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Case Report

Rare Primary Mandibular Chondrogenic Sarcoma in a Middle Aged Woman – Imaging Perspective

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Abstract: Chondrogenic sarcomas (CSs) are uncommon cartilaginous neoplasms with rare occurrence in the head and neck region where the maxilla is most commonly affected. Histologically, they are hypercellular lesions with abundant amount of hyaline type cartilage and they range from well-differentiated growth resembling benign cartilage tumour to high-grade malignancy with aggressive local behaviour and potential to metastasize. Only few cases of mandibular affectation have been reported with fewer cases involving the nasal cavity, larynx, skull and cervical vertebrae. Here a case of high grade, primary right hemimandibular CS in a 45-year-old woman presenting with right jaw swelling and a discharging sinus following tooth extraction is reported. Initial x-ray examination of the skull demonstrated 'sunray' pattern in the mass which raised high index of suspicion for an aggressive process. Computed tomography (CT) imaging of the jaws showed the lesion to be destructive with adjacent soft tissue involvement. It also aided definition of tumour extent which was necessary for surgical planning. The mass was excised and histologic evaluation of the specimen revealed atypical chondrocytes with mitotic figures on chondromyxoid background consistent with high grade chondrosarcoma. The aim of this case presentation is to provide insight into the role played by three dimensional imaging modalities (CT and MRI) in diagnosis and management of malignant jaw lesions.

Keywords: Chondrosarcoma, Mandible, Imaging

1. Introduction

CSs are a heterogeneous group of malignant bone tumors that share in common the production of chondroid matrix. They are rare tumours with estimated overall incidence of 1 in 200,000 per year [1]. They account for 11-20% of all primary malignant tumors affecting the human body.

Cranio-facial CS occur less frequently, accounting for approximately 1% to 3% of all head and neck tumours [2]. The sites which get affected at relatively high incidences include the long bones, the pelvis and the ribs. In the head and neck region, they affect mainly the maxilla, while relatively few arise from the mandible, nasal cavity, larynx, skull and cervical vertebrae [2].

CS can be divided into primary, when they arise from healthy tissue, or secondary, when they arise from previously existing chondromas or cartilaginous exostosis [3].

We present the case of a 45year old woman with right hemi mandibular swelling and imaging features consistent with high grade mandibular chondrogenic sarcoma.

2. Case Presentation

CE is a 45-year-old woman admitted via the dental clinic of the Nigeria Navy Reference hospital Ojo on account of progressively increasing jaw swelling following tooth extraction 1 year prior to presentation. There was history of associated pain, inability to close the mouth and weight loss.

There was no history of nasal discharge or obstruction. No history of epistaxis, diplopia, diminished vision or headache.

On examination, she was stable and in no respiratory distress. Her vital signs were normal. There was obvious right hemimandibular swelling that extended from the midline to the tragus. It was bony hard and caused bulging of the hard palate. There was associated dental anarchy and the mass was not attached to the overlying skin. A discharging sinus was seen at its infero-lateral border. There was no similar mass in any other part of the body and her vital signs were stable.

Laboratory investigations revealed low normal PCV of 37% (range 36-46%) and WBC of 7.4% (leucopaenia). Electrolytes, urea and craetinine results were normal.

Radiographs of the jaws (Figures 1 and 2) revealed poorly defined, expansile radiodense mass involving the entire right hemimandible and causing distortion of its normal anatomy. The mass demonstrated 'sunray' pattern with surrounding soft tissue extension and ipsilateral maxillary antra opacification. Dental anarchy was evident on the radiograph due to root resorption.

Cranio-facial CT examination (figures. 3 and 4) showed an expansile hyperdense mass arising from the right hemimandible and causing destruction of part of the bone. There was evidence of tumour extension into the surrounding soft tissue. 3D volume reformation of the image (Figure 5) clearly depicted the extent of bone involvement by the mass. There was no evidence of distant metastastasis found on imaging.

Subtotal mandibulectomy was carried out and specimen was sent for histology which reported findings of chondroid tissue populated by highly pleomorphic and atypical chondrocytes consistent with stage III chondrogenic sarcoma (Figure 6). Patient had uneventful post-op recovery and was discharged home for outpatient clinic follow-up.

