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**Dr. Gyneshwar Tank**  
LLRM Medical College  
Meerut, Meerut, Uttar  
Pradesh, India

**Dr. Sumit Agarwal**  
LLRM Medical College  
Meerut, Meerut, Uttar  
Pradesh, India

**Dr. Kalom Jamoh**  
Department of orthopedic,  
ESIC-PGIMS Basaidarapur,  
New Delhi, India

**Corresponding Author:**  
**Dr. Gyneshwar Tank**  
LLRM Medical College  
Meerut, Meerut, Uttar  
Pradesh, India

## **Osteochondroma transform into secondary low grade chondrosarcoma with similar histology features: Case report**

**Dr. Gyneshwar Tank, Dr. Sumit Agarwal and Dr. Kalom Jamoh**

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### **Abstract**

Osteochondroma is the most common form of benign bone tumors. Chondrosarcoma is a rare malignant bone tumor that may be primary or secondary. We report the case of a 19-year-old man with low grade chondrosarcoma secondary to malignant transformation of osteochondroma of the right distal femur. Here we discuss osteochondroma, chances of malignancy secondary to osteochondroma, chondrosarcoma, work up done in our case, histopathological features to differentiate osteochondroma from low grade chondrosarcoma, as well as treatment options that have shown significant benefits in patient's outcomes.

**Keywords:** Secondary chondrosarcoma; osteochondroma; transformation

### **Introduction**

Osteochondroma are benign chondrogenic lesion derived from aberrant cartilage from the perichondral ring that may take form of solitary osteochondroma or multiple hereditary exostosis usually present in the metaphysis of long bones and commonly present in the second and third decade of life<sup>1</sup>. The cartilage overgrowth is thick in children, then starts to thin out in adolescence, and by adulthood it usually measures less than 1cm<sup>2</sup>. Osteochondroma most often affects the long bones of extremity (around the knee), scapula, and the pelvis. They are the most common primary tumors of the axial skeleton and make up 35% of benign bone tumors overall<sup>3</sup>. Most cases of osteochondroma are painless mass, asymptomatic, and discovered as an incidental finding. If symptoms do present they present as limitation of motion and symptoms of neurovascular compression. Treatment of choice for osteochondroma is excision of the outgrowth or surgical removal of the solitary lesion<sup>2</sup>. The most serious complication of osteochondroma is transformation into malignant chondrosarcoma, which accounts for less than 1% of reported cases. Furthermore, 5%-10% of these patients are found to have hereditary multiple osteochondroma which is an autosomal dominant disorder characterized by two or more osteochondroma lesions<sup>4</sup>. This is a case report of solitary osteochondroma transformation into chondrosarcoma.

### **Case Presentation**

A 19-year-old male known case of osteochondroma of left distal femur presented to us with a painful mass on the inner aspect of left thigh distal third and difficulty in walking since the last 3 months. The mass has been present over the last 1 year and has been recently increasing in size and extremely painful. Physical examination showed a globular mass of size about 6cm x 6cm over the medial aspect left thigh distal third, tender, bony hard, well defined margin, irregular surfaced, not attached to skin and arising from the medial part of left distal femur and distal neurovascular are intact. A plain Radiograph was obtained and finding consist of bony outgrowth from medial condyle of left femur with popcorn shape calcification within the lesion with cortical breach at multiple site, suggestive of malignancy transformation so, MRI was obtained and its reveals ill-defined altered signal intensity lesion arising from metaphysis of lower femur with associated soft tissue component and extent anteriorly into vastus medialis muscle along the cortical margin and posteriorly infiltrating into Sartorius and semimembranosus muscle. Chest X-Ray shows no pulmonary metastasis. Based on the clinico-radiological finding, the patient was considered as a case of Chondrosarcoma secondary to osteochondroma of distal femur left side. Patient underwent a surgical excision of the tumor.

