

ATYPICAL CLINICAL PRESENTATION OF CHILDHOOD OSTEOSARCOMA

CASE REPORT

ABSTRACT

Aim: Highlight childhood Osteosarcoma as uncommon in a pre-adolescent female's scapula.

Presentation of case: 9-year-old female with left scapular outgrowth, weight loss, paleness, cachexia and axillary lymphadenopathy. Mass measured 27 by 53 cm, hard, tender with smooth, shiny unattached skin. Shoulder and Chest CT showed huge, well demarcated, heterogeneous 14.6cm by 13.4cm by 14.4cm soft tissue mass with lobulated margins on the left scapular with its lytic destruction. Multiple nodules were seen on ipsi-lateral lung involving adjacent pleura. Malignant mesenchymal moderately pleomorphic cells with large pleomorphic hyperchromatic occasionally mitotic nuclei in a fibrous background with neoplastic lacey bone trabeculae, alternating hypercellular and hypo-cellular areas, dilated congested vessels were visualized on histology. Metastatic Osteosarcoma was diagnosed. Neo-adjuvant chemotherapy was given. Surgical tumor debulking awaited.

Discussion: The peak age for childhood Osteosarcoma is reported as 10–14 years for females and 15–19 years in males which coincides with the adolescent growth spurt in both sexes (Tanner stage 3 or Sexual Maturation Rating (SMR) 3 in girls). Our pre-adolescent patient is noteworthy hence our documentation. In children Osteosarcoma occur primarily in the metaphyseal region of tubular long bones with 42% in the femur, 19% tibia and 10% humerus. Rarely in skull, pelvis or scapular (8%). The inflammatory features seen in osteosarcoma mimic osteomyelitis, cellulitis, trauma and benign bone tumors as strong differentials. Our patient's late presentation features rather mimic our diagnosis. Our patient's histology support Osteoblastic Osteosarcoma with high grade histology. A variant previously reported in sacrum, iliac crest and pubis. Late presentation seen has been cited as a major factor affecting surgical method as it will in this case. Otherwise limb salvage surgery with chemotherapy remains the management with best outcome.

Conclusion: Scapular Osteosarcoma in a pre-adolescent female is an uncommon presentation, hence this report.

Keywords: Osteosarcoma, Scapula, Pre-adolescent, Female, Atypical.

Introduction:

Osteosarcoma is the most common primary malignant bone tumor in children and adolescents globally¹ accounting for 21.6% of bone tumors in children aged 0-14 years in Enugu Nigeria². It accounts for about 3% of pediatric cancers globally and has a bimodal age distribution worldwide³. The first peak incidence occurs during the second decade of life at the adolescent growth spurt while the second peak is in older adults during the seventh and eighth decades of life^{3,4}. Generally males and African-American children are more affected than females and Caucasians respectively³. However peak age and incidence rates coincide with pubertal changes in bone growth, hormones and development in both sexes. Thus a later age and higher rates in males (age 15–19, peak rate of 9–15 cases/million population) compared to females (age 10–14, peak rate of 6–10 cases/million population) have been reported⁵

The etiology of Osteosarcoma in most patients is unknown however; the predilection for adolescent age of growth spurt and sites of maximum growth suggests a relationship with rapid bone proliferation⁶. Certain genetic conditions predispose to development of osteosarcoma; hereditary Retinoblastoma, Li-Fraumeni syndrome, Rothmund Thompson syndrome⁷. Also exposure to alkylating agents and irradiation has been linked to the development of the tumor⁷.

Osteosarcoma is defined by the presence of malignant mesenchymal cells-producing osteoid or immature bone. Its origin is majorly in the metaphyseal region of tubular long bones, with 42% occurring in the femur, 19% in the tibia, and 10% in the humerus⁸. Other common locations are the skull and pelvis which occur in about 8% of osteosarcoma cases⁸. In adults the tumour usually involves the axial skeleton and flat bones and occur as a result of malignant transformation from pre-existing conditions like Paget disease, irradiation, osteochondroma and other benign bone processes⁹

The common presenting features are pain, limping and swelling which can be easily dismissed as minor trauma due to the active nature of the age group. Plain X-ray, bone scans and magnetic resonance images are required for diagnosis however confirmation is by biopsy of the lesion.

