeratin (PanCK) stain: note lack of staining of lesional cells (PanCK IHC, x400); (D) Strong and diffusively positive CD99 stain: note membranous staining (golden brown coloration) outlining lesional cells (CD99 IHC, x400); (E) Moderate and patchily positive Synaptophysin stain: note cytoplasmic staining (golden brown coloration) of portions of lesional cells (Synaptophysin IHC, x400) (F) Negative CD45 stain: note lack of staining of lesional cells (CD45 IHC, x400).
H&E - Haematoxylin and Eosin; IHC – Immunohistochemistry.

Clinico-laboratory features include local pain, soft-tissue swelling, limitation of motion, weight loss, fever, leucocytosis, and elevated erythrocyte sedimentation rate. [2] Metastasis commonly

occurs to the lungs, bones, and bone marrow and connotes a poor prognosis.[2] In the index case, the characteristic radiologic features of Ewing sarcoma were present, including aggressive periosteal reaction, permeative lytic lesion, large soft-tissue mass, cortical destruction and frequently sclerotic reactions. Due to its close clinical and radiologic features, Ewing sarcoma can often be misdiagnosed as osteomyelitis, osteosarcoma, osteoma, giant cell lesion, lymphoma, cartilaginous tumour, or Bartonelosis. Differentiating them requires a comprehensive evaluation using multiple imaging modalities and microbiological testing. A CT scan can show a large soft-tissue mass of the foot, permeative lytic lesions of the bone, with aggressive periosteal reaction and cortical destruction. However, distinguishing between osseous remnants, reactive changes, and tumour matrix can be challenging. Images from a T2-weighted MR scan will not sufficiently differentiate tumours from necrosis, and lesion boundaries are often overestimated because of the presence of oedema and haemorrhage. This challenge is overcome with an MRI scan with contrast, wherein the rim enhancement pattern following contrast administration allows distinction between tumour and peritumoral oedema. Additionally, an MRI scan usually differentiates the large soft-tissue sarcomatous mass with a solid and cystic component around the involved bone from the abscess or oedema of osteomyelitis. However, to make a specific diagnosis, a biopsy of the tumour is required, with histology and immunohistochemistry to confirm it. The importance of having a biopsy with an adequate amount of representative tissue cannot be overstressed. In the current case, the initial biopsy was not representative (as it was taken from an ulcer base on the foot), and the diagnosis was not clinched until an USS-guided core needle biopsy was performed, with adequate, representative tissue strands obtained from the jaw and skull swelling.

In this report, we demonstrate a scalp and jaw lesion with morphological features in keeping with 'classic' Ewing sarcoma on Haematoxylin and Eosin, which was subsequently confirmed with an immunohistochemistry. To document local or distant metastasis of the tumour, staging before biopsy is important. In Ewing sarcoma, metastasis may involve the lungs alone, which is the most common site of metastasis.[9] But it could also involve the bone or bone marrow alone, skip metastases, or combined metastatic disease. [3,10,11] Bone metastasis was suggested by a CT scan of the skull, which showed a solitary lytic mass on the right posterior parietal bone. In contrast, the involvement of the bone marrow was demonstrated by MRI of the left foot, which showed marrow intensity alteration in the first metatarsal. For Ewing sarcoma, MRI is the preferred imaging modality due to its high sensitivity in detecting tumour spread to bone and bone marrow. MRI's accurate assessment of tumour extent is crucial for guiding treatment decisions, making it an essential tool in evaluating this condition.

Limitations

Although a duplex USS was suggestive of lymph node involvement, biopsy of the suspicious nodes would have been desirable to confirm lymph node metastasis. The observed lymph node enlargements could have been reactive and not tumorous. A biopsy of the metatarsal bone would also have confirmed the diagnosis and established a primary tumour site.

Conclusion

Presentation of Ewing sarcoma with the involvement of the flat and irregular bones is rare and uncommon. This case highlights the need for a high index of suspicion, utilising appropriate imaging modalities, and ensuring adequate and representative tissue sampling for diagnosis and treatment.

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Authors' Contributions: UKO, OIA, ODB conceived and designed the study. EKN, INI and OAC curated and interpreted the data. UKO, CBF, and OJG drafted the manuscript, and EOU and CBF revised it for sound intellectual content. All the authors approved the final version of the manuscript.

Conflicts of Interest: None declared.

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