International Journal of Otorhinolaryngology and Head and Neck Surgery
Satpute SS et al. Int J Otorhinolaryngol Head Neck Surg. 2024 Apr;10(2):232-235
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20240705

Case Report

Giant cell rich osteosarcoma of maxilla vs. giant cell granuloma: a diagnostic dilemma

Satish S. Satpute[1], Nidhin S. B.[1]*, Nitin M. Nagarkar[1], Ashish Gupta[2], Ripudaman Arora[1]

[1]Department of ENT, AIIMS Raipur, Raipur, Chhattisgarh, India
[2]Department of Pathology, AIIMS Raipur, Raipur, Chhattisgarh, India

Received: 20 January 2024
Accepted: 08 March 2024

***Correspondence:**
Dr. Nidhin S. B,
E-mail: sbnidhin@gmail.com

ABSTRACT

Giant cell-rich osteosarcoma is a rare histological variant of osteosarcoma, accounting for 3% of all primary osteosarcomas and rarely occurring in the head and neck region. GCRO poses a significant diagnostic challenge radiologically and histopathologically, as it often presents with a benign appearance despite being a clinically aggressive osteosarcoma with a high tendency for recurrence. Meticulous surgical planning, a radical surgical approach, and postoperative radiotherapy are essential to prevent recurrence. We are discussing the case of a 21-year-old male patient diagnosed with Giant Cell-Rich Osteosarcoma of the maxilla. This case highlights the diagnostic dilemma associated with GCRO, the surgical approach employed, and the subsequent reconstruction process.

Keywords: Giant cell rich osteosarcoma of maxilla, Central giant cell granuloma, Temporalis muscle flap

INTRODUCTION

Primary osteosarcoma of the head and neck region is comparatively rare and aggressive malignancy, representing 6 to 13% of osteosarcoma cases, 1 to 5% of all head and neck cancers. Giant cell rich osteosarcoma (GCRO) is an extremely rare histologic variant, accounting for only 1%-3% of conventional osteosarcoma.[1,2] GCRO is an undifferentiated high-grade sarcoma with numerous osteoclast-like giant cells and variable amount of tumor osteoid.[3] The radiological and histopathological differentiation of GCRO from other benign and malignant giant cell tumors (GCTs) is very difficult. It is important to differentiate them from other aggressive GCTs as the prognosis and treatment is different for both. In literature only few cases of GCO in head and neck been reported, GCRO in maxilla prior to this only two case is been published prior, here we are presenting a rare case report of 21 year old male patient diagnosed with GCRO of maxilla its diagnostic challenge, surgical management and reconstruction.

CASE REPORT

21-year-old male patient presented to OPD with complaints of right side cheek swelling with right sided nasal obstruction since 2 years, after thorough clinical examination and detailed radiological examination patient was diagnosed to have sinonasal mass underwent right side medial maxillectomy post-op histopathology came out to be giant cell granuloma, patient had disease-free period of 2 months then slowly he started noticing recurrence of swelling over the right side upper alveolus which later spread to right side palate then progressed to the right cheek with associated complaints of pain over the swelling and restricted mouth opening, on examination asymmetry of face noted single oval-shaped diffuse cheek swelling of size approx 4*3cm noted with surface appears to be smooth with old previous right lateral rhinotomy scar seen, (Figure 2) mass was superiorly extending from the lower eyelid to inferiorly 2 cm from the lower margin of mandible, medially up to median nasal bridge and laterally up to zygoma, skin over

the swelling appears to be smooth pinchable firm in consistency.