All soft tissue surgical margins were free of tumor the resected mass showed irregular 7.5 x 6 x 4.5 cm cartilaginous excrescence heavily calcified, that projected from the surface distal femur and extended into surrounding soft tissue. The mass was partially covered by a bluish cartilaginous cap (Thickness-4cm).

Microscopic examination revealed mature bony trabeculae covered by cartilaginous cap composed of mature hyaline cartilage, containing small chondrocytes arranged in columns with overlying thick and thin fibrous perichondrium without atypia cells. The tumor was considered to be a low-grade chondrosarcoma, grade I of III (Based on clinic-radiological features and thickness of cartilage capsule).

Patient received no postoperative chemotherapy or radiation and was considered for strict follow up in OPD.

## Discussion

In this report, we described a case of secondary chondrosarcoma arising from an osteochondroma on the distal femur of the left side. This unusual painful presentation of osteochondroma can be observed in <1% of patients [4]. Most solitary osteochondroma are found in children and adolescents with symptomatic lesions occurring in younger patients. They are typically asymptomatic and are discovered incidentally [5]. Clinical features of osteochondroma include a non-tender, painless, slowly growing mass [5]. Two types of osteochondroma include sessile type with a broad-based attachment to the cortex and pedunculated one with a long and thin stalk and bulbous tip as in our patient's.

The risk of malignancy in osteochondroma is generally low. With a follow up of osteochondroma, any alterations in radiological appearance, especially with ill-defined margin evolution and thickening of the cartilage greater than 15 mm, is highly suggestive of malignant transformation of osteochondroma to chondrosarcoma [6]. A major consideration in determining the malignant potential of an osteochondroma is the thickness of its cartilage cap; malignant transformation occurs with cartilage cap thicknesses greater than 2cm [8]. Many investigators have reported the incidence to be 0.5%-1% in osteochondroma and 5%-10% in patients who have hereditary multiple exostoses [4]. In the case of our patient, it was a solitary osteochondroma.

Hereditary multiple exostoses (HME) is an autosomal dominant disorder that is characterized by two or more exostoses in the axial and appendicular skeleton [7]. It is diagnosed by the presence of two osteochondroma that are detected by radiograph in the metaphyseal ends of the long bones [12]. It presents similarly to those with solitary osteochondroma during the second decade. Males are more commonly affected than females [10]. Most individuals with HME have a parent with the condition, however spontaneous mutations can be found in 10-20% of individuals with HME [13].

