Case Report

Recurrent Giant Cell-Rich Osteosarcoma of the Jaw- a Case Report and Review of the Literature

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ABSTRACT

Giant cell-rich osteosarcoma of the jaw (JGCRO) is very rare but it is diagnostically important because of its poor prognosis compared to other variants of osteosarcoma. We report a rare case of recurrent JGCRO initially diagnosed as an ossifying fibroma. The clinical presentation, radiologic and histologic features and the intermediate outcome of treatment of the case are highlighted.

INTRODUCTION

Osteosarcoma (osteogenic sarcoma) is a neoplasm in which the neoplastic cells directly produce immature bone or osteoid tissue[1,2]. It is the most common primary bone tumour. Most cases of osteosarcoma are high-grade tumours that have a bimodal age distribution, affecting young people 10-14 years old, and older persons over 40 years old. They affect the long bones of the extremities (distal femur, proximal tibia and proximal humerus)[2,3]. Six per cent of cases arise from the jaw. At this site, osteosarcomas rarely metastasize and occur 10-20 years later than do osteosarcomas at peripheral sites[3-5].

Giant cell-rich osteosarcoma (GCRO), first described by Bathurst et al, is a rare variant of conventional osteosarcoma. It comprises 3% of all primary osteosarcomas[6]. It is very uncommon in the jaw. To date, very few cases of JGCRO have been reported[7]. However, JGCRO is very important to recognize because it is locally aggressive and has a worse prognosis than other variants of osteosarcoma[8]. We present a rare case of recurrent JGCRO of the jaw in an African female which was initially diagnosed as an ossifying fibroma.

CASE HISTORY

A 37-year-old woman presented to the maxillofacial clinic at the University Teaching Hospitals Adult Hospital, in Lusaka, Zambia, after being referred from her local hospital for the management of a jaw tumour. She complained of a painful, progressively enlarging swelling on the left upper jaw for seven months. A toothache on the left upper jaw was the patient's first symptom. This was followed by progressive swelling which resulted in a blocked left nostril. She had no prior history of trauma or surgery. Her medical history was unremarkable. She had a history of taking alcohol.

The patient was fully conscious and oriented. She had no pallor or jaundice. She had obvious facial asymmetry. Both eyes were moving normally and there was no defect in hearing. Examination of the face revealed a non-tender tumour on the left

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maxillary area extending from the angle of the mouth to the lower border of the left eyelid and measuring approximately 5cm in widest diameter. There was no difference in the colour or temperature of the overlying skin to that surrounding it. Intraoral examination showed gingival swelling with preserved teeth and unaltered palate. There was no bleeding from the lesion both intraorally and extra-orally. The patient was admitted to the hospital. After the patient had a radiologic evaluation of the skull using X-rays (not availed to the authors), the attending doctor conducted an incision biopsy of the lesion and discharged the patient. The patient returned 23 days later for review with her incisional biopsy histology report. The tumour was diagnosed as an ossifying fibroma with a comment that malignancy could not be ruled out because of the small size of the tissue provided. The patient was discharged and asked to return a month later for excision of the tumour.

Twenty-nine days after her last review, the patient underwent an excisional biopsy of the mass. Intraoperatively, a maxillary tumour was noted involving the maxillary sinus and the inferior orbital floor. The tumour was excised piecemeal. On histologic evaluation (Figures 1 and 2), the sections showed a heterogenous spindle cell neoplasm characterized by areas with benign features and other areas with overt malignant features. The benign-appearing areas showed bland spindle cells with associated fibrosis. The malignant areas had sparsely distributed osteoclast-type giant cells and more than an occasional lacelike osteoid deposition. The spindle cells displayed fasciculate and vague storiform patterns. There were spindle cells characterized by moderate eosinophilic cytoplasm, hyperchromatic nuclei and atypical mitoses. The mitotic count was 17 per 10 high-power fields. There was also coagulative necrosis. We diagnosed the tumour as stage two (pT2)[9] GCRO with positive tumour margins.

