Case Report

Extraskeletal Osteosarcoma: A Case Report

Ndubuisi Okoh[1], *Mustapha Usman Ibrahim[2], Muhammad Nuhu Salihu[1], Yusuf Ibrahim[3]

[1]Department of Clinical Services, National Orthopedic Hospital, Dala, Kano, Nigeria. [2]Department of Surgery, Bayero University Kano and Aminu Kano Teaching Hospital, Kano, Nigeria. [3]Department of Histopathology Bayero University Kano and Aminu Kano Teaching Hospital, Kano, Nigeria.

Abstract

Extraskeletal osteosarcoma (ESO) is a rare form of osteosarcoma that has no attachment to bone and can be found in the thigh, retroperitoneum, chest, breast, orbit, retropharyngeal space etc. Literature about ESO is still mainly case reports and case series with few reported cases in our environment. We therefore report this 52-year-old with swelling at the left shoulder which progressed over a 3-year period. Pre-operative evaluation (both clinical, radiological and histopathological) were in favor of soft tissue sarcoma. A wide local excision showed no bony attachment intra-operatively. Histology of the specimen confirmed ESO. Patient had adjuvant chemotherapy and was followed up for 14months before this report with no sign of local recurrence or distant metastases.

Keywords: Extra skeletal; Osteosarcoma; Nigeria.

Introduction

Extraskeletal osteosarcoma (ESO) is a rare form of osteosarcoma that has no attachment to bones.[1] It is a form of soft tissue sarcoma with malignant mesenchymal neoplasm capable of osteoid, bone or chondroid matrix production, located in the soft tissues and without connection to the skeleton, as determined by radiographic and intra-operative examination.[2] It accounts for about 1% of all soft tissue sarcomas and 4% of all osteosarcomas.[3] It is very rare in children and most report found it to be common after 50yrs of age.[4] It commonly affects the limbs especially the thigh, but it has also been reported in the retroperitonium, chest, breast, orbit, retropharyngeal space etc.[5]ESO is a rare tumor worldwide and most publications on it are case reports and case series. In Nigeria, the largest case series is that of Alonge et al who found a total of 5 ESO cases amongst 112 sarcoma patients over a 20-year period.[6] Few other reports of ESO in Nigeria have been reported, hence, we report a case in a middle aged man who was managed at the National Orthopedic Hospital, Dala, Kano.

Access this article online

Quick Response Code:

Website: www.nigerianmedjournal.org

Case Summary

YAN is a 52-year- old lecturer who was first seen at the National Orthopedic Hospital, Dala Kano on the 6th of August, 2020 with complain of left shoulder swelling of 3years duration. The swelling was initially about 2cm in diameter, but progressively increased in size to about 30cm at presentation. There was associated pain over the preceding 6months which was worse at night. No skin changes or ulcerations. He had no anorexia or easy fatigability. No history suggestive of metastasis. No preceding history of trauma, prior exposure to irradiation or exposure to agro/ industrial chemicals or heavy metals. He had no history of prior treatment for a malignancy or family history suggestive of malignancy. He is a known hypertensive but controlled. He had no other comorbidities.

Corresponding Author: *Mustapha Ibrahim Usman Surgery Department, Bayero University Kano and Aminu Kano Teaching Hospital, Kano, Nigeria mustyibro90@gmail.com, miusman.sur@buk.edu.ng

How to cite this article: Okoh N, Ibrahim MU, Salihu MN, Ibrahim Y. Extraskeletal Osteosarcoma: A Case Report. Niger Med J 2022;63;(1): 86-90

On examination, there was a firm to hard fusiform mass measuring 20cm in its widest diameter at the posterolateral aspect of the upper arm, non-tender, attached to underlying structures but not to overlying skin. The distal neurovascular status was normal.

Plain radiographs of the left shoulder showed normal joint outline, no bony abnormalities or focal lesions and increased soft tissue shadow with no calcifications. See fig 1.

On Magnetic Resonance Imaging (MRI), the tumor showed a relatively well-circumscribed heterogeneous mass, which was isointense to skeletal muscle on T1-weighted sequence and hyperintense relative to skeletal muscle on T2-weighted sequence.

The pre-operative Tru-cut needle biopsy was suggestive of soft tissue sarcoma, but pathologist advised a repeat biopsy, illustrating the difficulty with making a definitive diagnosis using tru-cut needle biopsy technique. This has been well illustrated by Kasraeian et al in their study.

