

## Clinico Pathological Analysis of Chondroblastoma: A Tertiary Care Centre Experience

Sindhu Nair P<sup>1</sup>, Jayasree K<sup>1\*</sup>, Sreejith G Nair<sup>2</sup> and Venugopal M<sup>3</sup>

<sup>1</sup>Department of Pathology, Regional Cancer Centre, Trivandrum, Kerala, India

<sup>2</sup>Department of Medical Oncology, Regional Cancer Centre, Trivandrum, Kerala, India

<sup>3</sup>Department of Imageology, Regional Cancer Centre, Trivandrum, Kerala, India

\*Corresponding Author: Jayasree K, Professor and Head, Department of Pathology, Regional Cancer Centre, Trivandrum, Kerala, India.

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### Abstract

Chondroblastomas are benign chondroid neoplasms composed of chondroblasts usually arising in epiphysis or apophysis of skeletally immature patients. They account for less than 1% of all bone tumors. Even though long bones are most commonly affected it can affect craniofacial bones, talus, calcaneum, patella and vertebra. Within the craniofacial region the temporal bone is most frequently affected. Here we present a series of chondroblastoma cases diagnosed in our Department within last ten years with special emphasis on those cases occurring in rare sites like temporal bone, vertebra and talus. This study is a retrospective analysis of cases of chondroblastoma diagnosed in the Department of Pathology, Regional Cancer Centre, Trivandrum during the time period of ten years from 1<sup>st</sup> January 2009 - 30<sup>th</sup> September 2019. Clinical features like age, sex, site, presenting symptoms were analyzed along with radiology and histopathology in each case. A total of thirteen cases of chondroblastoma in the age range of 10 - 56 years of age and M:F ratio of 8:5 were received during the study period. Five cases were reported in long bones, four in temporal bone, three in talus and one in vertebra. Radiology of majority of cases showed lytic lesions with a sclerotic rim and some showed fluid-fluid level in MRI. Histopathology section showed chondroblasts admixed with variable number of osteoclastic giant cells and chondroid matrix. Most of them showed the characteristic chicken wire calcification while some had secondary aneurysmal bone cystic areas. Occasional cases show increased mitosis while some show cellular atypia also. We emphasize the need for a combined clinical, radiologic and histologic approach to the diagnosis of chondroblastomas especially in rare sites and with varied histology.

**Keywords:** Chondroblastoma; Temporal Bone; Talus; Vertebra; Aneurysmal Bone Cyst

### Introduction

Chondroblastomas are benign chondroid neoplasms composed of chondroblasts usually arising in epiphysis or apophysis of skeletally immature patients. They account for less than 1% of all bone tumors. More than 75% of the cases involve the long bones the most common anatomical sites are the epiphyseal region of the distal and proximal femur, proximal tibia and proximal humerus. Other classic sites of involvement include the talus, calcaneum and patella. Within the craniofacial region the temporal bone is most frequently affected [1]. Here we present a series of chondroblastoma cases diagnosed in our Department within last ten years with special emphasis on those cases occurring in rare sites like temporal bone, vertebra and talus.

## Materials and Methods

This study is a retrospective analysis of cases of chondroblastoma diagnosed in the Department of Pathology, Regional Cancer Centre, Trivandrum during the time period of ten years from 1<sup>st</sup> January 2009 - 30<sup>th</sup> September 2019. Clinical and radiological data were obtained from the medical records library and consultation files. The H&E stained slides of all the cases were retrieved and studied. The clinical data included age, sex, presenting symptoms, site of involvement and radiological findings. Histopathological analysis included the presence of chondroid matrix, chicken wire calcification, aneurysmal bone cyst (ABC) component, necrosis, atypia and mitosis.