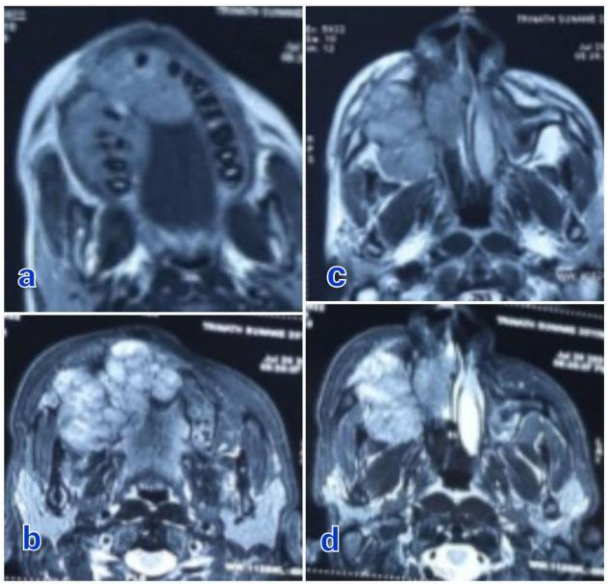

Figure 1 (a-d): CE MRI NOSE PNS showing Ill-defined heterogeneously enhancing lobulated lesion with multiple interspersed foci in right infratemporal fossa, superior margin from inferior orbital margin, inferiorly along nasolabial fold crossing midline to involve philtrum, the anteriorly upto right premaxillary region with infiltration of levator superioris muscle posteriorly destruction of the posterior wall of maxillary sinus extending into masticator space, abutting lateral pterygoid muscle laterally erosion of inner and outer cortex of right zygomatic arch, involving zygomaticus major vessels seen.

Oral cavity examination mouth opening less than two finger, single palatal growth seen arising from right gingival buccal sulcus region extending up to the midline.no significant neck nodes palpable. Right side nasal cavity mass was seen completely obliterating and pushing the septum to the left side. CECT nose PNS orbit done ill-defined minimally enhancing soft tissue lesion with multiple interspersed coarse calcific foci in right maxillary sinus noted, superiorly erosion of floor of the orbit, inferiorly erosion of right alveolus and hard palate, anteriorly it was seen infiltrating levator palpebrae superioris muscle and soft tissue, posteriorly up to the posterior wall of maxilla medially up to nasal septum and laterally up to the zygomatic process of the right maxillary brim (Figure 1). Multiple sub centimetric lymph nodes bilateral cervical region, 9 mm right level Ib. CEMRI Nose PNS Orbit was also done Ill-defined heterogeneously enhancing lobulated lesion with multiple interspersed foci in right infratemporal fossa was seen superiorly inferior orbital margin, inferiorly along nasolabial fold crossing midline to involve philtrum, the anteriorly right premaxillary region with infiltration of levator superioris muscle posteriorly destruction of the posterior wall of maxillary sinus extending into masticator space, abutting lateral pterygoid muscle laterally erosion of inner and outer cortex of right zygomatic arch, involving zygomaticus major vessels seen. Combined approach excision of tumour by enblock dissection i.e.; Right side total maxillectomy (Figure 2) with temporalis muscle flap reconstruction was done (Figure 3).

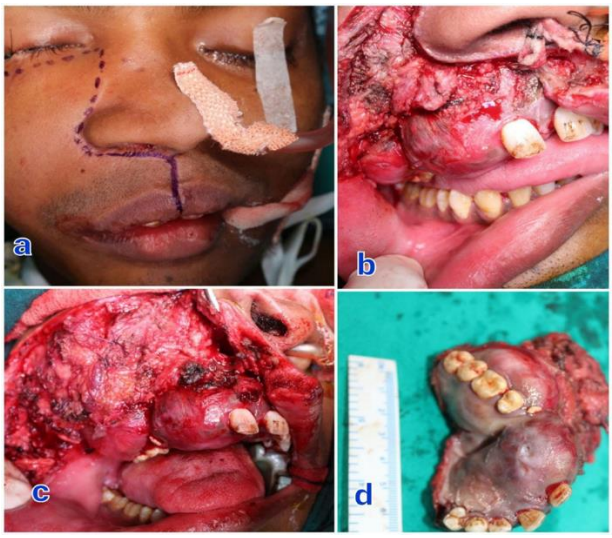

Figure 2: (a) right side weber fergusen incision markings over previous surgical scar site, (b) cheek flap elevated and tumour exposed, (c) tumour eroding posterior wall of maxillary sinus involving ITF, (d) tumour removed in-total.