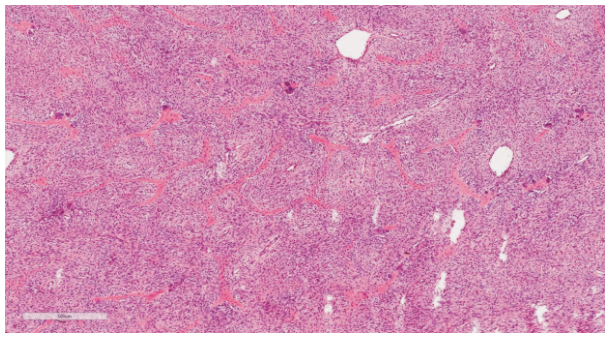


Figure 1

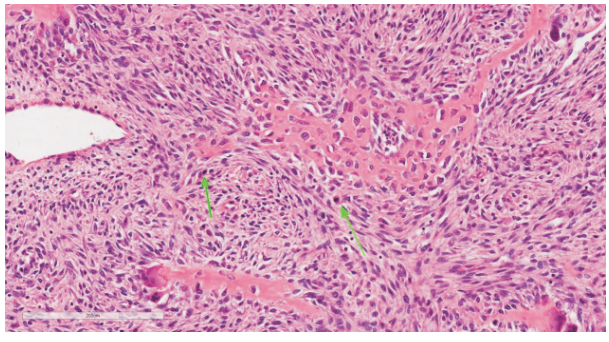


Figure 2

The patient returned for review to the maxillofacial clinic two months after the excision of her tumour. She reported that the tumour had grown back and had extended to the left eye, affecting her vision in the same eye (Figure 3). A CT scan was conducted (Figure 4). The CT-scan report revealed a large, enhancing and expansile left maxillary lesion (approximately 8.4 x 7.2 x 6.5cm) with intralesional dystrophic calcifications and hypodense areas. Bone windows showed erosion of the alveolar ridge, zygomatic arch, inferior and lateral orbital, and maxillary sinus walls.

The patient underwent a second excision biopsy. The excised tissue was evaluated histologically (Figures 5 and 6) and found to be consistent with the diagnosis of an incompletely excised pT2 GCRO.

We reviewed the tissue block upon which the earlier diagnosis had been made (Figures 7 and 8). We noted that there was a focus of an osteoclast-like giant cell, atypical bone and vaguely atypical

