A singular enchondroma presenting as a pathologic humerus fracture in a patient with multiple osteochondromas. A case report.

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Introduction and Objectives:
Hereditary multiple osteochondromas (hereditary multiple exostoses, diaphyseal aclasis) is a rare autosomal dominant disorder characterized by the formation of multiple osteochondromas, mostly in the long tubular bones (humerus, radius, ulna, femur, tibia, fibula), and the development of associated osseous deformities. The disease is typically diagnosed during childhood and requires a lifelong monitoring, mostly because of the risk of the malignant transformation into a secondary chondrosarcoma. Surgical treatment is sometimes needed in painful osteochondromas, in cases of local mechanical irritation of the soft tissue, or in cases of vascular compromise or nerve compression.

Benign bone tumors producing hyaline cartilage can occur intramedullary, either as a single lesion (enchondroma), or as multiple lesions (enchondromatosis). Ollier disease and Maffucci syndrome are the most common subtypes of enchondromatosis while the others like metachondromatosis are much less frequent. Ollier disease is a non-hereditary developmental disorder characterized by the occurrence of multiple cartilaginous masses, particularly affecting the long tubular bones of the limbs. When haemangiomas are also present, the disorder is referred to as Maffucci syndrome. Metachondromatosis is an extremely rare hereditary disorder involving the formation of both enchondromas and osteochondromas. It is distinct from hereditary multiple osteochondromas as the orientation of lesions in metachondromatosis is towards rather than away from the epiphysis, and there is a predilection for the hands and feet. There have been only approximately 50 cases reported worldwide.

We report the case of a 40 year old patient with multiple osteochondromas and a single enchondroma of the humeral proximal metadiaphysis.

Case presentation:
A 40 year-old patient with known multiple osteochondromas was referred to our hospital with pain in the left humerus after falling on the left arm. X-rays and contrast enhanced MRI of the left humerus showed a pathological fracture of the left proximal humeral metadiaphysis through the bizarre deformity due to known osteochondromas. In the area of the fracture, an intramedullary lesion was observed, with enchondroma-like typical popcorn calcifications. A biopsy was performed, and the histological examination revealed features of an enchondroma without evidence of malignancy. The treatment included curettage of the enchondroma, subsequent filling of the defect with beta-tricalcium phosphate bone graft substitute chronOS®, and Philos plate osteosynthesis. The histological examination of the curettage material confirmed the diagnosis of an enchondroma.

Discussion/Conclusion:
This case highlights the importance of clinical and even more radiological examination, and also raise awareness of the possibility of a patient having more than one tumor in the same location. Although radiological presentation of the enchondroma could be typical, bizarre bone deformity in patients with multiple enchondromas could linger radiological analysis. The treatment in the patients with two different, still benign lesions, should not differ the standard osteochondroma/enchondroma treatment, even in the cases of the pathologic fracture. Regular, lifelong check-ups are recommended.

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Keywords : Multiple Osteochondroma, Enchondroma, pathologic fracture
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