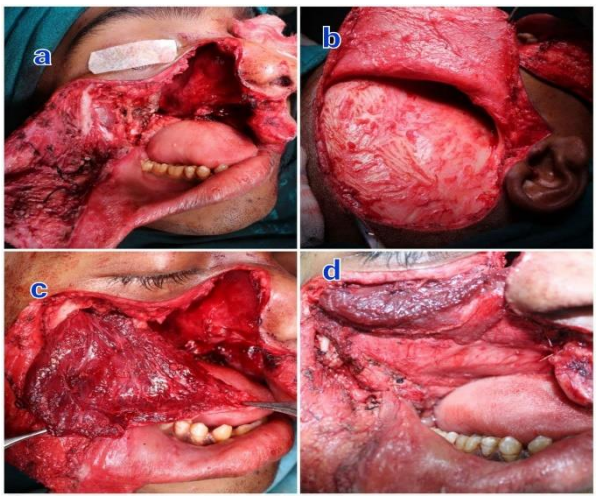

Figure 3: (a) post total maxillectomy tumour excision defect seen, (b) temporalis muscle flap harvested, (c) temporalis muscle flap tunneled below the zygomatic arch, (d) muscle layer approximated and sutured to opposite side thus reconstructing floor of orbit and nasal cavity.

Since patient had limited mouth opening, we did tracheostomy under local anaesthesia then general anaesthesia was induced through tracheostomy tube.

weber Fergusen incision given, cheek flap elevated tumour excised in total complete right maxilla removed along with left palate (Figure 2) also post excision total maxillary reconstruction done with temporalis muscle flap post-op histo-pathology came as microscopically lined squamous epithelium with multiple fragments of fibrocollagenous tissue with occasional giant cell seen more towards giant cell granuloma. Case was discussed in inter departmental meet clinical histopathology diagnosis was revised to giant cell rich osteosarcoma (Figure 4), postoperatively patient was started on radiotherapy 23 cycles of radiation given following patient was decannulated, patient comes for regular follow up till date no recurrence noted (Figure 5).

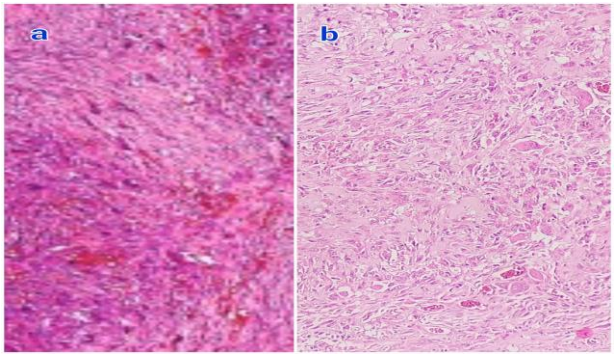

Figure 4: Histopathology image showing (a) giant cell granuloma, (b) giant cell rich osteosarcoma tumor cells were highly pleomorphic with some of them ovoid to round and some spindle shaped, with numerous osteoclast-like giant cells. Areas of lace like osteoid formation by the tumor cells were also seen as eosinophilic staining with glassy appearance and irregular contours.

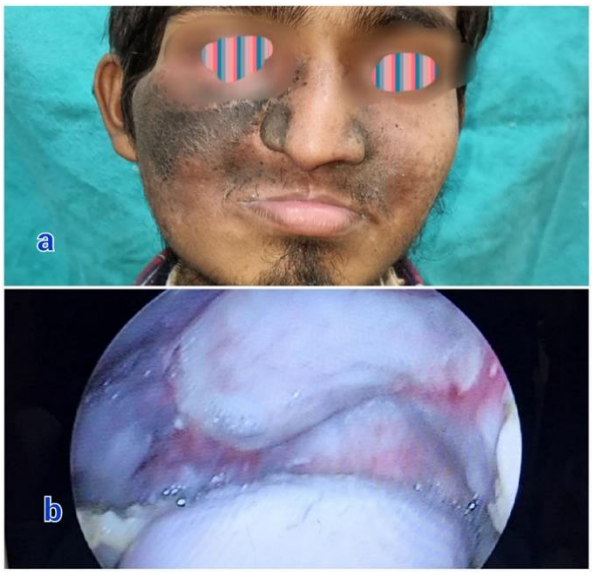

Figure 5: (a) post operative picture of the patient, (b) Intra oral picture showing healed oral cavity roof with adequate mouth opening.

