# Chondroblastic Osteosarcoma: A Review

**K. Kiran Kumar Patro**

*Intern, Institute of dental sciences, Siksha 'O' Anusandhan (Deemed to be University), Bhubaneswar 751003, Odisha, India*

## Abstract

The main purpose of this article to present a review on chondroblastic osteosarcomaand its prevalence in the different jawbones of the body. The word osteosarcoma mainly refers to a group of heterogeneous malignancies that involves bone formation or it can also results due to mesenchymal tissue with evidence of osteogenic disparity. It has been reported around 25% of the pattern of the chondroblastic osteosarcomas represents neoplasms.

***Keywords:*** *Osteogenic sarcomas, chondroblastic osteosarcoma jaw, bonetumors.*

## Introduction

Sarcoma is procured from the Greek word "SERX" which means "Flesh''.A sarcoma is a type of cancer that arises from the transformed cells of mesenchymal origin. These are primary connective tumors.[1]

Osteosarcoma is the most common type of cancer that primarily affects immature bone. It is the most common type of cancer and has a more male prediction ratio. Osteosarcoma occurring in the orofacial regions is extremely sparse, noticing around 1 % of all head and neck region and 7% of all osteosarcomas occurring in the body.[2]

**Types:**

The different types of osteosarcoma are:

**Central:**

- Lowgrade-parosteal;
- Intermediate-periosteal;
- Highgrade- Dedifferiatelyparosteal

**Surface:**

- High-grade 1.Conventional; 2. Telangiectatic; 3. Small cell; 4. Epithelioid; 5. Osteoblastoma-like; 6. Chondroblastoma-like; 7. Fibrohistolytic; 8. Giant cell-rich
- Low grade: 1.lowgrade-central; 2.fibrous dysplasia-like

**Intracortical:**

**Gnathic:**

**Extra Skeletal:**

- High grade
- Lowgrade

Depending on who classifies the chondroblastic osteosarcoma as a histological entity illustrious by the major occurrence of the chondroid matrix, which marks in the direction to give you an idea about an elevated degree of hyaline cartilage as well as is intimately associated through the non-chondroid elements like osteoid, bone matrix.[3]

**Epidemiology, genetic factors and risk factors:** Osteosarcoma is a condition which occurs in both children and adolescence group of people but it is found that it culminates more during the second decade of life. The occurrence rate in children and juvenile age group mostly variety stuck between 3.5 to 4.2 cases

**Corresponding Author:**
**K. Kiran Kumar Patro**
Intern, Institute of dental sciences, Siksha 'O' Anusandhan (Deemed to be University), Bhubaneswar 751003, Odisha, India
e-mail: kiranpatro156@gmail.com

per million inhabitants per year at the same time as the rate in old aged group of individuals is found less .i.e. approximately 1 to 2 cases per million population per year and it is found that middle age group of individuals shows around 1.5 to 4.6 cases per million population per year (25-60) years. Among the risk factors for osteosarcoma, fluoride exposure is found to be the predisposing factor for causing bone cancer. Later no evidence was found clearly regarding this factor.[4]

The exact cause is still unknown but several risk factors are been identified. It is been found that it has arisen from the hereditary multiple exostoses and diseases like Paget disease, fibrousdysplasia, etc. The re are different types of osteosarcoma of jawbones and they can be classified mainly into two groups-

a. Primary

b. Secondary

The cause of the primary subtype of osteosarcoma is still unidentified .but it has been predicted that it may take place due to possible risk factors like genetic as well as environmental factors in addition to the genetic predisposing factors includes the hereditary form of retinoblastoma .i.e. gene alteration,Li-Fraumeni condition. i.e. P53gene transformation, Rothmund – Thomson syndrome. The cause of the resultant subtype of osteosarcoma arises as of the late sequel to craniofacial irradiation, Paget's disease as well as fibrous dysplasia. And it is found that it mainly occurs in grown-up group of patients.[5]