## Results

There were 13 cases of chondroblastoma in the study period. 8 were males and 5 females. There were cases ranging between 10 - 56 years of age. Seven of them were in the age group 10 - 20 years, four in the group 20 - 30 years and 2 in the group of more than 30 years (Figure 1). Four presented within the temporal bone with tinnitus and hearing impairment, three in the talus with chronic pain, three in the tibia, two in the humerus and one in vertebra with pain and localised swelling (Figure 2). Those in the long tubular bones (n = 5) presented with lytic lesion with a sclerotic rim. Histopathology section showed the characteristic uniform round to polygonal cells with well-defined cytoplasmic borders, clear to basophilic cytoplasm and round to oval nuclei (chondroblasts) (Figure 3) often growing in sheets. They exhibited longitudinal grooves and one or more small or inconspicuous nucleoli. Osteoclastic giant cells were seen randomly distributed. Chondroid matrix (Figure 4) was present in all cases while chicken wire calcification (Figure 5) was present in three cases only. Mitosis was sparse in all the cases and there was no nuclear atypia. Aneurysmal bone cyst like area was present only in one case. In the temporal bone all four cases had CT scan which was suggestive of a well-defined lytic lesion in temporal bone with surrounding oedema. One case showed fluid-fluid level also (Figure 6). The histopathology section showed the typical sheet like proliferation of small to intermediate-sized round to polygonal cells having eosinophilic cytoplasm with a centrally placed nucleus and a central, longitudinal nuclear groove ("coffee bean" nucleus) some showing a tiny nucleoli. Three of them were osteoclastic giant cells rich while one showed only scattered osteoclastic giant cells. All of them showed chondromyxoid matrix and chicken wire calcification. All the four showed aneurysmal bone cyst like areas. Haemosiderin laden macrophages were prominent in temporal bone cases. Two of the cases showed mild atypia as the cells were large with enlarged nuclei and mild pleomorphism. Mitosis was sparse. One case showed brain parenchymal infiltration and it recurred thrice. Three cases in talus (Figure 7) also showed lytic lesions with calcification. One case was showing marked atypia of nuclei suggesting malignancy. But the radiology picture was benign. Another case showed extensive aneurysmal bone cyst areas with one tiny fragment showing the typical chondroid matrix and round to oval cells with the grooved nuclei, which clinched the diagnosis. In all cases osteoclastic giant cells were there, but the number was variable. One case showed focal necrosis. Mitosis was sparse in two cases. One case was mitotically active. But no atypical mitosis was noted. Chondroid matrix was there in all the three. But only one showed chicken wire calcification. The case in the vertebra was a 45 year old male with a lytic lesion. Histopathology was typical with chicken wire calcification and chondroid matrix. There were many osteoclastic giant cells with secondary aneurysmal bone cyst. But there was moderate atypia with enlarged pleomorphic nuclei. There was no necrosis and mitosis were sparse. Primary diagnosis of chondroblastoma was given by FNAC in two cases; the first was temporal bone case which recurred thrice and the other was the case in humerus, which were confirmed later by biopsy.

## Discussion

Chondroblastoma was first described in 1931 by Codman who originally described an "epiphyseal chondromatous giant cell tumor of the proximal humerus" with the diagnosis corrected to chondroblastoma of bone by Jaffe and Lichtenstein in 1942 [2,3]. Chondroblastoma is typically seen in an epiphyseal location and is characterized histologically by the proliferation of immature chondrocytes (chondroblasts) along with other secondary elements, such as mature cartilage, giant cells, calcification, and occasionally, aneurysmal bone cyst formation. Chondroblastoma tends to be a solitary lesion. The neoplastic cell is the chondroblast, a cell that normally populates areas of secondary ossification. In young people, it preferentially occurs in the epiphysis of long tubular bones, with the most common sites being

