

A Study on Clinical and Radiological Profile of Benign Cartilage Tumors of Long Bones

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Abstract: ***Introduction:** Cartilaginous tumors are second most primary bone tumors and are characterized by formation of hyaline or myxoid cartilage. Benign cartilage tumors are common than malignant ones³³. Enchondroma of appendicular skeleton are usually symptom free and found incidentally on evaluation of trauma or adjacent joint pathologies⁴. Benign cartilaginous lesions behave in an inactive fashion and do not require intervention, unless they are symptomatic or cause functional impairment or complications from fractures. The clinical management and prognosis depend on the site and the histological diagnosis. With limited tissue, the differential diagnosis may be problematic, particularly with regard to distinguishing enchondroma from low - grade chondrosarcoma. In the current WHO classification system Grade 1 Chondrosarcoma has been renamed as “Atypical Cartilage Tumors” (ACT). ACT never metastasizes and is not considered as malignancy⁵. Surgical treatment consists of curettage alone or curettage with bone grafting or bone cementing. **Materials and Methods:** Patients with benign cartilaginous lesions of long bone diagnosed clinically and radiologically from Orthopedic Oncology clinic at the Amala Institute of Medical Sciences from January 2012 to January 2018 were included in study. Sample size was found to be 48. Consecutive sampling was done. Patient details collected from tumor registry maintained in the Orthopedic oncology clinic. Patients was examined at 6 monthly intervals for a period of 5 years in the absence of symptoms or sooner in case of symptoms like recent onset of pain at site of lesion or visible swelling. Surgically treated patients will be subjected to clinical examination and radiological examination if symptomatic. Radiological modality used was plain radiographs. **Results and Discussion:** Forty - eight patients were included in the study. The mean age group of patients was found to be 43.15 ± 17.31 (19 - 83) years. Male were patients 21 (43.75%) and female patients were 27 (56.25%). Most common bone affected was Distal Femur 32 (66.67%). Pain was presenting complaint in 45 (93.75%) patients. In 3 (6.25%) patients the tumor was an incidental finding. Lytic lesion with specs of calcification was most common x - ray finding in 43 (89.58%) patients. Most common pre operative histological finding was Enchondroma in 32 (91.43%) patients, in 3 (8.57%) patients it was Grade 1 chondrosarcoma. Surgical intervention was done in 35 (72.92%) patients, while 13 (27.08%) patients were managed conservatively. In surgically treated patients 32 (91.43%) had post operative biopsy finding of enchondroma, in 3 (8.57%) patients report came as low - grade chondrosarcoma. In surgically treated patients 32 (91.43%) had final diagnosis of enchondroma, in 3 (8.57%) patients as Low - grade chondrosarcoma. In conservative management group 13 (100%) patients had enchondroma as final diagnosis. In conservative management group the cause of symptom was found to be unrelated to tumor. Two patients presented with pain during follow - up in surgical group, one patient was 65 years old female who underwent curettage and bone grafting for enchondroma of distal femur at third 6 monthly follow up, another patient was 65 year old male patient who underwent curettage and bone cementing for distal femur for grade 1 chondrosarcoma fifth 6 monthly follow. Both the patients underwent x - rays and it showed no signs of recurrence. No further complaints during subsequent follow up. None of the patients in surgically treated groups had recurrence. in patients managed conservatively the lesion was found to be stable and asymptomatic. **Conclusion:** In this study we have followed up 48 patients with benign cartilage tumours of long bones attended orthopaedic oncology clinic. Most commonly patients presented with pain. Tumor was an incidental finding in 3 patients. Most common radiological finding was lytic lesion with specs of calcification. Distal femur was involved in 32 patients. Surgical intervention was done in 35 patients, while 13 patients were managed conservatively. Patients managed conservatively had symptom unrelated to tumour and lesion was found to be stable in subsequent follow - up. Both groups had 6 monthly follow - ups. None of the patients had local recurrence or new onset of symptoms related to tumor. In the small group of conservatively managed patients, it appears to be a safe option if strict follow up is assured and symptoms are unrelated to tumour.*

Keywords: Enchondroma, Atypical Cartilage Tumors, Grade I Chondrosarcoma, Curettage, Bone grafting, Bone cementing

1. Introduction

Cartilage tumors account for majority of primary bone tumors and are characterized by formation of hyaline or myxoid cartilage. Benign tumors are common than malignant ones. Cartilage - forming tumors comprise of enchondroma and osteochondroma encompassing the overwhelming majority of benign cartilaginous³³.

Benign cartilaginous lesions behave in an inactive fashion and do not require intervention, unless they are symptomatic or cause functional impairment, or complications from fractures. The clinical management and prognosis depend heavily on the location and the histopathological diagnosis. Limited tissue from needle biopsy, the differential diagnosis may be problematic, particularly in distinguishing enchondroma from low - grade chondrosarcoma³⁴. These

tumors may show characteristic clinical and radiologic features that allow for this distinction; however, the histologic overlap in the well - differentiated lesions can make a definitive diagnosis difficult, and some studies have shown significant inter observer variability among pathologists. Enchondroma is a rest of hyaline cartilage within the medullary canal of long bones and tubular bones of the hands and feet comprising 12–24 % of benign bone tumours, they are often painless and incidentally found.

