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Commentary

A Brief Description on Multiple Osteochondromas Disease

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DESCRIPTION

Multiple Osteochondromas (MO) is a dominant skeletal disease, in this disease there is noncancerous bone tumors are developed these tumors are called osteochondromas. The estimated possible rate of this disease is 1:50,000 and it is higher in male. This disease is generally occurring to long tubular bones or at the surface of the flat bones in the body at in the time of first decade of the life and develops slowly. This osteochondromas is caused due to increase in chondrocyte proliferation, this is results to excessive bone growth in children's metaphysis. Generally these tumors can arise from each bone with a cartilage origin, but the possible percentages of occur are at distal femur is 90%, and at the proximal tibia is 84%, fibula is 76% and at humerus is 72% and the facial bones are not affected and the mean number of locations in the body is in between 15-18. This osteochondromas bone tumors are benign, but they can cause to several secondary complications, they are compression of blood vessels, nerves and tendons. The inequality in limb length, varus angulation of the knee and disproportionate short stature. The angular deformation of ankle or forearm is the most complicated orthopedic problem. The most serious complication in multiple osteochondromas is the malignant transformation of an osteochondroma toward a secondary peripheral chondrosarcoma, this is generally occur only in 1%-5% of patients and this malignant degeneration is increases with age. Most of the people prefer surgery to remove this osteochondroma for their functional and cosmetic reasons. Most of the patients with MO experience pain and half of are under estimate this pain and remaining seems to a problem, more pain tends to surgery and treatement.

The cause for this disease is mutation in protein coding gene Exostosin Glycosyltransferase 1 (EXT1) and Exostosin Glycosyltransferase 2 (EXT2). These genes are providing instructions to produce proteins of exostosin-1 and exostosin-2. These proteins function is to produce heparan sulfate protein, but there is a mutation of exostosin-1 and exostosin-2 is caused to nonfunctionals in the body, this is causes to osteochondromas disease. The members of the EXT family are characterized by their homology in the carboxyterminal region, a ubiquitous expression pattern and the presence of a signal peptide at the amino terminus.

The diagnosis process for this disease is carefully monitored because of its rarity. The patient's family history is carefully reviewed before proceeding to differential diagnosis. And specific histological and radiological expertises in the field are needed to diagnosis. The patient's blood is screened for germline mutations in the protein genes EXT1 or EXT2. The differential diagnosis methods for solitary and hereditary osteochondromas are Dysplasia Epiphysealis Hemimelica (DEH) is also known as Trevor's disease and Metachondromatosis (MC). The simple removal of n osteochondroma can improve forearm rotation and correct deformity, especially if there is an isolated tumour of the distal part of the ulna. It is more important that there is no new osteochondromas after the treatement. And it is important to regular follow-up to discover potential malignant transformation at an early stage to enable adequate treatment should be considered.

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