Huge Chondrosarcoma of Mandibular Ramus : A Case Report

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Introduction

Chondrosarcoma of the head and neck region is a rare disease, representing approximately 0.1% of all head and neck neoplasms.[6] This malignant tumor, in which the tumor cells form chondroid matrix, represents 10-20% of primary bone tumors and is the second most common sarcoma of bone origin.[9,10] They range from a well-differentiated growth resembling a benign cartilage tumour to a high-grade malignancy with aggressive local behaviour and the potential to metastasize. Only 5% to 10% of chondrosarcomas are known to occur in the head and neck region. Most chondrosarcomas of the head and neck region arise from the maxilla, with relatively few arising from the mandible.[2] Although chondrosarcoma occurs in patients of all ages, most of those affected are over 50 years of age.[3] A case of chondrosarcoma of the mandibular angle ramus region is presented here.

Abstract

A 43 year male patient complains of pain and swelling on right side of the face since 3 months. Patient was apparently normal 3 years back, when he developed a small painless swelling appro. 2×2 cm. over the preauricular region. Initially the swelling was small and gradually developed into a huge one. In past 3 months the swelling has became painful and increased rapidly to the present size.

Extraoral Findings

A diffuse 7-8cm large swelling was present over right side of face extending posteriorly upto posterior border of rumus. It was bordering inferiorly till lower border of mandible anteriorly upto angle of mouth. Superiorly,1cm below the infraorbital margin. overlying skin appeared normal. Swelling was hard to feel and mobile having slightly raised local temperature, and was tender with no signs of paresthesia.

Intraoral Finding

A diffuse swelling was present in the right retromolar region extending upto the buccal mucosa of the 16 region, slightly obliterating the buccal vestibule. Overlying mucosa here was inflamed due to irritation from interference in occlusion and recently performed incisional biopsy .

Investigations

Fine Needle Aspiration Cytology (FNAC) Revealed scanty cellularity composed of chondromyxoid stroma and dissociated cells which are round, oval or spindle shaped. Chronic inflammatory cells were also present. As the material obtained was insufficient for diagnosis so histopathology was adviced to confirm the diagnosis.

Radiograph

Extraoral Radiography

- PA- view revealed a very pathognomic **cotton-wool appearance** in angle ramus area
- Computed tomography shows destructive lesion with central electrodense region .
- 3D Reconstruction revealed sever e destruction of ramus of mandible.

Provisional Diagnosis

- Osteosarcoma
- Fibrous dysplasia

Gross Specimen

Hemi-mandibulectomy was performed, and received specimen showed lobulated structure which appeared brownish in colour. The specimen was approximately 6-7 cm in diameter. The specimen was firm in consistency. Muscle attachments were also seen. Hard tissue structure like bone and teeth were present.

Radiograph of gross specimen shows cotton wool appearance of ramus of mandible.

Histologically

H and E stained section shows lesional tissue showing lobules of hyaline cartilage which are separated by thin fibrous connective tissue septa. The tumor tissue is clearly demarcated from adjacent normal tissue and is composed of densely arranged collagen fiber bundles, fibroblasts , numerous blood vessels and chronic inflammatory cells. There are large number of chondrocyte arranged in pairs, some of the chondrocytes are trinuclated, few are seen singly, at places nuclear and cellular pleomorphism and mitotic figure are also noted. Features were suggestive of chondrosarcoma. Serial sectioning of the tissue specimen revealed the same histological picture, which helped in confirming the diagnosis as chondrosarcoma

Discussion

Chondrosarcomas are slow-growing, malignant mesenchymal tumours characterized by the formation of cartilage by the tumour cells. The exact origin of chondrosarcoms is obscure but the fact is that its basic proliferating tissue is cartilage. Chondrosarcomas are classified into three main types: (1) primary chondrosarcomas, arising from undifferentiated perichondrial cells; (2) secondary chondrosarcomas, arising from metamorphosed cells either in a central chondroma or a cartilaginous exostosis; (3)mesenchymal chondrosarcomas, arising from primitive mesenchymal cells. Surgical excision is the only effective treatment for chondrosarcomas as they are resistant to radiation and chemotherapy.[7] Evans et al. classified chondrosarcomas into 3 grades, from grade I to grade III, according to cellular density, nuclear differentiation, and the size of nucleus. This classification is still currently used.[8] Nevertheless, there have been efforts to classify chondrosarcomas simply into high grade and low grade for better correlation with prognosis.[6] Chondrosarcoma of the jaw occurs primarily in the anterior maxilla, where pre-existing nasal cartilage is present. Chondrosarcoma of the mandible is rare and occurs mostly in the mandibular symphyseal region.[4,5]

