Case Report - Oncology

Chondromyxoid fibroma of the mandible: Case report and review of the literature

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ABSTRACT

Introduction: Chondromyxofibroma (CMF) is exceedingly rare, accounting for 0.5% of the 10,065 bone tumors categorized by Unni and Inwards and 1.6% of their catalog of benign bone tumors. Only 2 of the 50 chondromyxoid fibromas included in their study occurred in the skull. Chondromyxoid fibroma of the maxillofacial region is typically seen in patients in the 2nd-3rd decade of life with slight female predominance. It is more commonly seen in the maxilla and is unusual in the sphenoid and ethmoid sinuses. The tumor is composed of hypocellular chondroid or myxochondroid tissue with multinucleated giant cells. **Case Report:** A 30 year old Nigerian house wife was seen at the Oral and Maxillofacial clinic of the Dental and Maxillofacial Department of the Federal Medical Centre Lokoja with a 4 year history of Rt mandibular swelling which was initially slow growing and painless and difficulty in eating. The whole lesion was removed and result confirmed the previous biopsy of chondromyxofbroma of the jaw. **Conclusion:** Patients with CMF need close monitoring due to high rate of recurrence with cases of malignant transformation at rate of 1-2%, and this seems to have occurred following irradiation.

Keywords: Chondromyxoid fibroma, disarticulation, mandible, resection

INTRODUCTION

Chondromyxoid fibroma (CMF) is a benign cartilaginous neoplasm first distinguished from other cartilaginous tumors by Jaffe and Lichenstein in 1948.[1,2] It is exceedingly rare, accounting for 0.5% of the 10,065 bone tumors categorized by Unni and Inwards and 1.6% of their catalog of benign bone tumors. Only 2 of the 50 CMF included in their study occurred in the skull. In another study of 76 cases of CMF including additional cases from the literature, 1 of 189 were in the skull, and in a review of 278 cases, 15 were in the skull or facial bones.[2] Formerly it was classified as myxoma or a myxomatous variant of giant-cell tumor, or mistaken for a malignant lesion, especially chondrosarcoma, chondromyxosarcoma or myxosarcoma.[1]

It is prevalent in males, and it mostly occurs between the 2nd and 3rd decades.[3] This benign neoplasm occurs most frequently in young adults and it is found in numerous anatomic locations, including long bones, flat bones and craniofacial bones.[2,3] The craniofacial region bones tumors are extremely rare, approximately 2% of the cases,[3,4] nearly 5.4% according to Won *et al.*, 2010[5] and they often involve the jaw and maxilla,[4] with a higher incidence in the mandible (76%) than in the maxilla (24%) and an equal sex distribution.[6]

It generally presents as an active stage lesion, locally destructive with a high recurrence (>25%) and does not undergo malignant transformation.[4] The clinical course ranges according to the area involved and generally associated with a long standing history of non-specific symptoms, mainly pain and edema.[3]

The computed tomography findings of CMF are non-specific and almost always suggest a benign lesion. They typically have a lobulated outline with sharp margins and the majority has a sclerotic rim. The cortex of the bone is usually thinned and expanded. In approximately 50% of cases, a portion of the cortex

may be absent. Up to one-third of cases show radiographic evidence of soft-tissue extension. The majority of tumors have purely lucent matrix. However, approximately 13% of tumors show some discrete areas of calcification.[4] CMF consists of lobulated areas of interspersed myxoid, fibrous and chondroid material,[2-4] multinucleated giant cells.[4] A search in the literature did not reveal any reported case of CMF in our environment.

We present here a 30-year-old Nigerian housewife with a CMF of the mandible and review the necessary literature.