In case of pathological fracture EC can present with pain. Peak incidence is in third decade of life. X rays show a central radiolucent lesion in the metaphysis with lobular margins and a narrow zone of transition. Matrix mineralization can be variable, but characteristically occurs as “rings and arcs” and is best seen on CT scan. Mineralization is usually absent in the hands and feet. MRI

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shows uniform low signal on T1 sequence. T2 sequence has a high signal due to the water content of hyaline cartilage with small areas of low signal representing mineralization. There should not be MRI enhancement or bone scan activity. Histology shows hyaline cartilage with minimal chondrocytes with no nuclear atypia or mitotic figures. Hand ECs looks aggressive under the microscope. Treatment consists of observation with serial radiographs, if asymptomatic. Curettage and grafting with optional internal fixation may be considered for multiple fractures, impending fracture, or painful lesions. EC should not progress or recur. Clinical indication of increasing pain or imaging showing progressive growth, endosteal scalloping cortical destruction, or a soft tissue mass needs a biopsy. Isolated lesions rarely transform.

Larger and more proximal lesions are having more risk. Hand ECs are rarely to transform despite their histologic appearance. Noninherited conditions with multiple ECs exist. They often affect one side of the body and are at greater risk of secondary transformation, in adulthood. Ollier's Disease consists of multiple EC whereas Maffucci Syndrome is multiple EC with soft tissue haemangiomas and or lymphangiomas. They are associated with 20–30 % chance of malignant transformation. In adulthood, these patients should be monitored with periodic chest CT and whole - body bone scan. Areas with activity on bone scan needs MRI. There is a fine line between the diagnosis of EC and grade 1 chondrosarcoma. The latter is treated with intralesional curettage and grafting. More aggressive chondrosarcomas are widely excised³¹.

Many authors have proposed radiographic follow - up instead of surgery for cartilaginous tumors in the long bones without signs of local aggressiveness. If the lesion is unrelated to symptoms it may lead to morbidity. Radiographic follow - up instead of surgery may prevent over treatment of this group of patients, resulting in less morbidity and lesser cost⁵

2. Review of Literature

Cartilaginous tumors form the second largest group of primary tumors affecting bone. They all have characteristic production of chondroid matrix by tumor cells. Cartilage tumors ranges from completely benign lesions to highly malignant. They are sub - divided by location in central, intramedullary and surface or peripheral sites. Cartilage lesions can be diagnosed using plain X rays or MRI. Differentiation between enchondroma versus low - grade chondrosarcoma, osteochondroma versus peripheral chondrosarcoma and chondroblastoma versus clear - cell chondrosarcoma are difficult. Biopsy of malignant lesions can be false negative due to sampling errors as most cartilage lesions have grades of malignancy and can even present benign sections.¹

Enchondromas are commonly diagnosed incidentally. They are commonly associated with nearby joint or soft tissue pathologies, which are main source of the symptoms. Even a small, well - defined lesions are often confused with a sarcoma or other malignancies, which may be due to the lack of exposure in the field of bone tumors for both the

radiologists and general orthopedic surgeons².

Omlor and et al suggested feasibility of intralesional curettage strategies for symptomatic benign to low - grade malignant chondrogenic tumors. Surgery, however, did not prove superior compared to conservative clinical and radiological observation in asymptomatic patients. Due to the low risk of transformation into higher - grade tumors and lessen morbidity, asymptomatic lesions might just be observed if continuous follow - up is assured³.

Akoh and et al reported Enchondroma growth was a rare event and typically occurred at two years follow - up series. Given the low risk for malignant transformation, we propose surveillance with plain radiographic follow - up for stable enchondromas every 3 - 6 months for the first year and then annually for at least three years of total follow - up. The most significant costs savings can be made by limiting MRI imaging in the absence of clinical or radiographic concern. Additional studies are needed to determine the long - term risk of growth or declaration of chondrosarcoma⁴.

Deckers and et. al followed up conservatively treated enchondroma and ACT, only 6% of the patients had a medical indication for surgery. This study shows that indication for surgery should be discussed more thoroughly. Based on results they recommend annual radiologic follow up for asymptomatic enchondroma or ACT in the long bones, irrespective of tumor size⁵. There is consensus that endosteal scalloping of more than two thirds of the cortical thickness indicates malignant transformation of enchondromas (EC), and that suspicious EC should be followed up with regard to their potential development to chondrosarcoma. There is no consensus to date on the frequency and duration of follow - up. Enchondromas that are smaller than 2 cm will remain benign if they are without cortical contact. Some of the signs suggestive of aggressive tumors and the recommendations for their management—e. g., tumor larger than 5 cm in the tubular bones, growth of more than 6mm over 12 months, and follow - up regimen after 12 months—are not fully accepted or scientifically proved yet⁶.

Cartilaginous tumors identified as low - grade on pre - operative imaging with or without additional image - guided needle biopsy can safely be managed as low - grade without pre - operative histological diagnosis. A few tumors may demonstrate high - grade features histologically, but the rates of recurrence are not affected⁷.

Benign lesions are best left untouched if free of symptoms and radiologically inactive such as enchondroma of the phalanges. En bloc resection of osteochondroma including the pseudo capsule is curative and results in a very low recurrence rate. Malignant cartilage tumors, if low - grade central chondrosarcoma of the long bones, can be treated with intralesional surgery in combination with some adjuvant (phenol, cryosurgery). High grade chondrosarcoma (including clear - cell chondrosarcoma) and all chondrosarcoma of the axial skeleton should be surgically resected with wide margins with an intermediate risk of local recurrence.