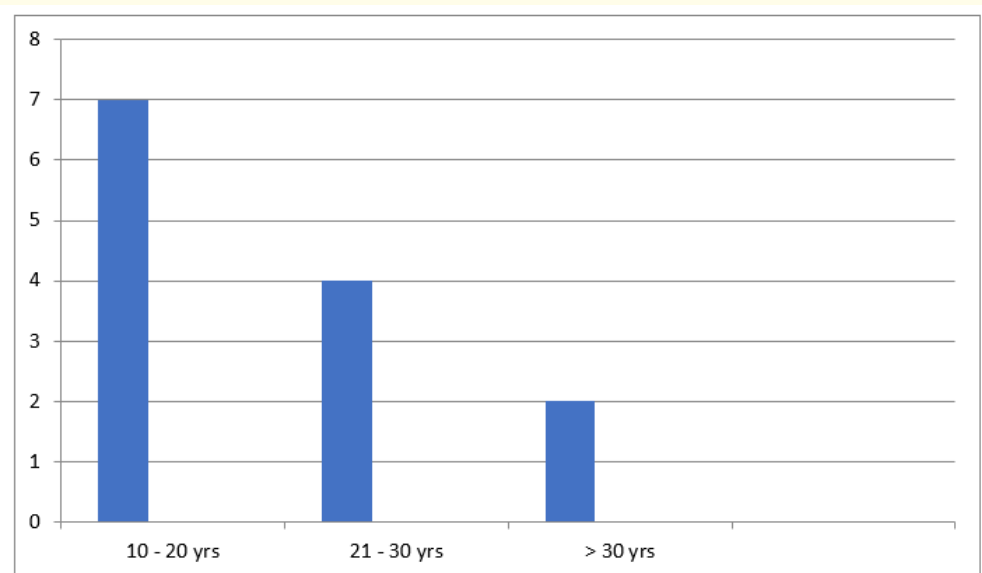


Figure 1: Age distribution.

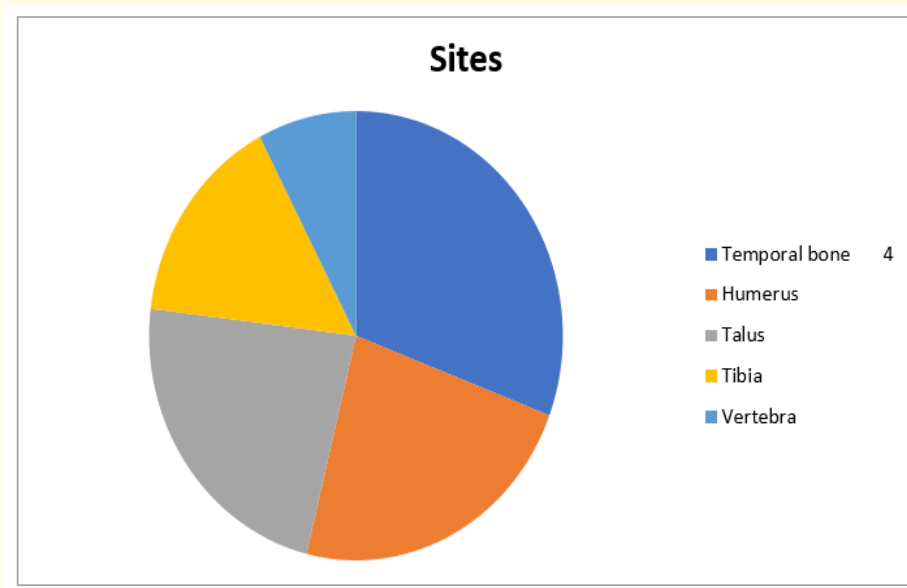
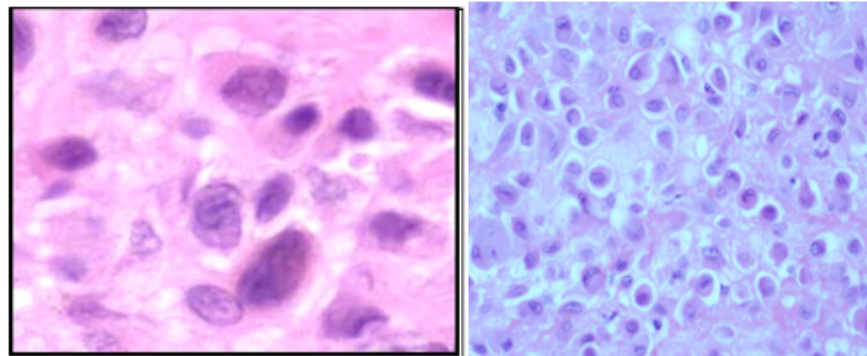
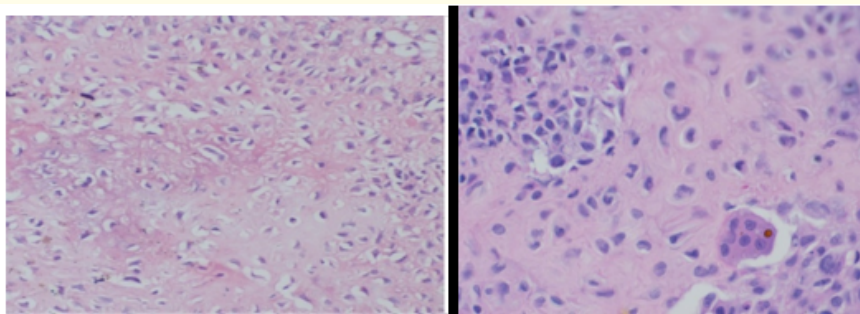