Having made a preoperative diagnosis of stage III soft tissue sarcoma, the patient was counseled and had wide local resection done on the 17thof August 2020.

The intra-operative finding was that of a firm 12 x 18cm mass with a well-defined capsule attached to the humeral periosteum distal to the deltoid insertion,as well as to the superficial aspect of the rotator cuff proximally.

A wide local excision with a 20mm margin beyond the pseudocapsule was done with enbloc excision of the biopsy scar. See figures II – VI.

The specimen was sent for histology. The histology report revealed sections showing sheets of mesenchymal cells and stroma invested in fibrous pseudocapsule, with the malignant osteoblasts having round to oval moderately pleomorphic vascular nuclei and moderate cytoplasm. Widespread trabecular osteoid deposition by osteoblast was seen. There were areas of loose fibrous tissue, zones of fibroblastic growth and ectatic vascular channels. Resection margins were free. The histological diagnosis was that of extraskeletal osteosarcoma. See figures VII – IX.

He was discharged home on the 7th day postoperatively. Adjuvant chemotherapy was given with cisplatin and doxorubicin. He had a total of 4 courses.

Patient is currently 14 months' post-surgery and no evidence of local recurrence or distant metastasis.

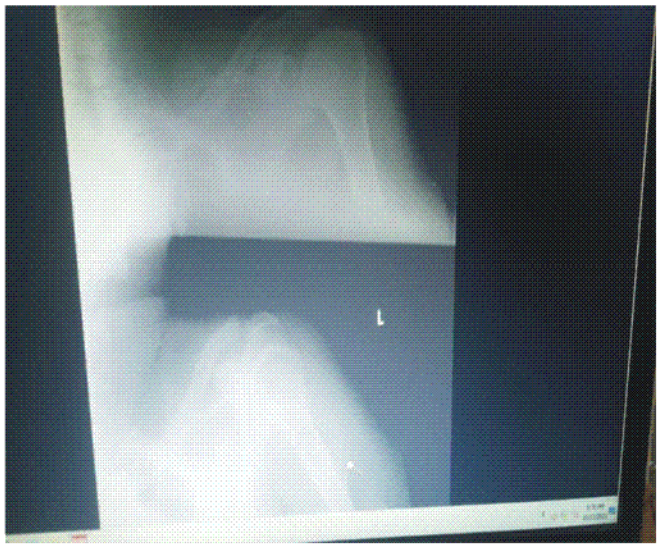

Figure 1: Plain radiograph showing the proximal left humerus and a soft tissue shadow around the deltoid muscle insertion