Feasibility of intralesional curettage strategies for symptomatic benign to low - grade malignant chondrogenic tumors was supported. Surgery, however, did not prove superior compared to conservative clinical and radiological observation. Due to the low risk of transformation into higher - grade tumors and better functional results, more lesions might just be observed if continuous follow - up is assured.⁹

Assessment of chondral lesions begins with a clinical evaluation and radiographs. Longitudinal follow - up with serial radiographs is appropriate in cases without evidence of aggressive radiographic features. Concerning radiographic features include periosteal reaction, soft - tissue extension, cortical destruction, endosteal scalloping of greater than two - thirds of the native cortex, larger lesions of size (≥ 5 cm), and location in the axial skeleton. Biomarkers include IMP3, SOX4, microRNA, and Periostin may be used as an adjunct in histological assessment which to help differentiate benign enchondroma from a low - grade chondrosarcoma¹⁰.

Advanced - imaging studies, such as computed tomography (CT), bone scans, magnetic resonance imaging (MRI), dynamic contrast - enhanced DCE - MRI, and Fluorodeoxyglucose Positron Emission Tomography (FDG - PET), may be considered for borderline cases. Aggressive or concerning X rays features should prompt evaluation with advanced imaging or referral to an orthopaedic oncologist.¹⁰

Enchondroma is a common benign intramedullary neoplasm, composed of mature hyaline cartilage. Enchondromas account for 10–25 % of benign bone tumours. Most common from the second to the sixth decades with a peak in the fourth and fifth decades. It is most frequently found on the small tubular bones of the hands and feet. Among the long tubular bones, enchondroma occurs frequently in the proximal humerus and proximal and distal femur and tibia. Histopathological features are variable according to the location of the lesion. It is very important to consider the site of the lesion together with the radiological findings and clinical features. Enchondromas are made of mature hyaline cartilage arranged in lobules. Cartilaginous lobules are surrounded by normal bone marrow.

Chondroma - This term is also applied to benign cartilage neoplasia of soft tissue or periosteal origin³².

Sites of Involvement

Most frequently on the small tubular bones of the hands, where it is the most common tumor. Among the long tubular bones, enchondroma occurs more frequently in the proximal humerus and proximal and distal femur and tibia. Rarely involves the pelvic bone, ribs, scapula, or sternum. Enchondroma in the spine is very rare. Almost never occurs in the craniofacial bones, that develop through intramembranous ossification³².

Clinical Symptoms and Signs

Usually found on routine radiologic examination or as hot spots on the bone scan without specific symptoms. The majority is asymptomatic. In the hands and feet, enchondromas present as palpable swelling with or without pain. In this location, they may frequently present with

pathological fracture and pain. Chondroid tumors located in hands and feet, in subperiosteal location, and in the lining of joint capsules are almost clinically benign. Behavior of a cartilage neoplasm is best predicted having in mind if the lesion is growing or not. Clinical, radiographic, and histological features should be interpreted in the light of this information. Enchondromas usually do not grow after puberty³².

Image Diagnosis Radiographic Features

A sharply defined border, mild endosteal scalloping, and calcifications resembling a "Ring or an Arc" characterize the Xray findings of enchondroma of the hand or foot. Spotty calcifications are commonly seen. Calcifications may be dense sometimes. (Figure 1) Cortical expansion or pathological fracture is a potential feature, predominantly in tumors of the hands and feet. Since enchondroma is a common type of benign tumor, solitary lytic bone lesions with well - defined margins and scalloping's of the endosteum in a bone of the hand should be considered an enchondroma until proven otherwise. Enchondroma also shows typical radiological features on other sites. In long tubular bones, pattern is of a central or eccentric location with an osteolytic lesion and calcification in the medullary portion. Radiographic findings of enchondroma occurring in flat or irregular, nontubular bone may be hard to recognize (Figure 2)³².



Figure 1: Enchondroma proximal phalanx. X ray shows well defined lytic lesion with stippled calcification in proximal phalanx



Figure 2: Enchondroma of distal femur. X ray shows conglomeration of stippled calcification

CT Scan

Provides better evaluation of the cortical endosteal surface in medullary located lesions of long tubular bones. It is a better investigation for the study of lesions located in areas of difficult radiographic assessment, as in case of axial skeleton.

MRI Features

The margin of the tumor is well defined by a low signal intensity on T1 - weighted images and a high signal intensity on T2 - weighted images. Lobulated margins of the tumor are characteristic, and also low signal intensity is seen at the calcified zones. The x - ray volume of the enchondroma overlaps the volume of the lesion as seen on MRI. (Figure 3)

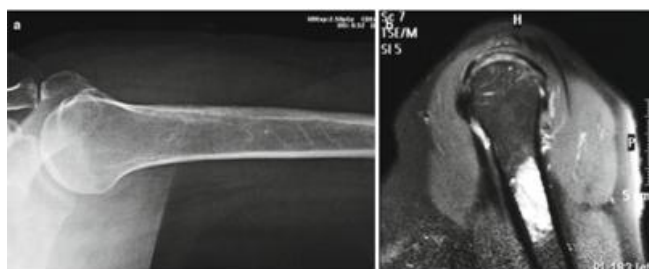


Figure 3: (a) X ray shows a lytic lesion in meta - diaphysis of humerus. (b) MRI T2 weighted image of same lesion. The radiographic volume of lesion overlaps with that of MRI.