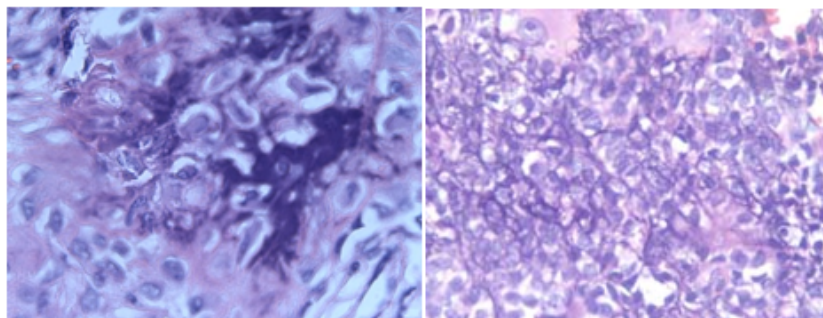
Figure 2: Site distribution.



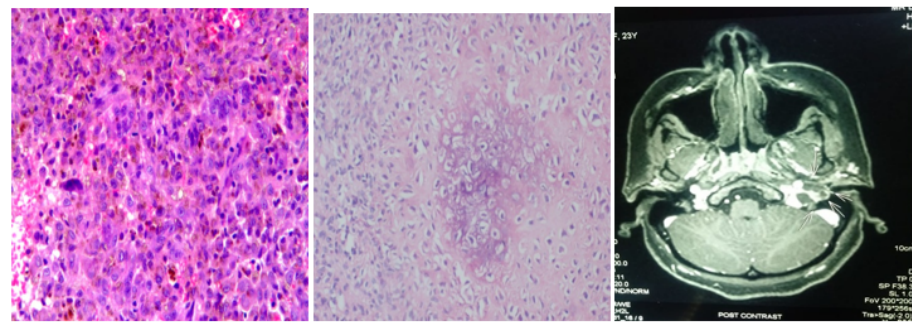
**Figure 3:** Chondroblast with grooved nuclei.