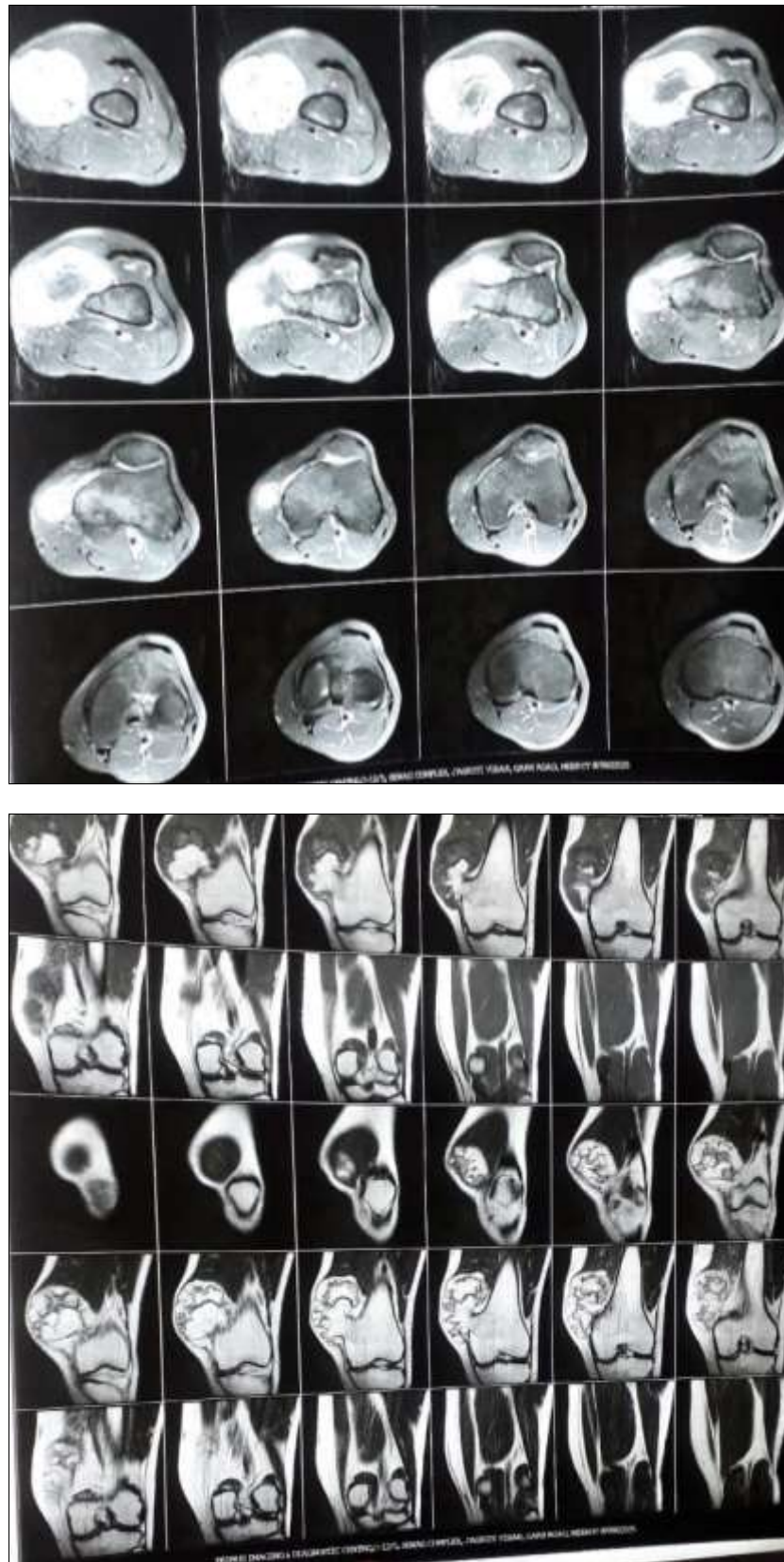
Secondary chondrosarcoma are rare and often difficult to diagnose due to the slow growth and late recurrence. Long-term follow up care is standard. One study found that most patients who died of chondrosarcoma, died due to local recurrence of the tumor [14]. Less than 5% of the patients in the study developed metastases, majority of which were found in the lung [14]. Other organ systems affected include nerve or vascular injury, bursa formation and configuration of a pseudo aneurysm [15].



**Fig 1:** Globular mass of size about 6cm x 6cm over the medial aspect left thigh distal third



**Fig 2:** Bony outgrowth from medial condyle of left femur with popcorn shape calcification within the lesion with cortical breach at multiple site

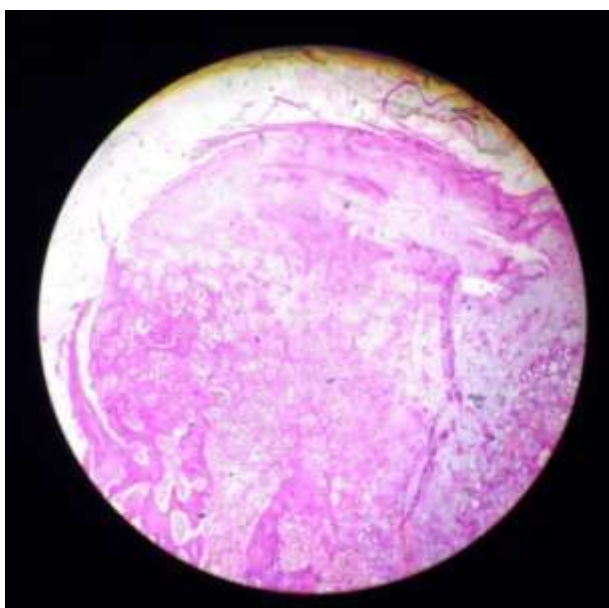


**Fig 3:** MRI- ill-defined altered signal intensity lesion arising from metaphysis of lower femur with associated soft tissue component and extent anteriorly into vastus medialis muscle along the cortical margin and posteriorly infiltrating into Sartorius and semimembranosus muscle