Image Differential Diagnosis

Low - Grade Chondrosarcoma - Differentiating between a low - grade chondrosarcoma and enchondroma is challenging, and radiology is useful adjunct in this diagnosis. Radiologically, chondrosarcoma of the long bone shows expansion of bone and thickening of cortex. On x - ray and CT scan of tubular bones, extensive endosteal scalloping, cortical expansion, cortical thinning or thickening, cortical spread, and ambiguous matrix calcification is suggestive of chondrosarcoma. Peripheral marrow edema around the lesion, subperiosteal edema, and mass in soft tissue, as seen in MRI, are critical findings that point towards a low - grade chondrosarcoma. MRI volume of the lesion bigger than x - ray volume is suggestive of malignancy. Epiphyseal location of the tumor also strongly indicates Chondrosarcoma³².

Pathology

Gross Features - Most enchondromas are curetted, so that it is rare to examine an intact gross specimen. Enchondromas are confluent masses of bluish, semi - translucent hyaline cartilage with a peculiar lobular arrangement.

The size of the lobules varies from millimeter to 1 cm. It is rare to find one greater than 5 cm in size. Periphery of the lesion is irregular, since individual lobules bulges into the surrounding marrow spaces. Punctuate calcifications seen on X rays can be appreciated grossly. Multiple enchondromas shows a similar gross appearance to that of solitary tumors. In multiple enchondromas, normal marrow elements encircle the well circumscribed nodules, which are mistaken for an aggressive pattern of permeation³².

Histological Features

Histological features are variable and on the location of the lesion. It is very important to consider the location of the lesion together with the X rays findings and clinical features. Enchondromas are made of mature hyaline cartilage arranged in lobules. Cartilaginous lobules are surrounded by normal bone marrow (Figure4).

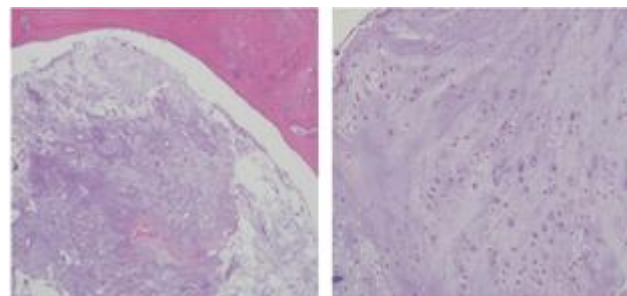


Figure 4: (a) Histology of enchondroma. Low power shows lobular hyaline cartilage with normal trabecular bone. b) The hyaline cartilage tumors are blue grey in color. They show lobulated pattern with low cellularity.

Enchondral ossification may also be seen. Other lobules of enchondroma are frequently partially encased by mature lamellar bone. In general, there is low cellularity and abundant extracellular matrix (Figure 5).

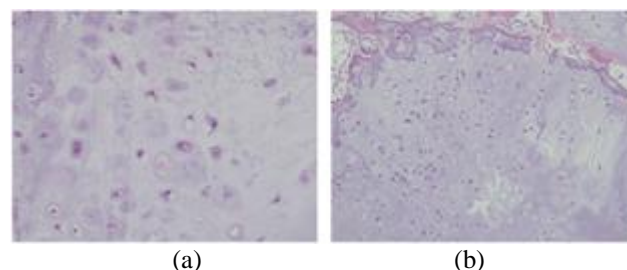


Figure 5: a) At high power the cytological features of chondrocytes are small dark nuclei, single nucleus in a lacunar space. b) Occasionally at periphery of cartilage lobule, calcification is evident.

In enchondromas of the hands and feet, there may be increased cellularity. Usually, chondrocytes are located inside lacunae, and the nucleus has a small round regular appearance. On a low power view, the nucleus is hardly visible. chondrocytes can be seen as clusters or sheets rarely.

Occasionally, chondrocytes can be seen as large cells or double - nucleated cells but they lack cytological atypia. Calcification can be seen as fine purple - colored granular precipitate or a big mass. Prominent myxoid change or necrosis is hardly observed in enchondromas. However, they can be seen occasionally in the small bones of the hands and feet. When these histological features are present, even if cellularity is low, the chance of malignancy should be considered. In case of small bones of the hands and feet, in order to make diagnosis of malignancy, there should be extreme hypercellularity, nuclei pleomorphism, and marked myxoid changes. Calcifying or ossifying enchondroma is a variant that can be present at the metaphyseal region of the long bones. In these cases, enchondroma can be seen as radiologically dense ringlike or flocculent agglomerated and heavily calcified and ossified radiopacities. Histologically, the lesion are made of mature hyaline cartilage and heavily calcified or ossified material with small scarce cartilage cells.

Pathologic Differential Diagnosis

- Low - Grade Chondrosarcoma - If arising from metaphysis of the long bone in middle - aged to elderly patients. Grossly, mucoid or myxoid changes are commonly observed in chondrosarcoma and rarely in enchondromas. Enchondroma do not show cortical erosion. Histologically, increased cellularity, marked myxoid change of the stroma, and cytologic features of malignancy are characteristic in chondrosarcoma. Separation of cartilage lobules by fibrous bands is observed in low - grade chondrosarcoma. The most important differential diagnostic feature is intramedullary invasion of tumor. In chondrosarcoma, the trabeculae of preexisting bone are embedded within the tumor tissue – entrapment of native trabeculae. X rays of enchondromas of the small bones of the hands and feet show cortical thinning; however, microscopic spread into the cortical tissue is not observed. Enchondromas of the pelvis, spine, ribs, and sternum are rare. Chondroid lesions occurring in these locations should be considered as aggressive cartilage lesions even if they are not reactive or metaplastic in nature.
- Fibrous Dysplasia - Chondroid differentiation observed in fibrous dysplasia is also different from enchondroma. This can be differentiated on the basis that in fibro chondroid dysplasia, fibro osseous elements are found in addition to cartilaginous nodules³².