CASE REPORT

This was a case report of a 30-year-old Nigerian seen at the Oral and Maxillofacial Clinic of the Dental and Maxillofacial Department of the Federal Medical Centre Lokoja with a 4 years history of the right mandibular swelling, which was initially slow growing and painless causing difficulty in eating [Figure 1]. There were no history of trauma and no other swelling in any other part of the body. Examination revealed a wasted lady and looking old for her age with a massive mandibular swelling extending from the preauricular region on the right to the body on the left and protruding out of the mouth. It was firm in consistency [Figure 1]. The mucosa over it had patches of ulceration. Intraorally, the lesion extended from retromolar on the right to 36 on the left. There was deranged occlusion with some teeth missing. There were no lymphadenopathies. Incisional biopsy revealed CMF of the mandible. Hematology and other ancillary investigations were done. Polycythemia vera was 26% while other investigations were within normal range. She was prepared up for surgery under general anesthesia. After a difficult intubation (Nasotracheal), she had resection with disarticulation (right temporomandibular joint disarticulation with resection at 36) via an extended Risdon incision and lip split approach [Figure 2]. The whole specimen was sent for histology. She had 4 units of blood (2 units preoperative and 2 postoperative). The histology confirmed CMF [Figure 3]. The patient is being follow-up and is doing well [Figure 4].

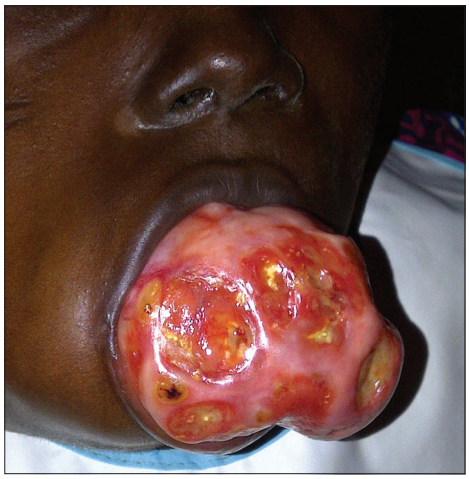

Figure 1: Patient with the lesion protruding out of the mouth

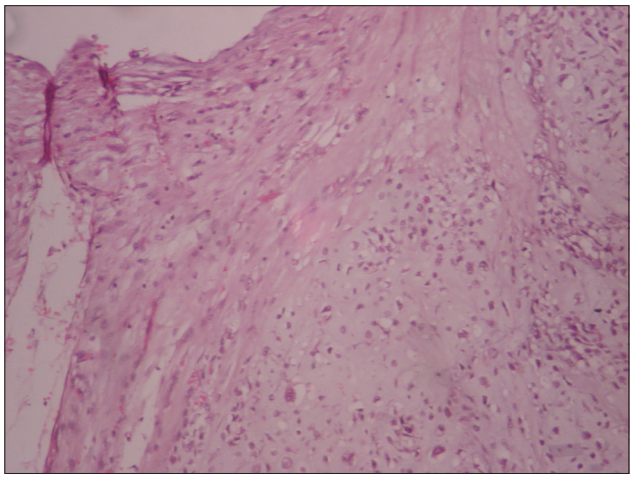

Figure 3: Photomichrograph of chondromyxoid fibroma H and E, ×100. It shows chondroid and fibromyxoid areas

DISCUSSION

CMF of the maxillofacial region is typically seen in patients in the 2nd to 3rd decade of life. The male to female ratio is 2:1[4,7] except when the cranium and facial bones are involved, as women then prevail in a 2:1 ratio.[3,7] In our case, the patient was a female of 30 years and the lower jaws was involved.

According to Semnic *et al*.,[8] the lesion is always greater than 5 cm in diameter at presentation. In our case, the lesion was 9 cm in diameter.