DISCUSSION

Osteosarcomas are malignant bone tumors which most commonly arise from long bones of the extremity. Osteosarcoma of head and neck region accounts for less than 10% of all cases of osteosarcoma.[4] Osteosarcoma have different histological variant of which Giant cell-rich osteosarcoma is a rare histological variant accounts for about 3% of all Primary intraosseous osteosarcomas, first described by Bathurst et al in head and neck region mandible is the common site, giant cell rich osteosarcoma of maxilla very uncommon as only two case being published in the literature regarding GCRO of maxilla till now, GCRO is aggressive, poor prognosis commonly seen in older age group and female predominance but in our case it seen in young age male its seen.[5] The differential diagnosis of giant cell lesions can present a diagnostic challenge, especially if they occur in an unusual location. In this case, it was difficult to differentiate between a giant cell granuloma from giant cell osteosarcoma. The major risk factors for development of head and neck osteosarcoma are radiation exposure and Paget's disease.[6]

Radiologically giant cell rich osteosarcoma are different from the conventional osteosarcomas as they mimic non mineralized benign or malignant bone tumors. In the long bones they present as osteolytic lesion with cortical thinning and ballooning without obvious cortical destruction.[7] While in our case CECT Nose PNS showed expansile heterogeneous mass filling the maxilla with extensive destruction of the all the walls of the maxilla, areas of speckled calcification within the tumor and adjacent sclerosis. The tumor was destroying the palate and extending into oral cavity. Radiologically, the differential diagnosis includes giant cell granuloma, aneurysmal bone cyst, chondrosarcoma, Chondromyxoid fibroma and malignant fibrous histiocytoma.[8] Surgical excision with post operative Radiotherapy is the treatment of choice. In our case combined approach radical tumour excision done ie total maxillectomy and reconstruction.[2] Temporalis muscle flap is used for reconstruction in this case as it has a reliable and hearty blood supply, can be tailored to provide reconstruction of most oropharyngeal defects, and arguably presents minimal donor site morbidity if performed correctly.[9] The flap is accessible through a hemicoronal incision that is hidden in the hair-bearing scalp and preauricular crease. The temporalis muscle flap lies in the same operative field as the defect, potentially reducing operative time and reducing motion-induced viability issues during the recovery period. The deep temporal fascia lines the flap and obviates the need for skin grafting. The temporalis muscle develops a mucosal layer with time.[10]

Giant cell tumor of bone (GCTB) Macroscopically, is usually a red-brown and friable tumor with a well-defined or locally infiltrative border. Histologically, GCTB comprises of stromal cells and numerous osteoclast-like multinucleate giant cells result from fusion of

mononucleate cells. Stroma composed of plump, round and oval cells together with a rich vascular network.[11] Giant cells are larger, more rounded with many nuclei (50-100) and no osteoid or new bone formation is appreciated. Fresh hemorrhage and hemosiderin deposits are not seen to differentiate it from central giant cell granuloma (CGCG).[11,12]

CGCG is a benign but sometimes locally aggressive tumor of the jaws, most commonly affecting the mandible of female pediatric patients, CGCG is unencapsulated and composed of proliferative spindle cells admixed with osteoclastic multinucleate giant cells, in a vascular and hemorrhagic background. Giant cells are grouped around haemorrhagic foci and they are relatively smaller, irregular, with relatively few nuclei in contrast to GCOS.[12,13] Foci of osteoid and new bone formation often seen in the middle of the lesion. Giant cell rich osteosarcoma is an undifferentiated sarcoma with scanty osteoid formation. The microscopic appearance of giant cell rich osteosarcoma is characterised by numerous osteoclast-like giant cells dominating the picture, however the presence of osteoid formation by the tumor cells gives a clue regarding the diagnosis. These giant cells appeared as nodular clusters admixed with fibroblasts or histiocyte-like cells, or alternated with bundles of collagen-rich fibroblastic tissue.[14] In the present case, the histology showed tumor cells arranged in fascicular pattern as well as in sheets. The tumor cells were highly pleomorphic with some of them ovoid to round and some spindle shaped, with numerous osteoclast-like giant cells. Areas of lace like osteoid formation by the tumor cells were also seen as eosinophilic staining with glassy appearance and irregular contours. Based on these histopathological features, a definitive diagnosis of giant cell rich osteosarcoma can be made.