Rationale of study

Benign cartilage tumors involving long bones are usually asymptomatic, and are commonly recognized as an incidental finding identified either on standard radiographs for unrelated trauma or joint disease or on a radionuclide bone scan for the investigation of unrelated musculoskeletal pathologies.

Regional pain is more frequently related to a nearby joint or a local soft - tissue disorder than to the tumor itself. An asymptomatic cartilage tumor in a long bone does not require treatment beyond follow up by sequential clinical assessments and radiographic evaluations to rule out progression. The morbidity of curettage or even biopsy usually outweighs any potential benefit for these indolent lesions.

In the absence of features of malignancy, a short period of conservative therapy (rest and non - steroidal anti - inflammatory medication) will often relieve symptoms and allow pain related to chondroid lesions to be distinguished from joint - mediated pain.

In an orthopedic oncology clinic, a large number of patients with cartilage tumors are seen, and often the clinical and radiographic findings easily point to the benign nature of the lesion. As such, the purpose of this study was to understand the natural course of benign cartilage tumors in such patients and recurrence in surgically treated patients.

Objectives

- 1) To evaluate natural course of benign cartilage tumors which are treated non operatively by clinical and radiological assessment.
- 2) To find out incidence of local recurrence in surgically

treated patients.

- 3) To find out incidence of cases which needed intervention during follow up.
4. To find out how to effectively follow up patients with benign cartilage tumors

3. Methodology

Study Design: Cohort study.

Study Setting: Department of Orthopaedics Amala Institute of Medical Sciences

Study Period: The duration is 3 years. Commenced after obtaining ethical committee clearance. May 2021 TO JANUARY 2023

Sample Size

Sample Size Calculation:

$$N = \frac{\sum (1 - \alpha/2)^2 X p X q}{d^2}$$

α = Significance level, p=Prevalence of distal femur cartilage tumor 67%5, q=1 - p, d=Absolute precision =20/100 n=48

Study Subjects

In this study we had included patients with benign cartilage tumors attending orthopedic oncology clinic in Amala Institute of Medical Science. Patient's data will be taken from tumor registry maintained in Department of orthopedics.

Inclusion Criteria

- 1) Patients with benign cartilaginous lesions of long bone diagnosed clinically and radiologically from Orthopedic Oncology clinic at Amala Institute of Medical Sciences from January 2012 to January 2018.
- 2) Age > 18 years
- 3) Cartilage lesions in the long bones of the extremities with follow - up time of minimum 5 years since initial diagnosis.

Exclusion Criteria

- 1) Patients with Olliers disease, Maffucci syndrome. Patients lost during follow up.

Sampling Technique

Consecutive sampling

Methods of Data Collection

Patients with benign cartilage tumors attending orthopedic oncology clinic will be evaluated clinically and radiologically initially. If patients are asymptomatic, they will be kept under follow up. Details will include patient symptoms, radiology report, our initial diagnosis, and any need for biopsy or surgical intervention like curettage as well as final histological diagnosis, Patient details will be collected from tumor registry maintained in Orthopedic oncology clinic. Patients during the period January 2012 to January 2018 will be included in the study who are meeting inclusion criteria.

Patients has been examined at 6 monthly intervals for a period of 5 years in absence of symptoms or sooner in case of symptoms like recent onset of pain at site of lesion or visible swelling. Surgically treated patients will be subjected to clinical examination and radiological examination if symptomatic. Radiological modality using for follow up will plain radiographs.

Outcome Measurement

Patients were kept under active surveillance by 6 monthly clinical examination and radiological examination for a period of 5 years. Patients were contacted through telephone regarding symptoms if they fail to come for review.

Factors taken into account: New onset of pain at the site of tumor. Suspicious radiological changes. Need for surgical intervention like biopsy or curettage. Incidence of pathological fracture during follow up.

Statistical Analysis

Descriptive analysis was carried out by mean and standard deviation for quantitative variables, frequency, and proportion for categorical variables. Non normally distributed quantitative variables were summarized by median and interquartile range (IQR). Data was also represented using appropriate diagrams like bar diagram, pie diagram and box plots. Categorical outcomes were compared between study groups using Chi square test /Fisher's Exact test (If the overall sample size was < 20 or if

the expected number in any one of the cells is < 5, Fisher's exact test was used.). P value < 0.05 was considered statistically significant. Data was analysed by using coGuide software - BDSS Corp. Released 2020. coGuide Statistics software, Version 1.0, India: BDSS corp.

4. Results and Discussion

In this study we studied clinical and radiological profile of benign cartilage tumors of long bones. Forty - eight patients met with the inclusion criteria and were included in the study. The duration of follow up was 5 years. The mean age of study population is 43.15 with SD of 17.31 years. The median age is 39 (19 - 83) years. In the study population there were 21 males (43.75%) and 27 females (56.25%).