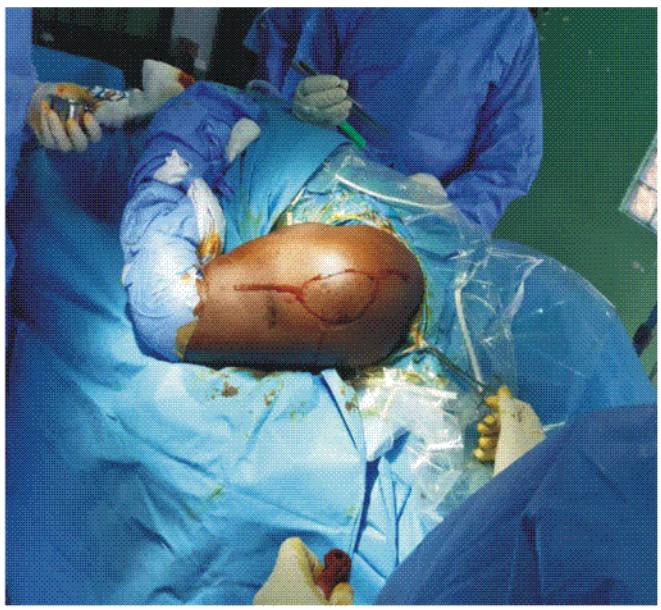

Figure 2: Intraoperative marking for incision

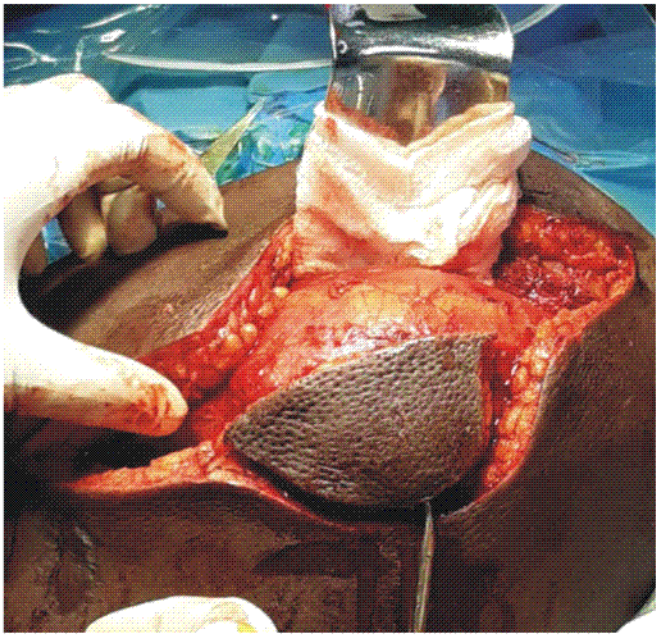

Figure 3: Surgical exposure with the biopsy scar and tumour being resected enbloc

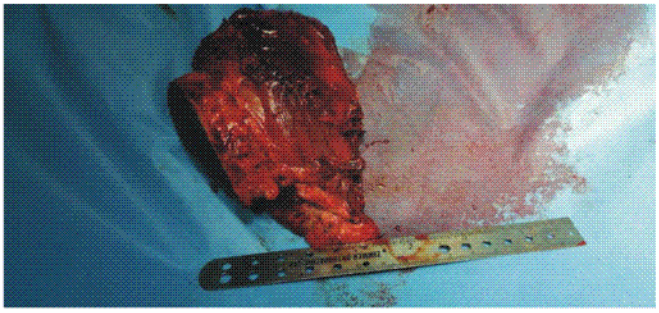

Figure 4: Resected specimen

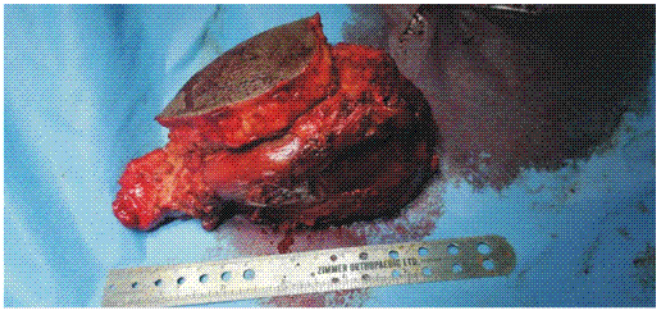

Figure 5: Resected specimen

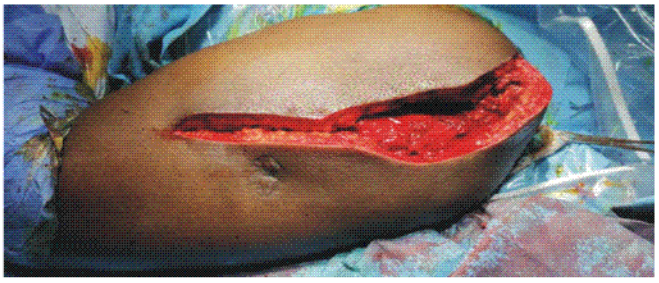

Figure 6: Post tumour resection

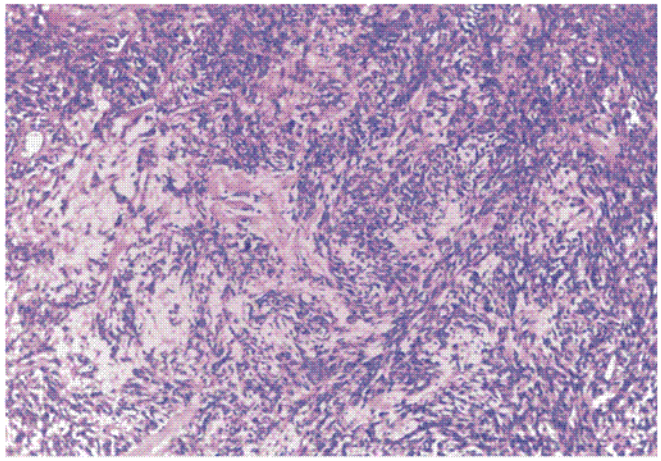

Figure 7: Histology shows sheets of malignant cells elaborating osteoid(H/E x 40)