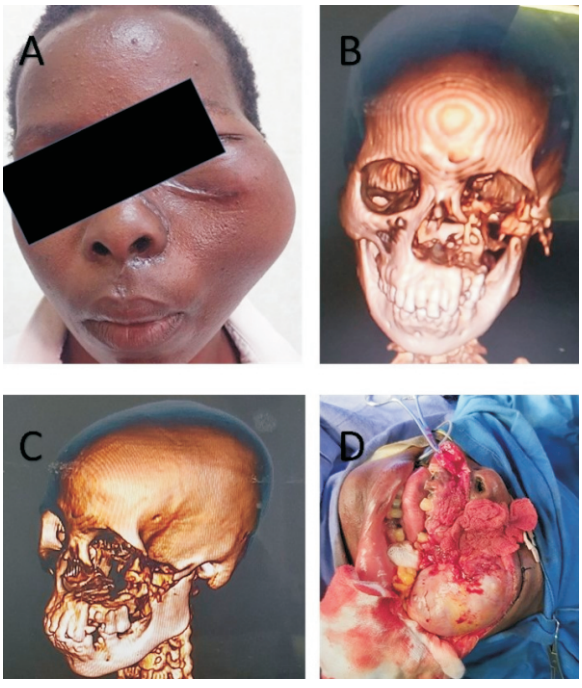


Figure 3

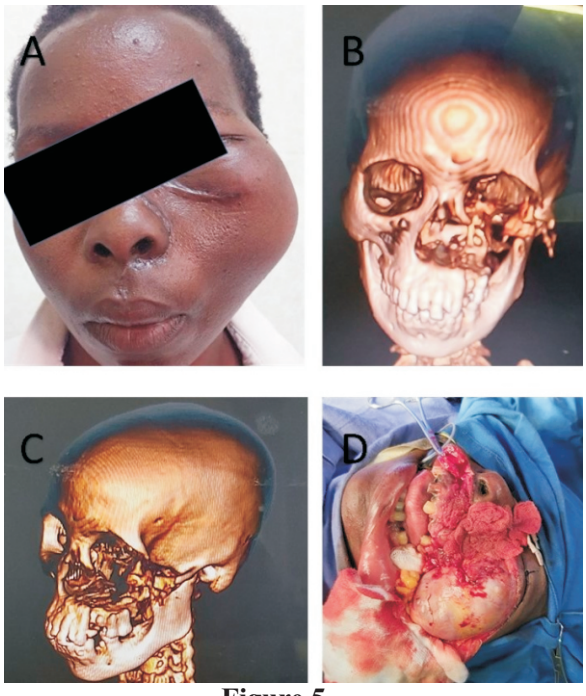


Figure 5

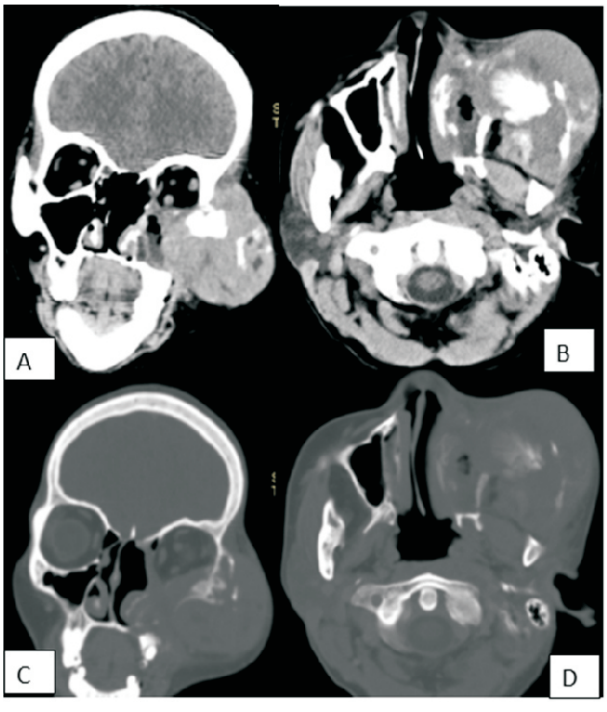


Figure 4

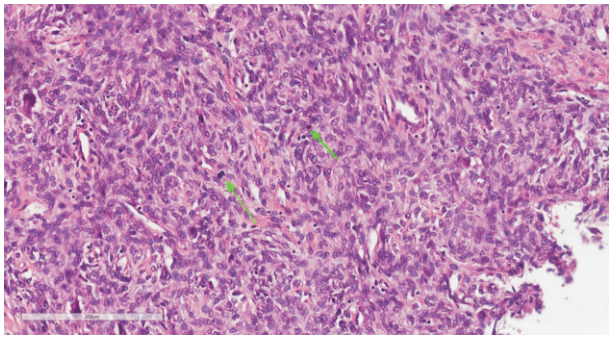


Figure 6

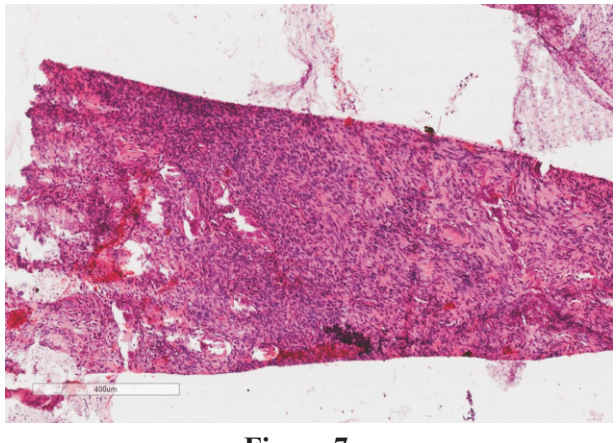
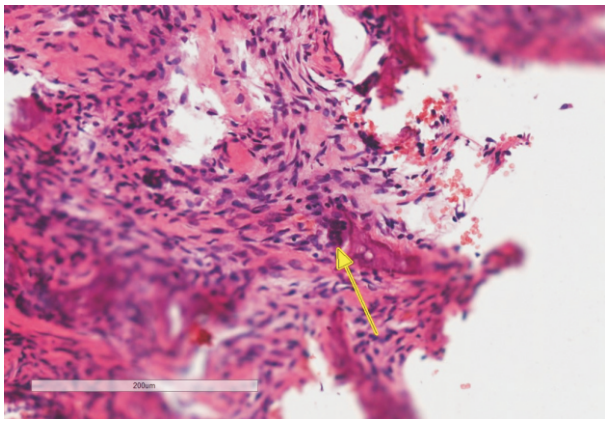