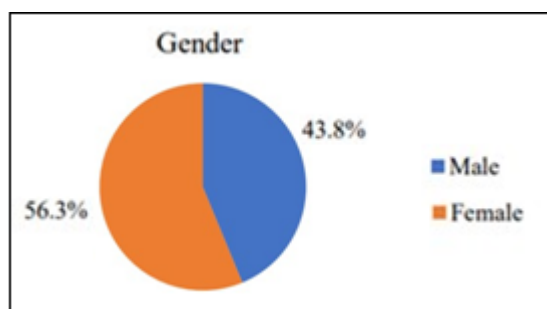


Figure 6: Pie chart of gender in the study population

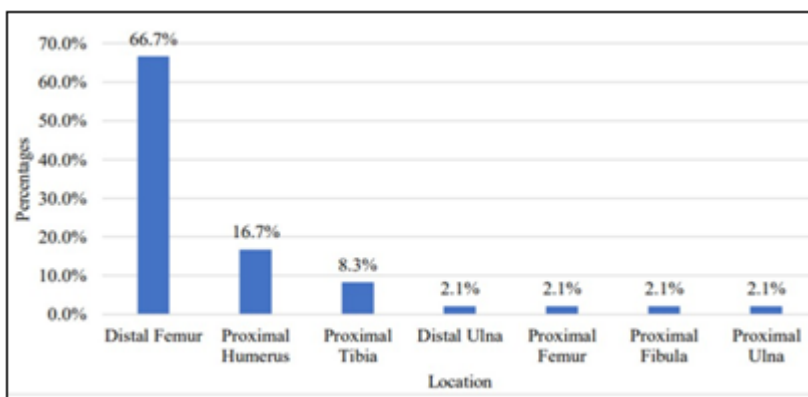


Figure 7: Bar chart of location of tumor in study population

Distal Femur is involved in 32 patients (66.67%). Proximal Humerus is involved in 8 (16.67%) patients. Proximal Tibia is involved 4 (8.33%) patients. Distal Ulna Proximal Femur, Proximal Fibula, Proximal Ulna are involved in rest of the study population one each 2.08% respectively. Deckers et al. studied natural course of enchondroma in 49 patients mean age of patients were 49 (20 - 76) years. Mean follow - up 67 months. Gender distribution was male 22 (45%) Female 27 (55%). Location of lesions were distal femur 33 (67%), proximal femur 3 (6%), proximal humerus 8 (16%), proximal tibia 3 (6%), distal tibia 1 (2%) and proximal fibula 1 (2%) 5.

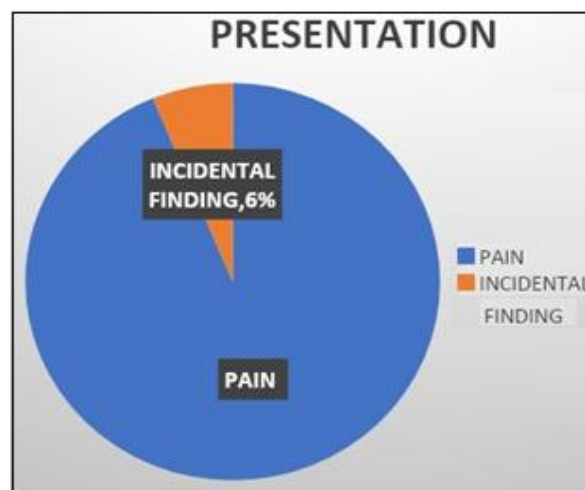


Figure 8: Pie chart showing cases which are incidental finding and cases presented with pain study population

Benign cartilage tumor was an incidental finding in 3 patients (6.25%), while regional pain was presenting complaint in 45 patients (93.75%). Donthineni and et. al retrospectively evaluated medical records of 115 patients with Enchondroma, among them patients, 88 (76%) presented with pain of the affected area and the rest were found incidentally².

Lytic lesion with specs of calcification was the x ray finding in 43 (89.58%) patients, calcified lesion in 4 (8.33%) and healed lesion in one patient (2.08%). The classical radiographic appearance of enchondroma includes an intramedullary, frequently eccentric, mineralized lesion with growth pattern. Calcifications classically are described as "ring and arc" pattern¹⁷.

CT scan was taken in 18 (33.33%) patients report came as enchondroma. CT demonstrates similar imaging features as radiographs, although CT allows for a more detailed assessment of the lesion of interest. It has been shown to be superior to radiographs in demonstrating internal mineralized matrix, cortical destruction, endosteal scalloping, and periosteal reaction¹⁸. It is of particular use in evaluating mineralization in the spine and pelvis, which may be difficult to assess on radiographs¹⁹. CT can be used to assess for soft - tissue extension in mineralized lesions; however, this is more apparent on MRI.