**Fig 4:** Gross examination- showing bony mass with outer surface white, encapsulated with cartilage cap (thickness 4cm)



**Fig 5:** Microscopic examination- mature bony trabeculae covered by cartilaginous cap composed of mature hyaline cartilage, containing small chondrocytes arrange in columns with overlying thick and thin fibrous perichondrium without atypia cells

Chondrosarcoma is a heterogeneous type of primary bone cartilage malignancies with highly contrasting clinical outcomes. Although recurrent mutations in the IDH genes and other genetic alterations including inactivation of CDKN2A and COL2A1 are commonly found in these tumors<sup>16</sup>, molecular testing of chondrosarcoma is usually not indicated due to lack of significant clinical value. An interesting recent study by Rémy Nicolle *et al.*, used multi-omics molecular profiles from a series of 102 cartilage tumors and found an mRNA classification that identifies two subtypes of chondrosarcoma defined by a balance in tumor differentiation and cell cycle activation. The microRNA classification revealed the importance of the loss of expression of the 14q32 locus in defining the level of malignancy. They also found that DNA methylation is associated with IDH mutations, and the use of multi-omics classifications may be able to predict outcome. Based on

their findings, they proposed an mRNA-only classifier to reproduce the integrated multi-omics classification, and its application to relapsed tumor samples showed the progressive nature of the classification. Thus, it may be possible to use mRNA-based signatures to detect patients with high-risk chondrosarcoma<sup>[16]</sup>. Heinritz W *et al.*<sup>[17]</sup>, reported new mutations of EXT1 and EXT2 genes in German patients with Multiple Osteochondromas<sup>17</sup>. No molecular studies on transformed cases were published yet. The most effective current treatment option reported for osteochondroma and chondrosarcoma is surgical removal of the exostoses mass, with minimal involvement of the musculature. As reported by Ngogang *et al.*<sup>[18]</sup>, procedures involving no muscle detachment resulted in reduced blood loss and decreased recovery time, which could be decreased further with the use of endoscopy techniques. We believe that the low- grade chondrosarcoma and the complete excision of the mass with safe margins will contribute to the favorable outcome.

The lesion we learned from this case is that

1. We should regularly follow up the patient with osteochondroma and remove a benign tumor as early as possible in order to avoid the risk of development of secondary chondrosarcoma may be wise because malignancy has a mortality rate of about 5%.
2. The histological features of low grade Chondrosarcoma may be similar to that of Osteochondroma as in our case, also reported by Dai Robert *et al.*<sup>[18]</sup>.

So differentiate between osteochondroma from the low grade conventional chondrosarcoma is based on (a) clinical presentation (pain, rapid increase in size), (b) radiological finding (size of tumor >5cm, cortical breach & soft tissue involvement) and (c) gross morphology of the tumor (cartilage cape >2cm) suggestive of chondrosarcoma.

It is our hope that this report raises awareness to clinicians and pathologists to this possible transformation of osteochondroma to low grade chondrosarcoma with similar histological findings and that continued investigation drives further development of efficacious diagnosis and safe treatments for improving patient outcomes.