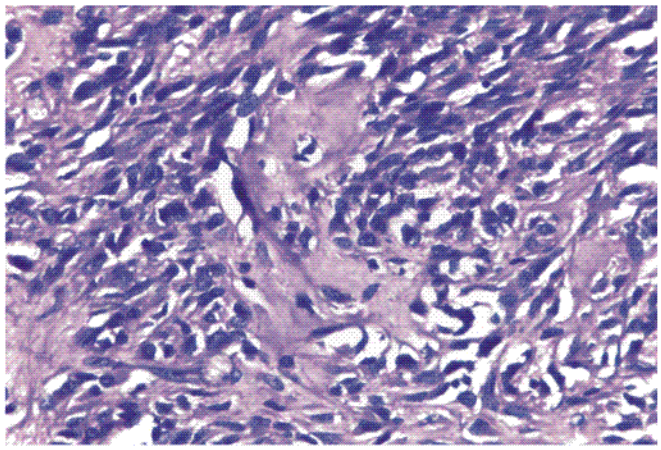

Figure 8: Histology shows malignant osteoblasts with pleomorphic hyperchromatic nuclei elaborating osteoid material (H/E X 100)

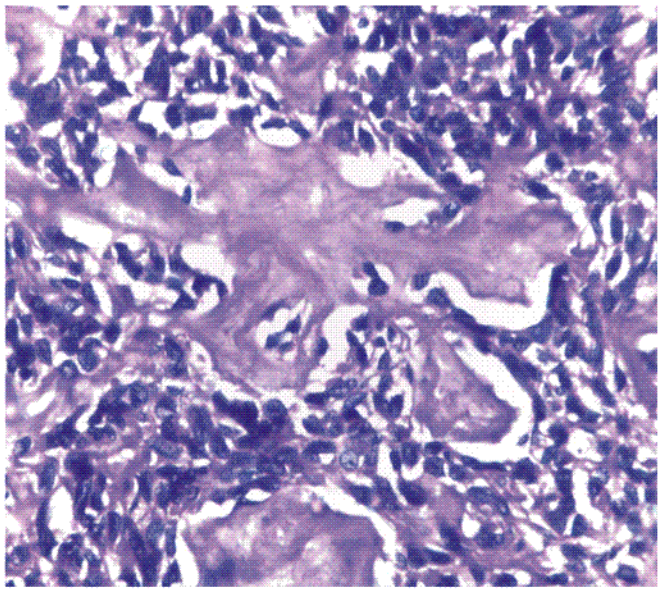

Figure 9: Osteoid material deposited by malignant osteoblast (H/E x 100)

Discussion

ESO was first reported in 1941 by Wilson et al.[] Since then, many series have been reported worldwide. The aetiology is still unknown like in most tumours; however, there are many associated factors that have been found like age, exposure to irradiation, intra-muscular injections and myositis ossificans.[] No clear genetic abnormalities have been associated unlike with osseous osteosarcomas.[]

Presentation of ESO is usually by a mass that grows insidiously to a very large mass of up to 60cm and may be associated with pain and ulceration.[] It commonly affects the thigh, upper limb, mediastinum, breast, orbit and the retropharyngeal space etc.[]Diagnosis is generally based on the absence of attachment to bone and the presence of malignant mesenchymal cells capable of producing osteoid or chondroid matrix.[–]

Plain radiographs will usually help delineate soft tissue mass and there may be some calcifications.[] Computerized tomographic scan and magnetic resonance imaging canboth help to further characterize the tumour.[] Definitive diagnosis is usually made after histology of part or whole tumour.[]

The mainstay of treatment is generally by surgical excision with or without radiotherapy and/or chemotherapy by neo-adjuvant or adjuvant means.[]

Our patient is 52year old man with an upper limb mass that was successfully treated as ESO. Chung EB et al in the US reported a median age of 59years and an average of 54years in their series.[]Longhi A et al reported 57years as the median age in their multi-center European report.[]

In this report, the patient presented with a shoulder mass that grew insidiously over 3years. The shoulder region is the second most commonly affected anatomic location by ESO as has been reported by many researchers.[]