In 27 (56.25%) patients MRI report came as Enchondroma, in 3 patients MRI report showed enchondroma/low grade chondrosarcoma. to assess on radiographs. On MRI, chondroid lesions are well - defined lobulated lesions that are predominantly fluid signal on all sequences and contain central areas of low - signal mineralized matrix. Post - contrast enhancement is typically septal and rim - like, which corresponds to areas of the lesion where fibrovascular septations lie between the lobules of hyaline cartilage. These imaging features are highly recognizable and can limit the imaging differential considerations to chondroid tumors. Lesions with predominantly intermediate signal on T1 - weighted sequences and adjacent bone - marrow changes elevate concern for malignancy²⁰. In addition to the radiographic features favoring malignancy, MRI can further discriminate benign and malignant by demonstrating soft - tissue tumor extension and peritumoral marrow edema, which is more commonly associated with chondrosarcoma

Dynamic contrast - enhanced (DCE) - MRI measures the concentration of gadolinium shortly after injection to provide data on the perfusion within the lesion. Studies have noted that early enhancement (<10 sec) may be indicative of a low - grade chondrosarcoma. compared with enchondroma. However, although DCE - MRI is able to differentiate low - grade from high - grade chondrosarcoma, in is limited in its ability to differentiate low grade chondrosarcoma from enchondroma, which adds little information to that provided by the more conventional imaging techniques^{21, 22}. As such, this modality is infrequently utilized.

PET scan was taken in 2 (4.17%) patients which showed low uptake. Pre operative histopathology report came as

enchondroma for 32 (91.42%) patients and for 3 (8.53%) patients it came as low grade chondrosarcoma.

The use of FDG - PET scan to differentiate between benign and malignant cartilaginous lesions has also recently been of interest. Results suggest that an SUVmax (maximum standardized uptake value) that is very low (< 2) can be used to exclude a high - grade malignancy, while neoplasms with an SUVmax of >4.4 are strongly suggestive of malignancy^{23 - 25}. However, as with DCE - MRI, there is inter - mediate range (SUVmax between 2.0 and 4.4) wherein PET struggles to differentiate benign and low - grade chondroid lesions.

Post operative histopathology report came as enchondroma for 32 (91.47%) patients and for 3 (8.53%) patients it came as low - grade chondrosarcoma. Biopsy should be considered for lesions that have concerning clinical or radiographic features. On histologic examination, typical features consistent with benign enchondroma include nodules of cartilage surrounded by marrow fat or the encasement of cartilage by lamellar bone²⁶. Concerning histologic bones². features include bone permeation, increased cellularity, atypia, binucleation, and myxoid changes. Any signs of marrow fat invasion are consistent with chondrosarcoma²⁶. The difficulty with basing treatment on a histologic diagnosis is the poor reliability in differentiating a benign enchondroma from a low - grade chondrosarcoma. A study by the Skeletal Lesions Interobserver Correlation among Expert Diagnosticians (SLICED) Study Group reported only fair interrater reliability (kappa = 0.443) among 9 expert musculoskeletal pathologists in the classification of cartilaginous lesions in long bones²⁷. The poor reliability of current histopathologic techniques highlights the difficulties of relying on histology alone to differentiate low - grade cartilaginous lesions.

In study population Enchondroma was diagnosed in 45 patients (93.75%) and low grade chondrosarcoma (grade 1 chondrosarcoma/ Atypical Cartilage tumors) in 3 patients (6.25%). Thirty - five patients (72.92%) underwent surgical management. Conservative management in case of 13 patients (27.08%). During third follow up one (2.86%) patient presented with pain over surgical site. Another patient (2.86%) during fifth follow up presented with pain. X rays were taken and found no signs of recurrence.

There is no incidence of recurrence in surgically treated patients and lesion was stable in conservative group. In case of distal femur 23 patients underwent surgery (65.71%) and 9 patients underwent conservative management (69.23%). One patient (7.69%) with distal ulna tumor underwent conservative management. Patient with tumor of proximal femur underwent surgical management (2.86%). In case of proximal humerus 6 patients underwent surgical management (17.14%) and 2 patients were managed conservatively (15.38%). In case of proximal tibia 3 patients underwent surgery (8.57%) and one patient underwent conservative management (7.69%). Patient with proximal ulna lesion underwent surgical management (2.86%). Figure 9.

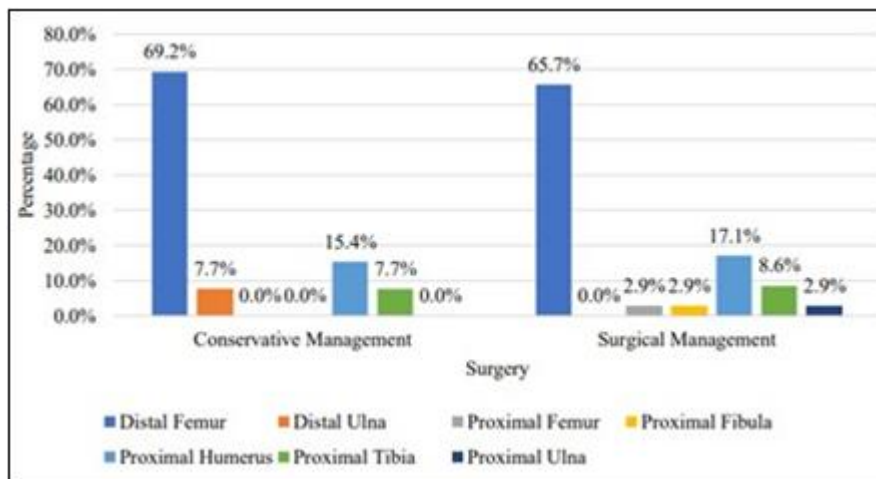


Figure 9: Bar chart showing location of lesion in conservative and surgical management group

In surgically treated patients 35 (100%) patients presented with pain. In conservative management group 3 (23.08%) patients had incidental finding and 10 (76.92%) patients presented with pain.