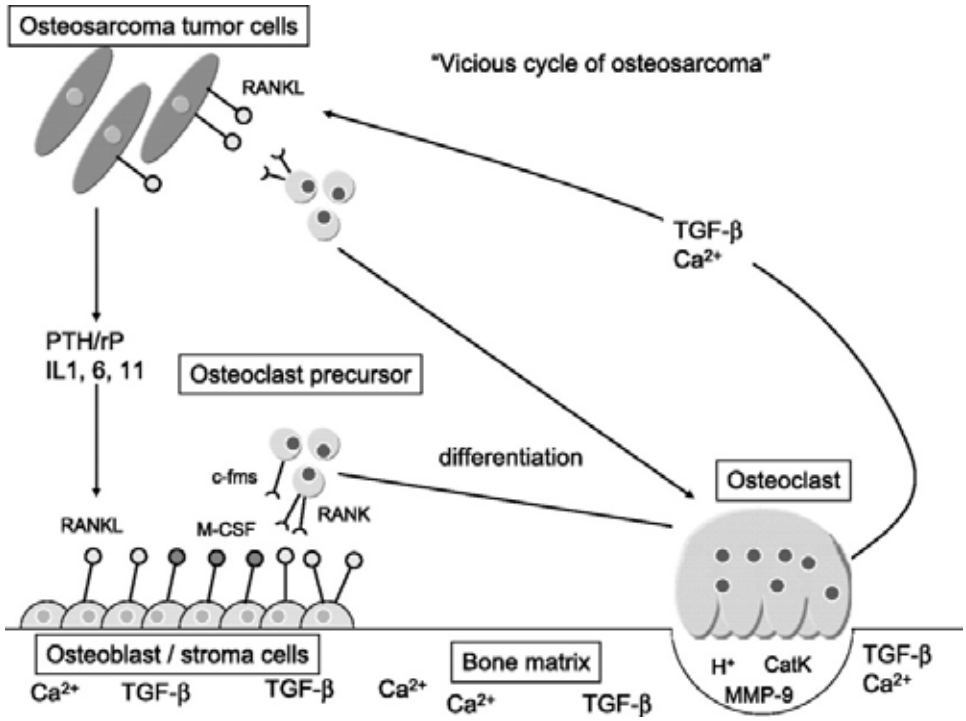



**Figure 1. Vicious cycle of osteosarcoma**

**Clinical Features:** It mainly occurs in the age groups of 20-40 years with predominance in males as compared to females and has a higher occurrence in the posterior mandible as compared to the mandible. In some cases, it has been found that the maxilla and mandible are involved with equal constancy. Sites that often participate in the mandible are at symphysis and the sites that mainly involve in maxilla are the alveolar ridge, palate, sinus floor etc.[6]

These lesions can be purely osteolytic, purely osteoblastic,or mixture of both. Common signs and symptoms include pain,swelling, or both in some cases. Lymphadenopathy is also found where there is unusual focal and regional lymph nodes involvement. The essential microscopic criteria to detect are the direct production of osteoid cells by malignant mesenchymal cells. In addition to these osteoid cells we find the tumor cells producing chondroid type of material and the

different fibrous connective tissue. Other complaints include numbness, facial dysphasia, mobility of the teeth, trismus, headache, nasalcongestion, inability of mouth opening, etc. But it is found that some patients have no prodrome at present time but their tumors can be located incidentally by radiographic imaging.[7]

At first parading, metastatic infection is accessible around 4- 5% of the patients. This occurs in a reduced amount in the patients among appendicular skeleton osteosarcoma. The lungs are the most habitually implicated sites of our body.[8]

**Investigations:** Radiography plays a major role in the initial diagnosis and it is seen that the classic radiographic presentation of osteosarcoma includes Sunray/Sunburst appearance. And this sunburst appearance arises due to osteogenesis of tumor cells. But the extent of the tumor in both hard and soft tissue is been better appreciated in the CT Scan, MRI imaging, Angiogram which mainly detect the vascular displacement and the relationship of the vessel to the lesion and the same time it also helps to determine the vascularity of the involved tumour.[9]

