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Secondary chondrosarcoma in metachondromatosis: a case report

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Introduction: Metachondromatosis is a rare, autosomal dominant, incompletely penetrant combined exostosis and enchondromatosis tumor syndrome. We present a patient with metachondromatosis in whom a chondrosarcoma was diagnosed at the site of an enchondroma. This is the only first documented case of secondary malignant transformation of a metachondromatosis enchondromatous lesion to chondrosarcoma.
Methods: A 29-year-old woman presented with pain localized at her right knee over 1 year. Radiography showed skeletal osteochondromas and enchondromas of the right femur and tibia, and an osteolytic lesion at the lateral tibial condyle. CT scan showed a cystic osteolytic lesion at the right proximal tibial metaphysis with erosion of the anterior and lateral cortex, and an adjacent proximal tibial enchondroma. MRI showed a destructive lesion, with erosion of the anterior and lateral cortex, without an associated soft tissue mass. Bone scan showed increased radioisotope uptake in the right proximal tibia and at the region of the right proximal femur, probably in the lesser trochanter. Based on the synchronous occurrence of skeletal osteochondromas and enchondromas, the diagnosis of metachondromatosis was established.
Results: CT-guided biopsy of the lesion was done; histology showed grade 1 chondrosarcoma. Complete curettage of the lesion using a high-speed burr, and application of phenol and bone cement as bone void filler was done; histology showed evidence of grade 2 chondrosarcoma. The patient was informed regarding immediate reoperation, and agreed to close follow-up evaluation using MRI; in case of local recurrence, wide surgical excision will be performed. Five years postoperatively, the patient is alive with no evidence of local recurrence or distant metastases.
Conclusion: We acknowledge that the treatment of grade 2 chondrosarcoma with intralesional surgery is inappropriate. However, the purpose of this study is not to present the treatment and prognosis of primary or secondary chondrosarcomas but to emphasize on metachondromatosis and present the malignant transformation of a metachondromatosis associated enchondroma to chondrosarcoma.