Chemotherapy and ablative surgery of primary tumor are the main stay of treatment⁷. Prognosis depends on the type, presence of metastasis and location of primary tumor. Up to 75% of patients are cured in non metastatic extremity tumors⁷.

We present this notable case of Osteosarcoma of the scapular in a pre-adolescent 9-year old female.

Aim:

Create awareness that childhood Osteosarcoma may occur at such uncommon sites as the flat bone of the scapular, a site reported more in adult patients.

Presentation of case:

A is a 9-year-old female who presented to the Children' Emergency Room of the Enugu State University Teaching Hospital, Parklane, Enugu, Enugu State, Nigeria, with a progressive, painful outgrowth over her left scapular noted three months prior to presentation, progressive weight loss that started two months prior to presentation and paleness of the body noted four days prior to presentation. The patient had visited a private hospital where excision biopsy was done before presentation. She had no prior exposure to alkylating agents and irradiation, no family or personal history of associated genetic syndromes.

At presentation, she was cachectic, pale, with significant axillary lymphadenopathy. She had a huge oval mass covering the entire left scapular and extending to the shoulder and the axilla and measuring about 27 by 53 cm in its widest diameter. Mass was firm to hard and tender to touch with a smooth, shiny unattached skin surface and a transverse incision scar across its longest diameter. Patient weighed 24.5kg which falls between 10th and 25th centile on the CDC chart. Respiratory system findings were essentially normal. Presumptive diagnoses of Rhabdomyosarcoma and Neuroblastoma were made. She was admitted and stabilized with antibiotics, analgesics, hematinics, intravenous fluids and several courses of blood transfusion.

Laboratory tests (Complete blood count, serum Electrolytes/Urea/Creatinine, Liver function) and Radiological investigations (posteroanterior Chest radio-graph and shoulder CT scans) were done. Complete blood count showed elevated total white cell count with neutrophil predominance, elevated platelet count and moderate anaemia. Serum electrolyte urea and creatinine results were within normal range while Liver function test showed elevated total bilirubin and aspartate transaminase, the viral screenings (HIV, HBV and HCV) were negative. Shoulder and Chest CT showed relatively huge, well demarcated, heterogenous soft tissue mass with lobulated margins that me

asures 14.6cm by 13.4cm by 14.4cm and is centered on the left scapular with its lytic destruction. Apparent involvement of adjacent pleura noted. Multiple ipsi-lateral, randomly distributed pulmonary nodules seen. An impression of advanced soft tissue sarcoma of the scapular region with lytic bone destruction and pulmonary metastasis (solitary fibrous tumor r/o osteosarcoma) was made.

Histopathology report: showed sections of malignant mesenchymal proliferation consisting of moderately pleomorphic cells in a fibrous background with neoplastic lacy bone trabeculae. Also there were hyper cellular areas alternating with hypo cellular areas. Dilated congested vascular channels were present. Malignant cells with moderate atypia, large pleomorphic hyperchromatic nuclei and occasional mitosis were visualized.

On account of the above findings, a final diagnosis of metastatic Osteosarcoma was made. Following extensive counseling of the family and work up, Neo-adjuvant chemotherapy with Doxorubicin and Cisplatin was commenced. Patient received three 21-day cycles of Doxorubicin daily on days 1-3 and Cisplatin on day one. Chemotherapy was well tolerated. Tumor size reduction was noted following commencement of Neo-adjuvant chemotherapy as diameter reduced to 29cm by 34cm. She is currently awaiting surgical tumor debulking and recommencement of chemotherapy.