Our patient had plain x-ray, MRI and Tru-cut biopsy that helped to establish the diagnosis before the definitive surgery was done. Diagnosis before definitive surgery is not done in some cases especially in developing countries as the surgeon may perform excision before definitive diagnosis is made. Some do not even subject the specimen to histology at all.[] This is because they usually think it is a benign soft tissue tumour. Alonge et al in his case series found 3 of the five cases had initial excision in a peripheral hospital without subjecting the specimen to histological evaluation.[]Most researchers do a histological evaluation of all patient pre or post definitive surgery.[]

In this report, our patient had wide local excision of the tumour. Wide local excisions with tumour free margin have been found to improve survival and decrease recurrence rate. Generally for soft tissue sarcomas, a resection margin of 5-10mm is adequate if frozen section was employed during surgery.[] However, if frozen section is not available during surgical resection a postoperative adjuvant radiotherapy is required.[] Without adjuvant radiotherapy, a minimum margin of 0f 20mm is required for adequate excision[]Torigoe T et al in a multicenter study in Japan found local recurrence to be absent when margin of resection is >5cm.[] Similarly the European musculoskeletal oncology group also found out that wide local excision gives better outcome and better long term survival.[]Our patient had a 20mm resection margin which was histologically free of tumour. This is in agreement with the general consensus of having tumour free resection margin of >5-10mm.[]

Multimodal treatment has been found to give better outcome of treatment in ESO.[–] Some surgeons treat the disease just like the osseous variety by given adjuvant chemotherapy with cisplatin, doxorubicin or ifosfamide which was found to increase the overall survival rate than when surgery alone was employed.[–] Our patient had four courses of adjuvant chemotherapy using intravenous Cisplatin and Doxorubicin

Conclusion

Extraskeletal osteosarcoma is still a rare tumor that is uncommonly encountered in our practice. Diagnosis and treatment is still a challenge in developing countries like Nigeria for varied reasons. We hope to report a large series of prospectively managed cases as they evolve.

References

1. Chung EB EF. Extraskeletal Osteosarcoma. *J cancer*. 1987; **60**:1132-42.
2. Hoch M, Ali S, Agrawal S, Wang C, Khurana JS. Extraskeletal Osteosarcoma?: A case report and review of the literature. *J Radiol case Rep*. 2013;**7**(Figure 5):15-23.
3. Longhi A, Bielack SS, Grimer R, Whelan J, Windhager R. ScienceDirect Extraskeletal osteosarcoma?: A European Musculoskeletal Oncology Society study on 266 patients *. *Eur J Cancer [Internet]*. 2017; **74**:9-16. Available from: http://dx.doi.org/10.1016/j.ejca.2016.12.016
4. Ogundiran TO, Ademola SA, Oluwatosin OM, Akang EE, Adebamowo CA. Primary osteogenic sarcoma of the breast. *World J Surg Oncol*. 2006; **4**:1-4.
5. Alonge TO, Obamuyide A, Ogun GO. Extraosseous osteosarcoma in Ibadan?: case series over a 20-year period Case Series. *rare tumours*. 2009;**1**:2-4.
6. Regan KO, Krajewski KM, Hornick JL, Butrynski J. Extraskeletal Osteosarcoma: Spectrum of Imaging Findings. *Am J Roentgenol*. 2012; (January):31-9.
7. Wang H, Miao R, Jacobson A, Harmon D, Choy E, Hornicek F, et al. rare tumors Extraskeletal osteosarcoma?: A large series treated at a single institution. *rare tumours*. 2018;**10**:1-7.
8. Lee BP, Prifchard SDJ, Nascimenfo AG. A Review of 40 Patients with Extraskeletal Osteosarcoma. *J cancer*. 1995;**76**:2253-9.
9. Ahmad BSA, Patel SR, Ballo MT, Baker TP, Yasko AW, Wang X, et al. Extraosseous Osteosarcoma?: Response to Treatment and Long-Term Outcome. *J Clin Oncol*. 2002; **20**:521-7.
10. Kasraeian S, Allison DC, Ahlmann ER, Fedenko AN, Menendez LR. A comparison of fine-needle aspiration, core biopsy, and surgical biopsy in the diagnosis of extremity soft tissue masses. *Clin Orthop Relat Res*. 2010; **468**:2992-3002.