Figure 7



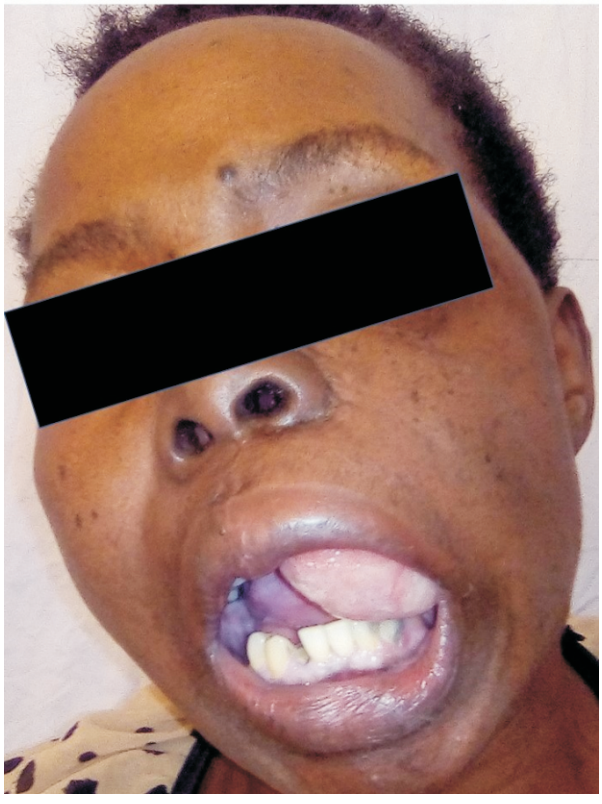


Figure 9

spindle cells. We also noted a comment in the previous report that malignancy could not be entirely ruled out, despite concluding the case as being benign. Therefore, the diagnosis of GCRO was upheld.

The patient was referred for further management to the University Teaching Hospitals' Cancer Diseases Hospital based on the histologic diagnosis of GCRO. The patient was treated with eleven fractions of radiotherapy up to the time of writing this article. She was scheduled to undergo a total of twenty-five fractions of radiotherapy. The patient reported that throughout her treatment with radiotherapy, she had a persistent headache and progressive swelling of the left side of her face (Figure 9).

DISCUSSION

This is the first case of JGCRO reported of an African patient. As of early 2020, there were only six cases of JGCRO which had been reported in the world literature[7]. None of these cases was from Africa. The rarity of this entity in the jaw can therefore pose a diagnostic challenge.

Among the cases that were previously reported, all of them affected adults, there was a 1:1 ratio of occurrence among males compared to women and a 2.5:1 ratio of occurrence in the mandible compared to the maxilla[7]. Our case is an addition to the numbers among females and those affecting the maxilla.

JGCRO in our patient was locally aggressive and showed progressive growth after excision biopsies and after radiotherapy. This supports previous reports that JGCRO is more aggressive than other variants of osteosarcoma of the jaw[8].

In our patient, the diagnosis of GCRO on the incision biopsy had proved to be challenging, because of the scant tissue available for histologic evaluation. The atypia of the mononuclear cells in GCRO can be obvious or suble[10], as observed in histologic sections of the excision biopsies of our patient. It is therefore imperative that pathologists should conduct a careful evaluation of the histologic slides and correlate their findings with radiologic findings and the clinical behaviour of a tumour. GCRO should be distinguished from other giant cell-rich tumours and other variants of osteosarcoma[1,11,12].

Immunohistochemistry for S100 can be utilised to highlight malignant cells in GCRO in obscure cases[12]. Furthermore, clinicians should collect adequately representative samples of a lesion.

Multimodality treatment is recommended for the management of osteosarcomas, including surgery and adjuvant or neoadjuvant chemotherapy with or without radiotherapy[13]. JGCRO, just like GCRO of the temporal bone were treated in the same manner as other osteosarcomas[7,12]. Until our last contact with our patient (Figure 9), she had demonstrated that her major symptoms (pain and growth of the lesion) had persisted despite undergoing thirteen cycles of post-surgical radiotherapy.