Figure 1 : Morphology of disease



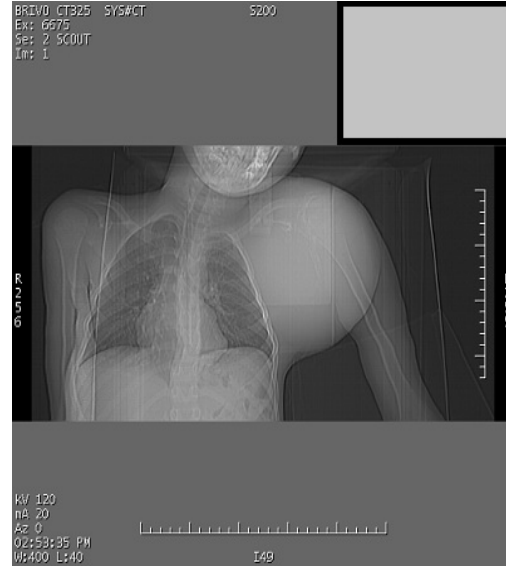
Fig.1a: External appearance of the osteosarcoma (back).



Fig.1b: External appearance of the osteosarcoma (front).



(A)



(B)

Fig.2: Morphology of the osteosarcoma. (CT-Scan)

Discussion:

The peak age for childhood Osteosarcoma is reported as 10–14 years for females and 15–19 years in males⁵. Our patient's slightly earlier age of presentation is thus noteworthy hence this documentation. This peak age coincides with the adolescent growth spurt in both sexes. The adolescent growth spurt occurs in consonance with Tanner stage 3 or Sexual Maturation Rating (SMR) 3 in girls, a stage characterized by breast and areola enlargement as well as darker, curly pubic hair¹⁰. However, our pre-adolescent index patient's SMR is not in consonance with the reported normal variants, thus our interest.

We report this unlikely presentation of childhood Osteosarcoma in the scapula, which is at variance with sites reported in children such as 42% in the metaphyses of the femur, 19% in the tibia, and 10% in the humerus⁸. Other common locations are the skull and pelvis which occur in about 8% of osteosarcoma cases⁸. Generally, scapular tumors are rare in childhood^{11,12} although Osteochondroma, Osteoid Osteoma, Chondrosarcoma, Osteosarcoma, Ewing's sarcoma⁴ may develop. The inflammatory features; pain and tenderness in the affected limb or bone, swelling and a noticeable mass in the arm, leg or area⁷, restriction of movement and differential warmth seen in oste

osarcoma as in the index case mimic osteomyelitis, cellulitis, trauma and benign bone tumors as strong differentials^{13,14}. These therefore would have been our presumptive diagnoses but for the patient's late presentation thus its features rather mimic our preferred presumptive diagnoses.

Our patient's histology with mitosis, background bony trabeculae and pleomorphic cells support Osteoblastic Osteosarcoma with high grade histology¹⁵ over the chondroblastic, telangiectatic and fibroblastic subtypes of conventional osteosarcoma⁴. This variant has equally been reported in uncommon areas of the axial skeleton such as sacrum, iliac crest and pubis¹⁵. The classic radiographic appearance described as sun burst pattern clearly suggests the aggressive bone-forming nature of the lesion¹¹. Thus histopathological and radiological evidences are both diagnostic and supportive in our index patient.

The late presentation seen in our patient, common in resource-scarce settings like ours has been cited as a major factor underlying the choice of management, necessitating radical surgical procedures such as 'above knee' amputation in some instances¹⁶. Our patient who has had the benefit of chemotherapy will likely get surgery and post surgical chemotherapy since limb salvage surgery or amputation in addition to chemotherapy is the reported management with best outcome⁷. Hopefully this will extend her mean survival time despite the poor prognostic index which her late presentation pose.

Conclusion:

Osteosarcoma in a pre-adolescent female as well as its affection of the scapular are uncommon presentations of this relatively rare childhood tumour, so should be reported.

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Authors' contributions: Author N.U.F wrote the initial draft of the manuscript, did the literature searches and took the photographs, Author C.I.E designed the study, reviewed the draft and wrote the final manuscript with the references. Both authors read and approved the final manuscript.

Consent :Written consent was obtained from the Institution’s Ethics Review Board and the patient’s mother before this report.

Ethical approval: This was obtained from the hospital’s Ethics Committee.

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