In surgical management group 35 (100%) patients had lytic lesion in their x rays, in conservative group 8 (61.54%) patients had lytic lesion, one patient (7.69%) had healed lesion and calcified lesion in 4 (30.77%) patients. In surgical group 13 (37.14%) patients had CT scan finding of enchondroma, in conservative management group 3 (23.08%) patients.

In the surgically treated group 23 (65.71%) patient had enchondroma as MRI finding, in 2 (5.71%) patients report came as enchondroma/low grade chondrosarcoma. In conservative management group, 4 (30.77%) patients had finding of enchondroma, in one patient (7.69%) report came as enchondroma/low grade chondrosarcoma.

PET scan was taken in 2 patients one (2.86 %) in surgical and one (7.69%) in conservative group, shows low uptake.

In surgically treated patients 32 (91.43%) had pre operative biopsy finding of enchondroma, in 3 (8.57%) patients report came as low - grade chondrosarcoma.

In surgically treated patients 32 (91.43%) had post operative biopsy finding of enchondroma, in 3 (8.57%) patients report came as low - grade chondrosarcoma.

In surgically treated patients 32 (91.43%) had final diagnosis of enchondroma, in 3 (8.57%) patients as Low - grade chondrosarcoma/ACT. In conservative management group 13 (100%) patients had enchondroma as final diagnosis.

No fresh complaints were reported in conservative management group. In surgically treated patients during third follow up one (2.86%) patient presented with pain over surgical site. Another patient (2.86%) during fifth follow up presented with pain. X rays were taken and found no signs of recurrence. None of the patients in both surgically treated and conservatively treated had recurrence, transformation or increase in size of lesion.

Omlor et al.⁹ studied outcome of conservative and surgical treatment of enchondromas and atypical cartilaginous tumors of the long bones: retrospective analysis of 228 patients. Comparison of all conservatively and surgically treated lesions showed excellent clinical results, however, the functional limitations were increased after surgical treatment. Surgically treated patients with histological diagnosis of enchondroma showed worse results for functional limitations, pain, and the MSTS score compared to conservatively treated patients. Sub - analysis of cases with larger tumor size (> 4.4 cm) displayed significantly worse clinical results with regard to pain and the MSTS score in the surgery group compared to the conservative cohort. For surgically treated patients, the complication rate has to be considered before carrying out treatment. Seven of the

75 surgically treated patients had complications requiring revision surgery. The revisions were necessary due to five peri - implant fractures and two intra - articular screws, which suggests lower complication risk in cases receiving the more conservative approach without osteosynthesis but otherwise non - inferior results – a finding, which was statistically significant. This underscores higher complication risks in cases treated with more sophisticated surgical strategies 28 - 30.

Case 32 A 48 year old female presented with pain over left knee for one month. Xray shows lytic lesion proximal fibula. MRI taken reported as enchondroma/low grade chondrosarcoma. Patient underwent proximal fibulectomy.



Figure 10: Pre Operative X ray Left knee AP view shows lytic lesion with specks of calcification over proximal fibula.



Figure 11: Post Operative X Ray after proximal fibulectomy and specimen

CASE 30 A 55 year old female presented with pain over right knee. X ray shows calcified enchondroma in distal femur which was managed conservatively.



Figure 12: X RAY right distal femur AP and lateral view showing calcified enchondroma

5. Conclusion

Based on the study conducted Enchondromas are most commonly diagnosed cartilage tumours. They are frequently associated with adjacent joint or soft tissue pathologies, which are main source of the symptoms. Some of the tumours are found incidentally during radiological evaluation of a nearby joint. Diagnosis work up consist of plane radiographs, CT – scan and MRI as imaging modalities. In plain x - rays enchondromas are lytic lesions with specs of calcification. CT and MRI help in additional confirmation regarding size and extent of the lesion and pre operative planning. PET scan is useful when the clinical, radiology, histology points towards a malignant nature of tumour. In our study we have followed up 48 patients with benign cartilage tumours of long bones attended orthopaedic oncology clinic. Most commonly patients presented with pain. Tumour was an incidental finding in 3 patients. Most common radiological finding was lytic lesion with specs of calcification. Distal femur was involved in 32 patients. Surgical intervention was done in 35 patients, while 13 patients were managed conservatively. In surgically treated patients 32 (91.43%) had post operative biopsy finding of enchondroma, in 3 (8.57%) patients report came as low - grade chondrosarcoma. In surgically treated patients 32 (91.43%) had final diagnosis of enchondroma, in 3 (8.57%) patients as Low - grade chondrosarcoma\ACT. In conservative management group 13 (100%) patients had enchondroma as final diagnosis. Patients managed

conservatively had symptom unrelated to tumour and lesion was found to be stable in subsequent follow - us. Both groups had 6 monthly follow - ups. None of the patients had local recurrence or new onset of symptoms related to tumour. In the small group of conservatively managed patients, it appears to be a safe option if strict follow up is assured and symptoms are unrelated to tumour. The indication for surgery should be discussed more thoroughly. If the symptoms are unrelated to tumour provided adequate evidence these benign cartilage tumours can be conservatively managed through clinical and radiological follow up. Thus, morbidity can be reduced in these patients and asymptomatic ones. Ultimately the decision for a surgical intervention depends on individual patient profile and thorough discussion with the patient regarding morbidity caused by surgery and about the chance of malignant transformation.

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