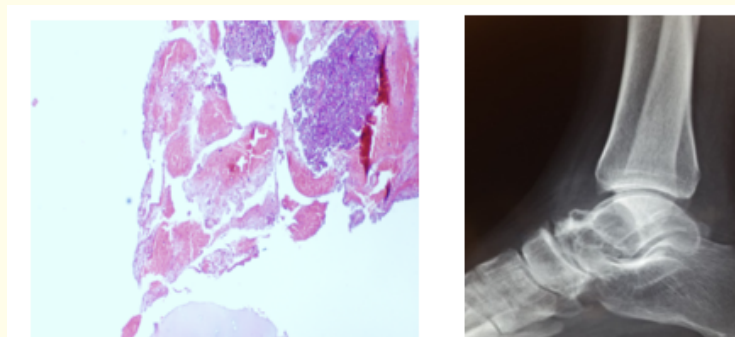
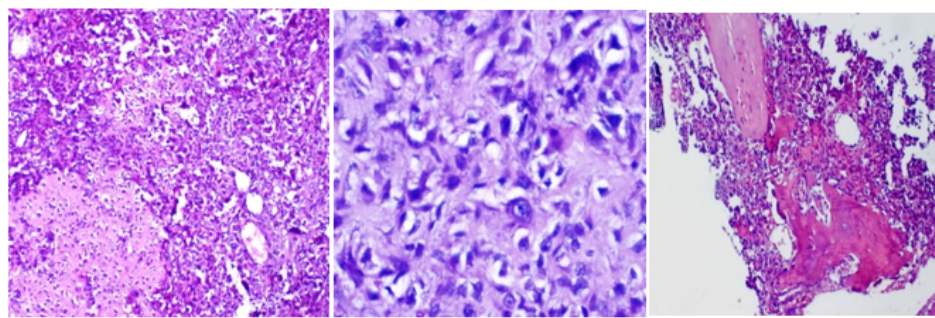
**Figure 4:** Chondroid matrix and osteoclastic giant cells.



**Figure 5:** Chicken wire calcification.



**Figure 6:** Temporal bone CB with chicken wire calcification, chondroid matrix and hemosiderin laden macrophages.



**Figure 7:** Lesion talus with atypia, permeation to soft tissue and secondary ABC.

the proximal tibia or femur, distal femur, and proximal humerus. In older individuals, the location is much more variable, and tumors may involve nontubular bones such as the craniofacial skeleton or bones of the hands and feet. Tumors in unusual locations, atypical clinical presentations, or complicated by aneurysmal bone cyst may pose diagnostic difficulty. Allgner, *et al.* in his study suggests that chondro-



blastoma should be classified as a specific bone-forming rather than cartilage-forming neoplasm [4]. The extracellular matrix composition and gene expression pattern analysis of chondroblastoma shows that type II collagen, which is the main component of all cartilage matrix, is not expressed by chondroblastoma cells and is not deposited into the extracellular tumor matrix. Instead, osteoid and fibrous matrix is formed, with its typical biochemical composition. There is multifocal expression of aggrecan proteoglycan in most chondroblastomas, which explains the bluish, pseudochondroid appearance of some matrix-rich areas.

While the age range is broad (2 - 83 years), most chondroblastomas are diagnosed in the second to third decade of life, with an average age of 19 to 23 years with a male predominance [5,6]. For example, in a study of 495 chondroblastomas, Kurt., *et al.* found that the mean age for patients with skull and facial bone lesions was 44.1 years. For those with flat bone lesions, the mean age was 28.4 years; for hand and foot lesions, 20.8 years; and for patients with long bone lesions, 19.1 years [7]. Our patients were in the age range 10 - 56 years, 7 of them between 10 - 20 years. Our temporal bone cases were above 20 years except for the one which was detected at the age of 14 years. Among the three cases in talus two were above 20 years and one was 19 years old. Our vertebral case was 45 years and all our long bone cases were under 20 years. As in the previous studies [5,8,9] in our study also males dominated with a M:F ratio of 8:5. The predominant bone affected was the temporal bone (4 cases) in our series which was against other studies in the literature, may be due to referral bias and less number (n = 13) of cases compared to other large series [5,7-9]. According to Luke B Reid., *et al.* there are a total of 81 reported cases worldwide of chondroblastoma of the temporal bone in the English Literature [10].