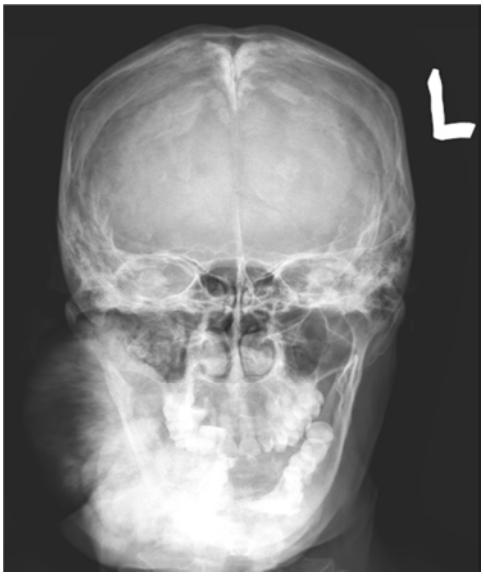

Figure 1. *Frontal skull radiograph showing radiodense right hemimandibular mass with sunburst pattern, surrounding soft tissue involvement and dental anarchy. Note ipsilateral maxillary antrum opacification.*

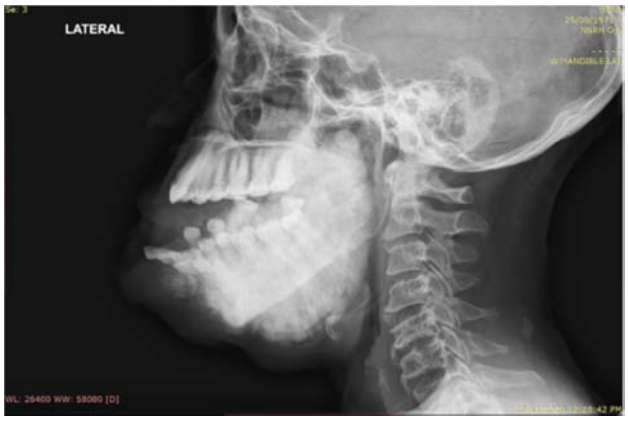

Figure 2. *Lateral skull radiograph showing the right hemimandibular mass with soft tissue infiltration and dental anarchy. Note background cervical spine degenerative changes.*

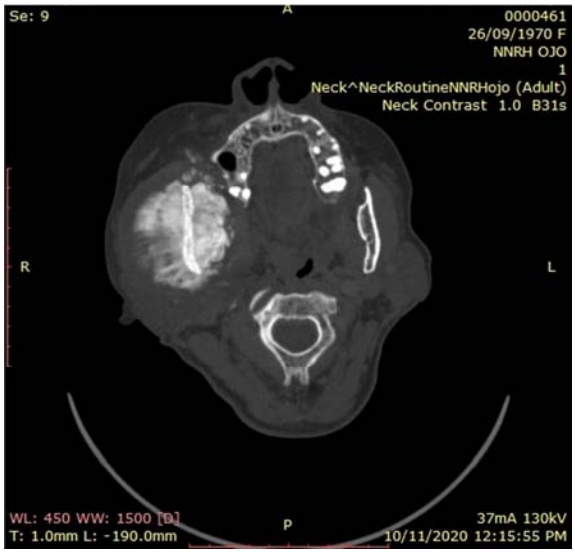

Figure 3. *Axial CT image (bone window) at the level of the body of mandibles, showing juxtacortical, calcific dense, right hemimandibular tumour. Note the surrounding soft tissue extension and calcifications.*

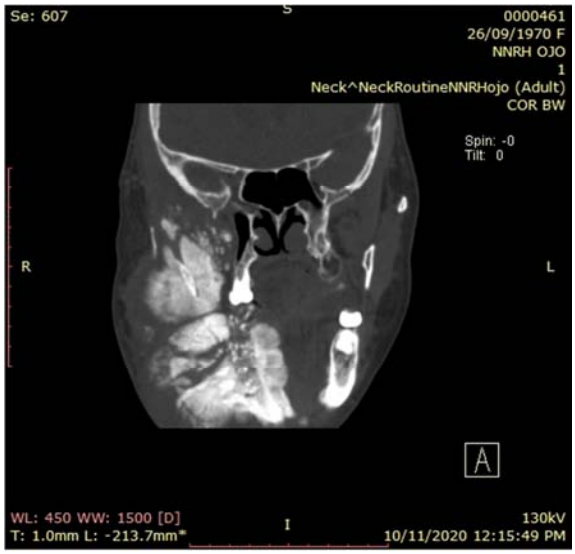

Figure 4. *Coronal cranio-facial CT image (bone window) showing destruction of the right hemimandible by the tumour.*

