

ATYPICAL CLINICAL PRESENTATION OF CHILDHOOD OSTEOSARCOMA

CASE REPORT

ABSTRACT

Aim: Create awareness that childhood Osteosarcoma may occur in pre-adolescent females and at uncommon sites like the flat bone of the scapular, a site reported more in adult patients.

Case presentation: 9-year-old female with three months history of left scapular outgrowth, weight loss, two months and paleness of the body. She was cachectic, pale, with significant axillary lymphadenopathy. Mass measured 27 by 53 cm, hard and tender to touch with a smooth, shiny unattached skin surface and a transverse incision scar across its longest diameter. Shoulder and Chest CT showed relatively huge, well demarcated, heterogenous soft tissue mass with lobulated margins that measures 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 ipsilateral, randomly distributed pulmonary nodules seen. Malignant mesenchymal moderately pleomorphic cells in a fibrous background with neoplastic lacy bone trabeculae, hypercellular areas alternating with hypocellular areas, dilated congested vascular channels, malignant cells with moderate atypia, large pleomorphic hyperchromatic nuclei and occasional mitosis 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 males⁵ which coincides with the adolescent growth spurt in both sexes (Tanner stage 3 or Sexual Maturation Rating (SMR) 3 in girls)¹⁰. Our affected pre-adolescent patient is noteworthy hence our interest and documentation. In children Osteosarcoma occur primarily in the metaphyseal region of tubular long bones with 42% in the femur, 19% tibia and 10% humerus⁸. Rarely in skull, pelvis or scapular (8%)^{8,11,12}. The inflammatory features seen in osteosarcoma⁷ mimic osteomyelitis, cellulitis, trauma and benign bone tumors as strong differentials^{13,14}. Our patient's late presentation features rather mimic our diagnosis. Our

patient's histology support Osteoblastic Osteosarcoma with high grade histology^{4,15}. A variant previously reported in sacrum, iliac crest and pubis¹⁵. Late presentation seen has been cited as a major factor affecting surgical method¹⁶ as it will in this case. Otherwise limb salvage surgery with chemotherapy remains the management with best outcome⁷.

Conclusion: Scapular osteosarcoma in a pre-adolescent female is an uncommon presentation, hence the need to report this case.

Keywords: Scapular-Osteosarcoma, 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. The tumor accounts for about 3% of **paediatric** cancer cases 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, osteochondromas 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.

CASE PRESENTATION

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 radiograph 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, heterogeneous soft tissue mass with lobulated margins that measures 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 ipsilateral, 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 lace bone trabeculae. Also there were hypercellular areas alternating with hypocellular 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



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 consonant with Tanner stage 3 or Sexual Maturation Rating (SMR) 3 in girls, a stage characterised by breast and areolar enlargement as well as darker, curly pubic hair¹⁰. However our pre-adolescent index patient's SMR is not in consonant with the reported normal variants, thus our interest.

In children Osteosarcoma occur primarily 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. The scapular is a small bone in which many tumors namely; Osteochondroma, Osteoid Osteoma, Chondrosarcoma, Osteosarcoma, Ewing's sarcoma and others⁴ can develop. However scapular tumors are rare in childhood^{11,12}. We report this unlikely presentation of childhood Osteosarcoma in the scapular. 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 osteosarcoma 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.

Osteosarcoma has 4 pathological subtypes of conventional osteosarcoma: osteoblastic, chondroblastic, telangiectatic and fibroblastic⁴. Our patient's histology with mitosis, background bony trabeculae, pleomorphic cells support Osteoblastic Osteosarcoma with high grade histology¹⁵. The classic radiographic appearance described as sun burst pattern clearly suggests the aggressive bone-forming nature of the lesion¹¹. This variant has equally been reported in uncommon areas of the axial skeleton such as sacrum, iliac crest and pubis¹⁵. Thus histopathological and radiological evidences serve as both diagnostic and supportive evidences 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. Limb salvage surgery or amputation in addition to chemotherapy remains the management with best outcome⁷. Our patient had the benefit of chemotherapy and is been worked up for surgery which will be followed up with post surgical chemotherapy. Hopefully to extend the mean survival time inspite of the poor prognostic index which her late presentation pose.

CONCLUSION

Scapular osteosarcoma in a pre-adolescent female is an uncommon presentation, hence the need to report this case.

CONSENT FOR PUBLICATION

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

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