## References

1. Vikram V Kadu, Saindane KA, Ninad Gogate, Neha Gogate. Osteochondroma of the Rib: A rare radiological appearance. J Orthop Case Rep. 2015;5:62-64.
2. Tessa AH Wilpshaar, Judith VMG Bovee. Atlas Genet Cytogenet Oncol Haematol. 23(5):133-136. Biermann J. Sybil Section Editor(s): Stanitski, Carl L. Editor Journal of pediatric orthopaedics. 2002;22:268-273.
3. Tianjun Lan, Xin Liu, Pei-Sheng Liang, Qian Tao. Osteochondroma of the coronoid process: A case report and review of the literature. Oncol Lett. 2019;18:2270-2277.
4. Cho HS, Han I, Kim HS. Secondary Chondrosarcoma from an Osteochondroma of the Proximal Tibia Involving the Fibula. Clin Orthop Surg. 2017;9:249-254.
5. Javdan M, Hekmatnia A, Ghazavi A, Basiratnia R, Mehrzad M, Hekmatnia F, *et al.* Case report of osteochondroma with unusual clinical and imaging presentation. Adv Biomed Res. 2015;4:2.

6. Murphey MD, Choi JJ, Kransdorf MJ, Flemming DJ, Gannon FH. Imaging of osteochondroma: Variants and complications with radiologic-pathologic correlation. *Radio-graphics*. 2000;20:1407-1434.
7. Shah ZK, Peh WC, Wong Y, Shek TW, Davies AM. Sarcomatous transformation in diaphyseal aclasis. *Australas Radiol*. 2007;51:110-119.
8. Gordon SL, Buchanan JR, Ladda RL. Hereditary multiple exostoses: report of a kindred. *J Med Genet*. 1981;18:428-430.
9. Lichtenstein L, Jaffe HL. Chondrosarcoma of bone. *Am J Pathol*. 1943;19:553-589.
10. Spjut HJ. Tumors of bone and cartilage. In: Spjut HJ, Dorfman HD, Fechner FE, Ackerman LV, eds. *Atlas of tumor pathology*. 2nd series Washington, DC: Armed Forces Institute of Pathology, 1971, 84-110.
11. Bredella MA, Stoller DW, Johnston JO. Bone and soft-tissue tumors. In: Stoller DW, ed. *Magnetic resonance imaging in orthopaedics and sports medicine*. 3rd ed. Philadelphia, Pa: Lippincott Williams & Wilkins, 2006; 2065-2070.
12. Vlok SCS, Wagener GWW, Zaharie D. Secondary chondrosarcoma: Malignant transformation of pre-existing hereditary and non-hereditary cartilaginous lesions. *S Afr J Rad*, 2014, 18.
13. Schmale GA, Conrad EU, Raskind WH. The natural history of hereditary multiple exostoses. *J Bone Joint Surg Am*. 1994;76:986-992.
14. Ahmed AR, Tan TS, Unni KK, Collins MS, Wenger DE, Sim FH. Secondary chondrosarcoma in osteochondroma: report of 107 patients. *Clin Orthop Relat Res*. 2003;411:193-206.
15. Recht MP, Sachs PB, LiPuma J, Clampitt M. Popliteal artery pseudoaneurysm in a patient with hereditary multiple exostoses: MRI and MRA diagnosis. *J Comput Assist Tomogr*. 1993;17:300-302.
16. Nicolle R, Ayadi M, Gomez-Bouchet A, Armenoult L, Banneau G, Elarouci N, *et al*. Integrated molecular characterization of chondrosarcoma reveals critical determinants of disease progression. *Nat Commun*. 2019;10:4622.
17. Heinritz W, Hüffmeier U, Strenge S, Mitterski B, Zweier C, Leinung S, *et al*. New mutations of EXT1 and EXT2 genes in German patients with Multiple Osteochondromas. *Ann Hum Genet*. 2009;73:283-291.
18. Ngongang FO, Fodjeu G, Fon AC, Fonkoue L, Guifo ML, Bitang A Mafok LJ, *et al*. Surgical treatment of rare case of scapula osteochondroma in a resource limited setting: A case report. *Int J Surg Case Rep*. 2019;61:130-134.