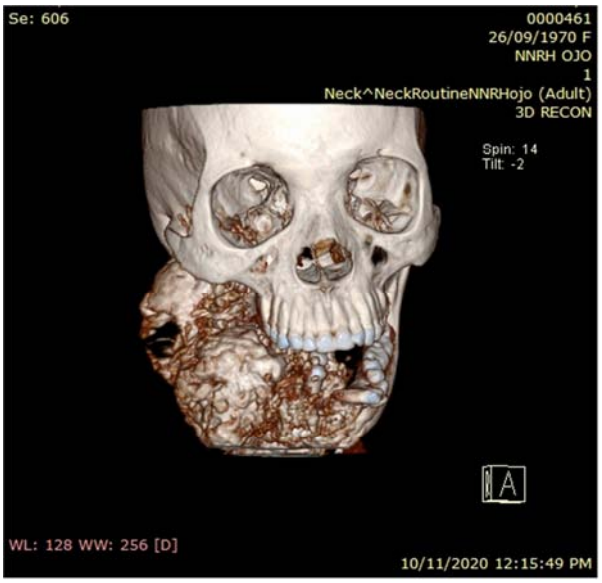

Figure 5. *3D Volume reformatted image of the cranium demonstrating involvement of the entire right hemimandible by the tumour. Note associated dental anarchy.*

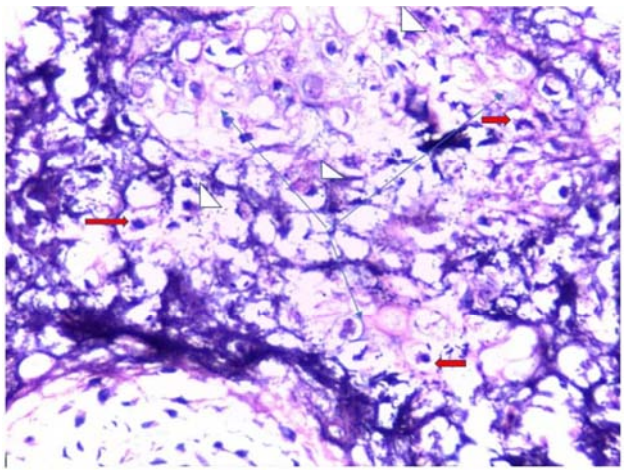

Figure 6. *High power photomicrograph of the specimen showing lobules of tumour cells (thin arrows) on chondromyxoid background. Atypical chondrocytes (white arrowheads) are seen within lacunae and few abnormal mitotic figures (red block arrows) are seen at this magnification (H & E 400x).*

3. Discussion

CS is a collective term for a group of tumours that consist predominantly of cartilage and that range from low grade tumours with low metastatic potential to high grade, aggressive tumours [4]. The usual age of incidence is from fifth to seventh decades of life with slight predominance in males [5]. CS of the jaws do not show sex predilection and the mean patient ages range from 35 to 45 years, although patients younger than 20 years of age have been reported [5]. Our patient was a female in her mid fourties with obvious right jaw mass that distorted her facial appearance.

The signs and symptoms of head and neck chondrosarcomas vary from pain, swelling, headache and hearing loss with neurological problems depending on the tumor location [6]. Jaw lesions may be associated with limited mouth opening, separation or loosening of teeth, expansion of cortical plates, and premature exfoliation of teeth [6]. The index case had jaw swelling with associated pain, inability to close the mouth, loosening of teeth and discharging sinus.

While lesions of the jaws very often have characteristic features that allow the clinician to arrive at differential diagnoses, it cannot be overemphasized that imaging evaluation is key in arriving at accurate diagnosis.

Certain radiographic appearances are typically associated with specific disease processes. It should be remembered, however, that radiographic appearance of a lesion is never pathognomonic but can help narrow the possibilities.

'Sunray' pattern in bony lesion as was seen on the radiograph of our patient narrowed the differentials to few pathologic processes which include osteosarcoma and chondrosarcoma.