CONCLUSION/ RECOMMENDATIONS

GCRO is rare in the jaw. It is challenging to diagnose on small biopsies and must be distinguished from benign and malignant jaw tumours. It is locally aggressive and shows progressive growth after incomplete excision and radiotherapy. An adequately representative sampling of tumour by a clinician and careful histologic evaluation by pathologists (with clinical and radiologic correlation) is key to making an appropriate diagnosis.

CONSENT FOR PUBLICATION

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

CONFLICTS OF INTEREST

The authors have no conflicts of interest.

FUNDING STATEMENT

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REFERENCES

1. Picci P. Osteosarcoma (Osteogenic sarcoma). *Orphanet J Rare Dis Rev.* 2007;4:4-7. doi:10.1186/1750-1172-2-6
2. Fletcher CD., Bridge J, Hogendoorn PC., Mertens F. Osteogenic tumours-Osteosarcoma. In: *WHO Classification of Tumours of Soft Tissue and Bone*. Lyon: IARC Press; 2013:290-296.
3. Baumhoer D, Lopes M, Raubenheimer E. Osteosarcoma. In: El-Naggar AK, Chan JKC, Grandis JR, Takata T, Slootweg PJ, eds. *WHO Classification of Head and Neck Tumours*. 4th ed. Lyon: International Agency for Research on Cancer (IARC) 69372 Lyon Cedex 08, France; 2017:244-246.
4. Kassir RR, Rassekh CH, Kinsella JB, Segas J, Carrau RL, Hokanson JA. Osteosarcoma of the head and neck: meta-analysis of nonrandomized studies. *Laryngoscope*. 1997;107(1):56-61. doi:https://doi.org/10.1097/00005537-199701000-00013
5. Elkordy MA, Elbaradie TS, Elsebai HI, Abd A, Amin E, Khairalla SM. Osteosarcoma of the jaw : Challenges in the diagnosis and treatment. *J Egypt Natl Canc Inst.* 2018;30(1):7-11. doi:10.1016/j.jnci.2018.02.001
6. Bathurst N, Sanerkin N, Watt I. Osteoclast-rich osteosarcoma. *Br J Radiol.* 1986;59(703):667-673. doi:10.1259/0007-1285-59-703-667
7. Malick A, Shah N, Mahmud AS, Das SK. Giant cell-rich osteosarcoma – A rare case. *J Oral Maxillofac Pathol.* 2020;24(Suppl 1):S67-S72. doi:10.4103/jomfp.JOMFP_251_19
8. Kumar R, Gaurav V. Primary Giant Cell Rich Osteosarcoma of Maxilla : An Unusual Case Report.*J Maxillofac Oral Surg*. 2011;10(June):159-162. doi:10.1007/s12663-010-0066-z

9. AJCC. Bone. In: Amin MB, ed. *AJCC Cancer Staging Manual*. Eighth. Chicago: American College of Surgeons; 2018:471-486. doi:DOI 10.1007/978-3-319-40618-3
10. Fletcher CDM. Conventional osteosarcoma. In: *Diagnostic Histopathology of Tumours*. Fourth. Philadelphia, PA 19103- 2899: Saunders- Elsevier; 2013:1894-1896.
11. Mullapudi SV, Putcha UK, Boindala S. Odontogenic tumors and giant cell lesions of jaws - a nine year study. *World J Surg Oncol.* 2011;9(68):1-8.
12. Hong G-B, Wu Q-L, Gou X-M, Gu L-J. Annals of Clinical Case Reports Giant Cell-rich Osteosarcoma in Temporal Bone : Case Report and Literature Review. *Ann Clin Case Reports.* 2016;1:1-4.
13. Ferrari D, Moneghini L, Allevi F, Bulfamante G, Biglioli F. Osteosarcoma of the Jaw: Classification, Diagnosis and Treatment. In: Honoki K, Weiss K, eds. *Osteosarcoma- Biology, Behavior and Mechanisms*. IntechOpen; 2017:197-220. doi:http://dx.doi.org/10.5772/67564