The diagnosis is made by biopsy from different sites to establish the malignant nature of the lesion and to differentiate the chondrosarcomas from chondromas. The histology as first defined by Lichtenstein and Jaffe,[11] includes a richly cellular stroma with plump cells, some with multiple nuclei, hyperchromatism of the nuclei and irregularity in the size of the cells and their nuclei. According to Batsakis,[12] mitotic figures may be infrequent and their absence should not rule out a diagnosis of chondrosarcoma. Evans and coworkers have further classified chondrosarcomas into grades I, II and III on the basis of the mitotic rate, cellularity and nuclear size. The differential diagnosis of chondrosarcoma from chondroma is often difficult. In chondroma, a benign neoplasm, most of the cell nuclei are small and single. It is also less cellular than its malignant counterpart. It may be impossible to distinguish a well-differentiated (Grade I) chondrosarcoma from a chondroma.[13]

Conclusion

Survival rate of chondrosarcomas of the jaws in general appear to be poorer than that of chondrosarcomas in other parts of body. Lesions of the jaw account for 1% of all chondrosarcomas reported. Benign cartilage-producing tumours within the jaws are extremely uncommon, but most ultimately prove to represent low-grade chondrosarcomas. therefore, even apparently benign chondrogenic tumours of the jaws should be considered malignant until proven otherwise. In one study, 32% of patients with an initial diagnosis of benign chordoma, chondroma or osteochondroma had a final diagnosis of chondrosarcoma; the median interval before correct diagnosis was 12 months.[2] Chondrosarcomas are generally radioresistant. High-grade chondrosarcomas commonly do metastasize to regional lymph nodes and to long bones more than do other sarcomas.

Moreover, the response of chondrosarcoma to chemotherapy is much poorer than that of osteosarcoma.

These clinicobiologic characteristics of chondrosarcomas, specially the high-grade ones have rendered them to have a protracted clinical course and high recurrence rates, which in turn stresses on the importance of a proper pre-operative clinicopathological evaluation and a regular periodic post-operative screening. Our patient lost to follow 2-3 months after surgery.

References

1. Rajan Saini, Noor Hayati Abd Razak,;Shiafulizan Ab Rahman, Abdul Rani Samsudin,Chondrosrcoma of mandible ; A case report. JCDA www.cda-adc.ca/jcda March 2007, Vol. 73, No. 2
2. Ruark DS, Schlehaider UK, Shah JP. Chondrosarcomas of the head and neck. World J Surg 1992; 16(5):1010-5.
3. Garrington GE, Collett WK. Chondrosarcoma. II. Chondrosarcoma of the jaws: analysis of 37 cases. J Oral Pathol 1988; 17(1):12-20.
4. Izadi K, Lazow SK, Solomon MP, Berger JR. Chondrosarcoma of the anterior mandible. A case report. N Y State Dent J 2000; 66(7):32-4.
5. Ormiston IW, Piette E, Tideman H, Wu PC. Chondrosarcoma of the mandible presenting as periodontal lesions: report of 2 cases. J Craniomaxillofac Surg 1994; 22(4):231-5.
6. Koch BB, Karnell LH, Hoffman HT, Apostolaskis LW, Robinson RA, Zhen W, et al. National cancer database report on chondrosarcoma of the head and neck. Head Neck. 2000;22:408-425.
7. Nishimura Y, Amano Y, Ogasawara H. Chondrosarcoma of the nasal septum: surgical considerations on Le Fort I osteotomy. Eur Arch Otorhinolaryngol 1993; 250: 59- 62.
8 It is written 12 Evans HL, Ayala AG, Romsdahl MM. Prognostic factors in chondrosarcoma of bone. Cancer. 1977;40:818-831.
9. Fechner RE, Mills SE. Atlas of tumor pathology. Armed Forces Institute of Pathology; 1993. (3rd series. Fascicle 8).
10. Barnes R, Catto M. Chondrosarcoma of bone. J Bone Joint Surg. 1966;48:729-764.
11. Lichtenstein and Jaffe HI (1943): Chondrosarcoma of bone, Am J Pathol, 19: 553-89.
12. Batsakis JG: Tumours of the head and neck, Clinical and pathological considerations, (2nd ed.), Baltimore, The Williams & Wilkins Company, page: 384.
13. Hackney FL, Aragon SB, Aufdemorte (1991): Chondrosarcoma of the jaws, clinical findings, histopathology and treatment, Oral Sur Oral Med Oral Path, 71: 139- 43.