Although the radiological pattern of chondrosarcomas is variable, findings often include opacification of air spaces, densely calcified bone mass and root resorption [7]. Also, ground glass appearance or a sunburst pattern may be seen as earlier noted.

Secondary malignant transformation should be suspected when radiographs of benign cartilage tumours show the following findings; expansile growth of lesion, increased lysis, endosteal erosion, permeative lesion with destruction of the cortex as depicted by the radiograph of our patient (figures 1 and 2), and presence of soft tissue mass.

A single important imaging feature differentiating a malignant chondroma from a benign tumor is the permeative pattern of the former with true infiltration of the cortical and or the medullary bone, and /or extension into surrounding soft tissue as were noted in the index case.

CT and MR imaging are excellent for assessment of jaw tumours as they depict the exact extent and are very useful for lesion staging. Chondroid matrix mineralization is well demonstrated with CT and the non-mineralized portion of the tumour appears as area of low attenuation [8].

Usually, CSs manifest with bone destruction as was the case in our patient (figure 4). True ossification may also be present, which sometimes corresponds to residual osteochondroma in cases of secondary CS.

3D (volume rendered) CT images (figure 5) are often valuable in assessing the true extent of jaw tumors, particularly with reference to planning reconstructive procedures.

MRI defines the full extent of tumor and is particularly useful in anatomically complex areas. Chondroid lesions in general are of inhomogeneous or homogeneous high signal intensity on T2-weighted spin-echo images but appear as low to intermediate signal intensity on T1W sequence [9]. Intravenous administration of Gadolinium compounds typically shows focal or diffuse enhancement on T1-weighted fat suppressed sequences [9].

It has been suggested that CT or MRI should be used to determine nodal stage of head and neck tumors because clinical estimation of nodal involvement is incorrect in up to 25% of cases [9].

Radionuclide bone scans show increased uptake of tumour in cases of central chondrosarcomas and reveal metabolically active areas in peripheral chondrosarcomas. Whole body bone scans can also be used to identify tumour metastasis [9].

Although CT, MRI and radionuclide scans are quite valuable in determining the nature and extent of the lesion, definitive diagnosis is by histologic evaluation.

CSs have been graded into I, II, and III based on mitotic rate, cellularity and nuclear size [10]. Grade I lesions resembles benign cartilage and they do not metastasize. Grade II lesions demonstrate more myxoid stroma. They recur locally more often than Grade I lesions and have 10% incidence of metastasis. Grade III lesions have a more cellular pleomorphic appearance. They have spindle cell proliferation with a marked increase in the number of mitotic figures. The incidence of metastasis in these lesions is more than 70% [11].

Although Findings on histology of specimen from our patient revealed atypical chondrocytes with a few abnormal mitotic figures (Figure 6) consistent with high grade tumour, there were no evidences of distant metastasis found as at the time of evaluation.

Surgery is the primary treatment for chondrosarcoma. Complete, wide surgical resection is the preferred method when it is feasible [12]. Our patient had subtotal hemimandibulectomy and did excellently well post-op.

CS are relatively resistant to radiotherapy and chemotherapy hence these play limited roles in primary treatment [12]. An exception is their use as adjuvant therapy or palliative treatment for tumours in surgically inaccessible areas or diffuse metastasis.

Tumour grade and resectability are the most important prognostic factors for head and neck CS. Tumour site is another important prognostic determinant [13]. Factors indicating poorer prognosis include histologically positive margins and high-grade tumour differentiation (Grades II and III) [14].

Good prognosis for the lesion is achieved with early recognition and diagnosis followed by wide surgical resection performed as soon as possible.

4. Conclusion

Adequate clinical history and physical examination can provide clues to diagnosis of bone tumors. However, imaging is key in evaluating tumours involving the cranio-facial region and defining their nature weather benign or malignant.

Plain films provide satisfactory features in evaluation of bony architecture and the pathologic processes involved but detailed assessment as in the case discussed above requires CT and MR imaging.

Three-dimensional modalities (CT and MRI) enable surgeons to appreciate the true anatomic location and extent of jaw tumors and aid surgical planning.

Although many tumors and tumor-like conditions have characteristic imaging appearances that almost pin-point their diagnoses, tissue diagnosis is mandatory prior to implementation of therapy.

En-bloc surgical resection of malignant jaw lesions provides patients with the greatest chance of long term survival and cure.

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