Histopathologic examination of all cases showed the typical round to oval cells with typical grooved nuclei admixed with variable number of osteoclastic giant cells. Even though chondroid matrix was present in all cases, chicken wire calcification was present in 9 cases. These are bluish or purple granular calcium deposits seen in the cytoplasm or stroma, where they demonstrate a delicate pericellular lace like or "chicken-wire" appearance' diagnostic of chondroblastoma. According to W Chen., *et al.* it is seen in approximately one-third of cases, most commonly in long bone tumors [11]. Chicken wire calcification was seen in all of our cases in the temporal bone and vertebra but absent in two out of three cases in talus and two out of five cases in long bones. According to W Chen., *et al.* the osteoclastic giant cells are variable in chondroblastoma [11]. But de Silva., *et al.* in their study of 46 cases of chondroblastoma described 11 cases with abundant giant cells [12]. In our study the cases from long bones had a smaller number of giant cells. Rest of the cases except one in temporal bone had numerous giant cells. When the number of giant cells increases the chances of them misdiagnosing as giant cell tumour of bone are high. But in GCT, the number of giant cells is more and they are evenly distributed compared to chondroblastoma. Adequate sampling will reveal the chondroid matrix, the chondroblast with typical grooved/convoluted nuclei and chicken wire calcification in chondroblastoma.

Aneurysmal bone cyst is not uncommonly seen as a secondary feature in chondroblastoma and therefore if a biopsy includes prominent ABC-like areas, the primary lesion may be overlooked. Careful attention must therefore be paid to clinicoradiologic features to allow distinction between these two entities, and specimens should be sampled thoroughly. Findings such as the location in the epiphysis, the presence of calcification, and histologic evidence of solid areas with chondroblastic differentiation should lean one toward a diagnosis of chondroblastoma in challenging cases [1]. ABC areas were noted in all our temporal bone cases and that from vertebra. But it was present only in one case each from long bones and talus. In the case from talus ABC areas were extensive and after thorough search we could identify a fragment with chondroid matrix. According to W Chen., *et al.* secondary ABC is seen in chondroblastoma of hands and feet while that of temporal bone cases show haemosiderin deposits. All our temporal bone cases showed ABC and hemosiderin laden macrophages. Huvos., *et al.* in 1970 found aneurysmal bone cyst "engrafted on" 20% of 25 chondroblastomas [13] while Dahlin., *et al.* in their study identified ABC changes in 20 of their 125 cases (16%) [14]. Schajowicz., *et al.* observed ABC changes in 17% cases [15].

Worrisome features like cellular atypia and suspicious soft tissue extension was noted in one of the cases in talus which was referred as osteosarcoma. Other unusual features are increased mitosis and necrosis [11,12]. Among 13 of our cases only one case in talus was mitotically active. But no atypical mitosis was noted in that case. According to WHO the number of mitosis usually present in chondroblastoma is 1-3/10 HPF [1]. Atypia was present in four of thirteen cases. Two in temporal bone, one in talus and the one in vertebra showed atypia.

It was mild in temporal bone cases but was moderate in talus and vertebra, but none of them showed increased mitosis. It was sparse in these cases. Necrosis was there in three out of thirteen cases. On follow up, these cases are doing well without disease.

Although chondroblastoma is a benign lesion, it may show aggressive features such as large size, pulmonary metastases, joint and soft-tissue infiltration and local recurrence. It shows a more aggressive behaviour in the flat bones than in the long bones [16]. One of our temporal bone case recurred thrice as it infiltrated adjacent brain parenchyma at initial presentation itself. It showed mild atypia, but mitosis was sparse. Temporal bone chondroblastomas may complicate the diagnosis because of their different histologic characteristics [17]. In temporal bone cases a diagnosis of chondroblastoma is given after thorough sampling for the characteristic histological features and radiological correlation.

### Conclusion

Increased awareness of the morphologic spectrum of chondroblastomas will enable pathologists to avoid diagnostic pitfalls. We emphasize the need for a combined clinical, radiologic and histologic approach to the diagnosis of chondroblastomas especially in rare sites and with varied histology.

### Conflict of Interest

Nil.

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