Immunohistochemical characterization plays an important role in the diagnosis of osteosarcoma which includes mainly the osteonectin and osteocalcin-osteocalcin is specific for osteoblast cells but osteonectin is not specified for osteoblasts and the other biomarkers that are used apart from osteonectin and osteocalcin are cytokeratin, P53, P16, Ki67, CD99, S100, caveolin-1, etc. Some laboratory standards like lactate dehydrogenase, alkaline phosphate serum levels possibly will be escalated in a small number of collections of patients. Even though they do not keep up a correspondence with disease proportions, exceptthey may have anegative prognostic importance.[10]

**Codman's Triangle:** It is formed primarily in the angle between the elevated periosteum and the cortex's underlying surface.

In many cases, CORTICAL BREACH is commonly observed. Histologically, there is the presence of belligerent malignant mesenchymal cells that produce undeveloped bone. And the stroma mainly consists of malignant connective tissue with anaplastic spindle cells surrounded in the mesenchymal parenchyma.[11]

Chondrosarcoma comprises of cartilage along with varying degrees of cellularity and maturation. It is found that normal chondroid lacunar formation is visible, but this feature may be rarelyseen in poorly differentiated tumors. The tumor can be often seen in globule growth patterns with thin fibrous connective septa-separated tumor lobules.

The central areas of the lobules show the highest degree of maturation. Calcification or ossification may occur within the chondroid matrix. Neoplastic cartilage may be replaced by bone-like normal endochondral ossification.[12]

It can be divided into 3 histopathologic grades of malignancy and this grading system is well correlated with the growth rate of tumors and the prognosis of extragnathic skeleton chondrosarcomas.

**Grade I:** Chondrosarcomas powerfully bear a resemblance to the manifestation of chondromas, consisting of the chondroid matrix as well as chondroblasts displaying no more than a slight deviation of normal cartilage outer shell. The cartilaginous matrix calcification or ossification is prominent, as well as the mitosis is rare.

**Grade II:** Chondrosarcoma shows a larger portion of moderately sized nuclei and increased cellularity, especially in the lobules with a less prominent hyaline matrix. However, the mitotic rate is low.

**Grade III:** Chondrosarcoma is exceedingly cellular with may show prominent spindle cell propagation mitoses may be important. Simply identifiable cartilaginous matrix containing lacunae cells may be scarce. Eventually develop into higher grade osteosarcoma with poor prognosis.[13]

**The different histopathologic variants are as follows:**

1. Conventional type-osteoblastic, chondroblastic, and fibroblastic osteosarcoma
2. Telangiectatic/osteolytic type osteosarcoma
3. Diminutive cell osteosarcoma
4. Low downgrade central osteosarcoma
5. Periosteal osteosarcoma
6. Periosteal osteosarcoma
7. Secondary osteosarcoma
8. Elevated grade surface osteosarcoma
9. Extraskeletal osteosarcoma