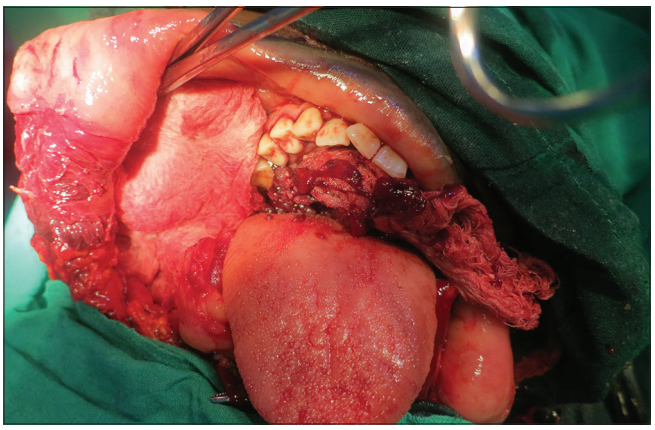

Figure 2: The patient intraoperative after resection and disarticulation

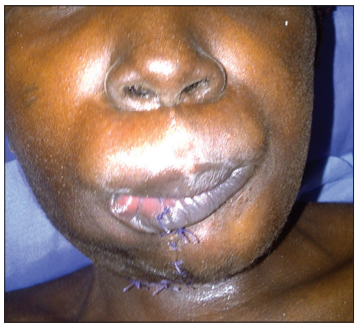

Figure 4: The patient 5 days after surgery and before suture removal

Rare examples of malignant CMF and sarcomatous transformation of CMF have been reported. However, it is unclear whether malignant CMF actually exists or whether it represents another type of sarcoma that has been misdiagnosed.[9]

The clinical course ranges according to the area involved and generally associated with a long standing history of non-specific symptoms, mainly pain and edema.[4] In most cases, CMF is slow growing and incidentally found on routine radiography for reasons not specifically related to symptoms caused by these lesions. When symptomatic, cases are associated with a long history of chronic local pain (85%), swelling (65%), edema (65%) and possibly a palpable soft tissue mass resulting in restriction of movement and more rarely, pathologic fracture.[7] In our case, there was history of pain, swelling, and inability to feed with the tumor protruding out of the mouth.

The radiological appearance is characteristic, but misses specificity. Osseous destruction areas, well-defined lobulated margins, partial cortical erosion and septations are viewed.[4] In our own case, the cortical outline was destroyed and there was soft-tissue infiltration.

The computed tomography findings of CMFs are non-specific and almost always suggest a benign lesion. They generally have a lobulated outline with sharp margins and the majority has a sclerotic rim. The cortex of the bone is usually thinned and expanded. In approximately 50% of cases, a portion of the cortex may be absent. Up to one-third of cases show radiographic evidence of soft-tissue extension. The majority of tumors have purely lucent matrix. However, approximately 13% of tumors show some discrete areas of calcification. It exhibits low signal intensity on T2-weighted images due to chondroid and myxoid tissue with an inhomogeneous pattern of enhancement.[3]

The differential diagnosis is made mainly with chondrosarcoma, which also has a lobular standard and may have myxoid areas. CMF differs for its fibroid elements separating the lobes and also has well-distinguishable margin between the tumor and the normal tissue. The chordoma is another differential diagnosis, and may be distinguished from CMF in immunohistochemical studies, since it expresses antigens like: Epithelial membrane antigen, protein S100 and cytokeratins, whereas this has only protein S100.[4]

Microscopically, it showed encapsulated mass composed of nodules cartilage found in between fibromyxoid areas. The chondrocytes were plump to spindly in shape, with no nuclear atypia, had indistinct cell borders and were disposed in sparsely cellular lobules of myxoid or chondroid matrix. Furthermore seen were cellular zones of the tumor, spindle cell stromal areas and areas of calcification.

In the treatment of craniofacial CMF, most authors now recommend thorough curettage of the tumor followed by careful surveillance.[7] However, when curettage is employed, CMF shows a recurrence rate approaching 25%. In many circumstances, especially cases involving the long bones, the preferable treatment is excision, generally due to the functional and cosmetic difficulties following en bloc resection.[4,7,8] When the tumor is surgically inaccessible, radiation therapy is indicated.[4,7] Local recurrences are more often encountered in younger patients, possibly due to the friable nature of the tumor, but there have been no reports of metastases. There have, however, been reports of the tumor undergoing malignant transformation at a rate of 1-2%, and this seems to have occurred following irradiation.[7] In our case, the patient had resection with disarticulation and is being considered for delayed reconstruction despite our limited resources and skill.

CONCLUSION

CMF patients even though they need close monitoring due to recurrence rate approaching 25%, the problem in our environment is that once the lesion has been removed, the patients disappear. Late presentation in our patient due to financial constraint left her with a huge defect. She also was malnourished as a result of the size of the tumor.

Furthermore we are limited in our environment for any form of reconstruction, therefore restoration of function and esthetics are very difficult.

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