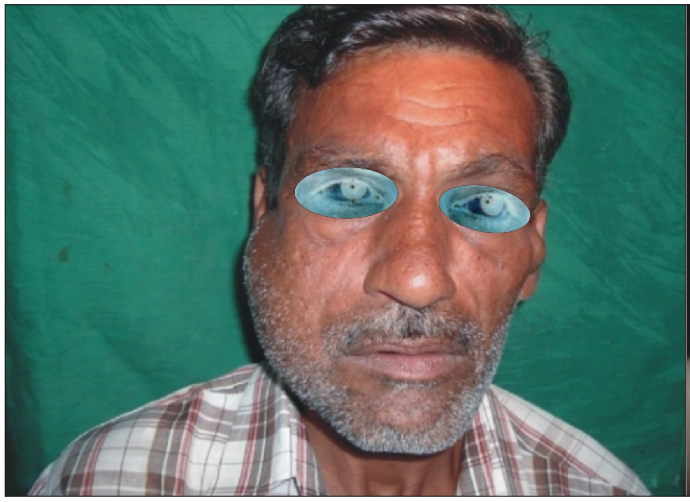

Fig. 1 : Extra Oral Photograph.

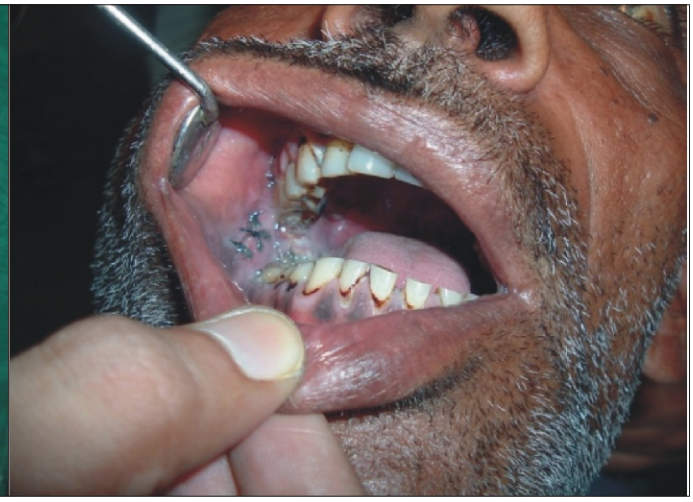

Fig. 2 : Extaoral Photopgraph.

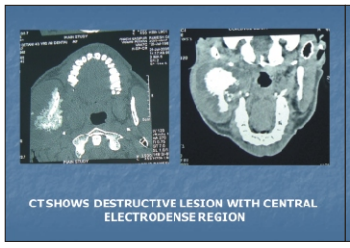

Fig. 3 : CT Scan.

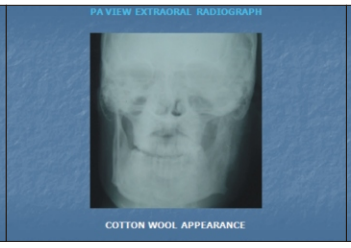

Fig. 4 : PA View.

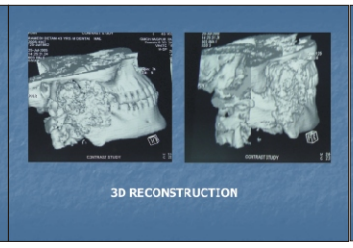

Fig. 5 : 3D Reconstruction.

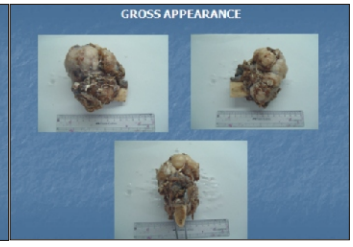

Fig. 6 : (a) Gross Specimen

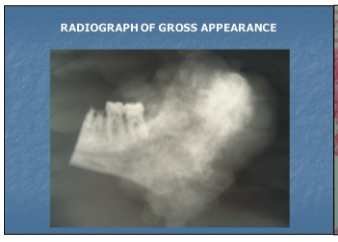

Fig. 6 : (b) radiograph of Gross Specimen

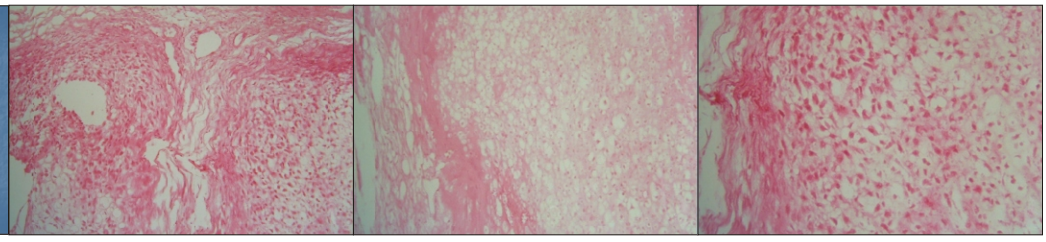

Fig. 7, A. B. C. Histopathology