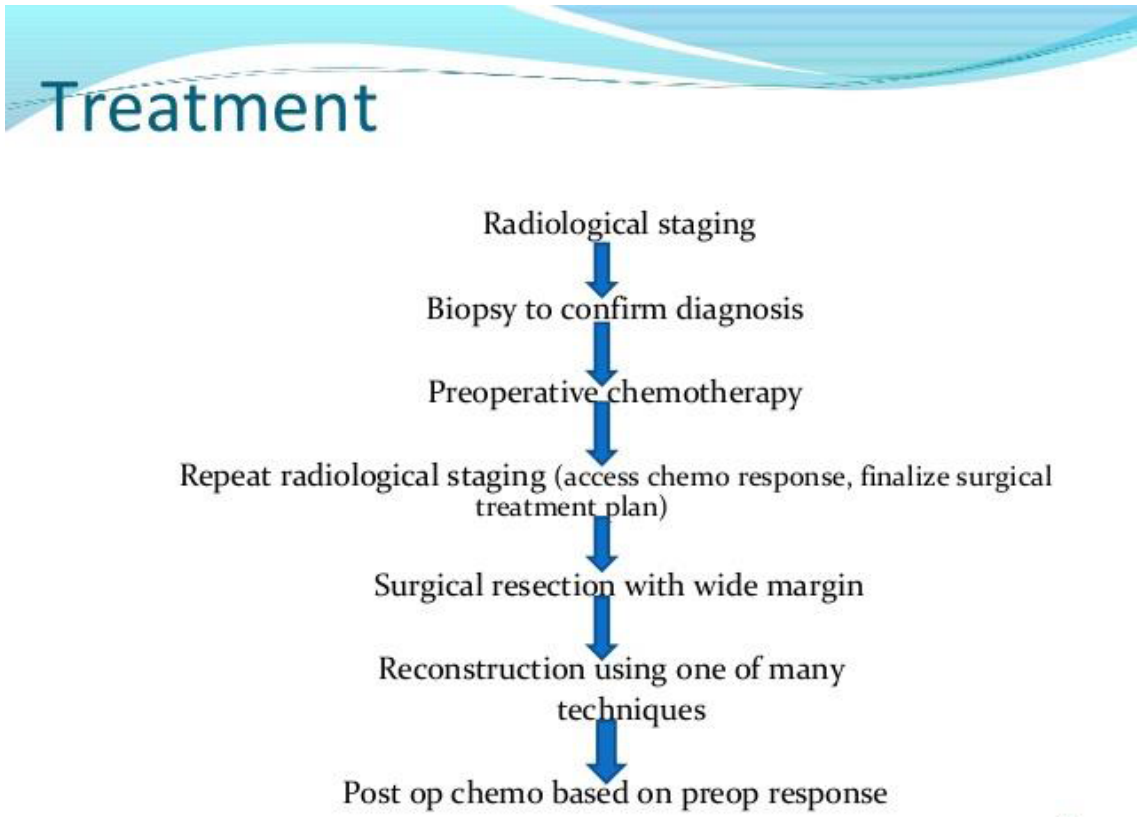
The prognosis feature is mainly evaluated by the Enneking system which for the most part evaluates the histological grade of the tumor (G), the extent of the primary tumor (T) as well asmetastasis to the close by lymph nodes or other concerned organs (M).

Amongst all the diverse histological types as well as subtypes chondroblastic type is more impervious to treatment and exhibits unfavorable prognosis. Whereas we can find Fibroblastic types that have a rather better prognosis and as well as responds to the treatment.

**Treatment:** The management is finished with a permutation of therapies together with surgery, chemotherapy as well as radiation therapy. However, they have to undertake three months of chemotherapy, acknowledged as neo-adjuvant therapy previous to surgery if the patient is originated to have high-grade tumors.[14]

**Surgery:** As these cases for the most part lies in the skeletal orientation, surgery is a central component of osteosarcomas treatment too in the head as well as neck region. The modality of the management, principles of the surgical management largely depends on the position of the tumor, the dimension of the tumor. Obtaining disease-free resection margins is of course dictatorial, to keep away from the risk of local reappearance. on the other hand, this objective is even extra complicated to reach when dealing through the head as well as neck osteosarcomas, as resecting few millimeters, more often means imperil significantly functional structures, through a noticeable reduction in the patient's quality of life. While in the intraoperative strength of mind of resection margins might represent a useful tool in other head as well as neck malignancies. Intraoperative pathological examination does not consent for the evaluation of bone margins. In the meantime, only soft tissue margins can be evaluated through the intraoperative consultation. It is found that the best management results is been obtained when the criteria .i.e. the histopathologic evaluation, biopsy of the desired site and the extent of the neoplasm is been determined, before the treatment. Multimodal mode of treatment is indicated or we can say that the best analeptic option is when we are dealing with the high grade of osteosarcomas but only alone surgery can be done if the neoplasm comes under the low grade, parosteal and central group of osteosarcomas. Provided if the metastatic potential of the neoplasm is found to be absent.[15]