CONCLUSION

Giant cell osteosarcoma is a rare histological variant of osteosarcoma mimicking radiologically and histologically towards benign giant cell granuloma, it's extremely difficult to diagnose GCRO from Giant cell granuloma and as well as from conventional osteosarcoma. We recommend the holistic approach in which we incorporate clinical, radiological and histological findings along with at least H3F3A sequencing or immunohistochemistry for derived mutant proteins for all head and neck giant cell-containing lesions so as to prevent the misdiagnosis. Aggressive surgical resection with post operative radiotherapy provides better 5-year survival.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Verma RK, Gupta G, Bal A, Yadav J. Primary giant cell rich osteosarcoma of maxilla: an unusual case report. J Maxillofac Oral Surg. 2011;10(2):159-62.
2. Mallick A, Shah N, Mahmud SA, Das SK. Giant cell-rich osteosarcoma A rare case. J Oral Maxillofac Pathol. 2020;24(1):S67-72.
3. Misaghi A, Goldin A, Awad M, Kulidjian AA. Osteosarcoma: a comprehensive review. SICOTJ. 4:12.
4. Lim S, Lee S, Rha SY, Rho JK. Cranofacial osteosarcoma: Single institutional experience in Korea. Asia Pac J Clin Oncol. 2016;12(1):e149-153.
5. Bathurst N, Sanerkin N, Watt I. Osteoclast-rich osteosarcoma. Br J Radiol. 1986;59(703):667-73.
6. Krishnamurthy A, Palaniappan R. Osteosarcomas of the Head and Neck Region: A Case Series with a Review of Literature. J Maxillofac Oral Surg. 2018;17(1):38-43.
7. Sato K, Yamamura S, Iwata H, Sugiura H, Nakashima N, Nagasaka T. Giant cell-rich osteosarcoma: a case report. Nagoya J Med Sci. 1996;59(3-4):151-7.
8. Zambo I, Pazourek L. Giant cell-rich lesions of bone and their differential diagnosis. Cesk Patol. 2017;53(2):61-70.
9. Hanasono MM, Utley DS, Goode RL. The temporalis muscle flap for reconstruction after head and neck oncologic surgery. Laryngoscope. 2001; 111(10):1719-25.
10. Nagasaka T. Reconstruction of midface defect with temporalis myofascial pedicle flap: a review and case series. MOJS. 2021;9(1):186-8.
11. Lee J-C, Huang H-Y. Soft Tissue Special Issue: Giant Cell-Rich Lesions of the Head and Neck Region. Head Neck Pathol. 2020;14(1):97-108.
12. Jeyaraj P. Management of Central Giant Cell Granulomas of the Jaws: An Unusual Case Report with Critical Appraisal of Existing Literature. Ann Maxillofac Surg. 2019;9(1):37-47.
13. Aragão M do S, Piva MR, Nonaka CFW, Freitas R de A, de Souza LB, Pinto LP. Central giant cell granuloma of the jaws and giant cell tumor of long bones - an immunohistochemical comparative study. J Appl Oral Sci. 2007;15(4):310-6.
14. Sun L-M, Zhang Q-F, Tang N, Mi X-Y, Qiu X-S. Giant cell rich osteosarcoma of the mandible with abundant spindle cells and osteoclast-like giant cells mimicking malignancy in giant cell tumor. Int J Clin Exp Pathol. 2015;8(8):9718-22.

Cite this article as: Satpute SS, Nidhin SB, Nagarkar NM, Gupta A, Arora R. Giant cell rich osteosarcoma of maxilla vs. giant cell granuloma: a diagnostic dilemma. Int J Otorhinolaryngol Head Neck Surg 2024;10:232-5.