**The treatment procedure can be understood as follows:**



## Conclusion

Chondroblastic osteosarcomas of the mouth remain "Delphic''. Anamount of difficulty associated with their diagnosis as well as treatment is yet to be determined. True synchronous multicentric osteosarcomas of the mouth are tremendously uncommon but like other osteosarcomas of the jaw, have a successful result. The palliative resection modality of the management in such lesion through difficult however can show the way to an enormously enhanced quality of life in addition to may even offer the window for cure. Bountiful studies have been reported that the molecular pathogenesis of the osteosarcoma as well as the related biomarkers associated with means of the tumor. Even though many studies are exceedingly favorable as well as there has been no upgrading in the reoccurrences along with survival rates of osteosarcoma. Our facts and data of the pathway caught up in sarcoma genesis deficient, as well as new perceptiveness are enthusiastically awaited in the perspective of developing a forceful target therapy to come together with surgery.

**Ethical Permission:** Not required

**Conflict of Interests:** None

**Funding:** None

## References

1. Parkas O, Varghese BT, Mathews A, Nayak N, Ramchandran K, and Pandey M. Radiation-induced osteogenic sarcoma of the maxilla. World J Surg Oncol. 2005; 3:49. 2.
2. Lima JJ, Manzi R, Silva FL, Baptista MZ. Extra-skeletal osteosarcoma in the cervical region - literature review and case report. Radiol Bras. 2002; 35:315–9.
3. Chindia ML. Osteosarcoma of the jawbones. Oral Oncol. 2001; 37:545–7.
4. Silva HR, Borges AC, Pizza M, Borsato ML, Castro HC, Luporini SM, et al. Osteosarcoma and acute myeloid leukaemia - two cases in children. Rev Bras Hematol Hemoter. 2006; 28:76–8.
5. Guerra RB, Tostes MD, da Costa Miranda L, Pires de Camargo O, Baptista AM, Caiero MT, et al. Comparative analysis between osteosarcoma and Ewing's sarcoma: Evaluation of the time from onset of signs and symptoms until diagnosis. Clinics (Sao Paulo) 2006; 61:99–106.
6. Alves MT, Miiji LN, Marinho LC, Petrilli AS, Jesus-Garcia R, Tolledo SC, et al. Human osteosarcoma with high immunoreactivity of P 53, erbB-2 and P-glycoprotein, and correlation with anaplasia parameter. Bras Patol Med Lab. 2008;44:107–14.
7. Castro HC, Ribeiro KC, Bruniera P. Osteosarcoma: Experience of the pediatric oncology service of holy house of mercy in São Paulo. Rev Bras Ortop. 2008;43:108–15.
8. Rech A, Castro CG, Jr, Mattei J, Gregianin L, Di Leone L, David A, et al. Clinical features in osteosarcoma and prognostic implications. J Pediatr (Rio J) 2004;80:65–70.
9. Neville BW, Damm dd, Allen CM, BouquotJE oral and maxillofacial pathology,3rd edition. philadelphia: Saunders,2009.
10. Longhi A, Errani C, De Paolis M, et al. Primary bone osteosarcoma in the pediatric age: state of the art. Cancer Treat Rev 2006;32:423–36.
11. Shirazian S, Agha-Hosseini F. Oral osteosarcoma: A case report and analysis of previously reported cases. N Y State Dent J 2014;80:50-4.
12. Nissanka EH, Amaratunge EA, Tilakaratne WM. Clinicopathological analysis of osteosarcoma of jawbones. Oral Dis 2007;13:82-7.
13. Paparella ML, Olvi LG, Brandizzi D, Keszler A, Santini-Araujo E, Cabrini RL. Osteosarcoma of the jaw: An analysis of a series of 74 cases. Histopathology2013;63:551-7.
14. Yildiz FR, Avci A, Dereci O, Erol B, Celasun B, Gunhan O. Gnathic osteosarcomas, experience of four institutions from Turkey. Int J Clin Exp Pathol2014;7:2800-8
15. Neville BW, Damm DD, Allen CM, Bouquot J. Oral and Maxillofacial Pathology. 3rd ed. St Louis: Mosby Elsevier; 2013