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Chondrosarcoma of hand and feet – Review of 37 cases from single institution

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Introduction: Solitary enchondromas (ECH) first described by Lichtenstein and Jaffe in 1943 are rather frequent bone hamartomas. They are believed to be the most common benign tumor of the hand, however their true incidence is unknown as they are mostly asymptomatic. M.Altay et al. reports rate of malignization 4,2% at average age of 49,8 years (7,7years from the diagnoses). Despite the commonness of ECH, chondrosarcoma (ChSA) of hand and accounts for less than 2% of all ChSA of the body and is believed to have an indolent course. Nevertheless the biggest yet published series of 111 patients from Mayo Clinic found chondrosarcoma of small bones to have the potential to be fatal. At present still neither radiotherapy nor chemotherapy are of practical value. Therefore adequate surgery is utter most imporant in achieving a cure. Nonetheless no consensusMatherial and methods:We reviewed charts of all patients operated in our clinic form 1965 to present day and found total of 45 patients treated for acral CHSa. Tumors originated in distal antebrachii or crus with extension to periphery, as well as soft tissue CHSa, were not included. We also excluded patients with mesenchymal, dedifferentiated and clear cell chondrosarcomas were not included as they have a different biological behaviour and only patients with diagnosis of convenctional bone CHSa were included in the study. Patients were evaluated for demografic data, duration and type of presenting symptom, localization, type of surgery, local recurrence and metastatic disease. 37 patients with follow-up of minimum 2 years, who met all inclusion criteria were included in the study.
Results: Of these patients 25 had tumor in hand and 12 bellow ankle. Men and female were equaly affected. Mean age at the time of diagnosis was 50,1 years (6y to 87y) with peak incidence in 7th decade. The most common presenting symptom was a bulge enlarging for relatively long time (40,7 months in average) before the patients seeked medical attention. Pain was a rare symptom and few cases manifested via pathological fracture (n=5). CHSa most often originated in metacarpal (metatarsal) region followed by proximal phalanges, with 3rd and 5th ray of the hand beeing the most commonly affected. Tumors in the foot were more often higher grade, when compared to those in hand. The great majority of CHSa in our series were secondary. The most frequent underlying condition was enchondroma (in 5 cases associated with Olliere disease and in 1 case with Maffucci sy.), followed by osteochondroma. In 5 cases no previous laesion was indentified. The treatment was always surgical. Of all the patients 4 (10,8%) developed distant pumonary metastases that were fatal for two of them, one patient died of unrelated causes and one is alive with disease. Patients with GI CHSa tend to be younger (40,6y vs 58,2y) and except for one all have been primarily treated by intralaesional surgery. Nine of these patients (75%) suffered local recurrence at an average of 131 months after the primary surgery. In all cases of multiple LR we encountered propensity to reccur in a higher grade then previous tumor (two of these patients susequently developed distant pulmonary metastases). On the other hand majority of patients with GII-III CHSa had an wide resection as a primary treatment. Four of them had an intralaesional surgery and all suffered early LR at an average of 5,75 months.
Conclusion: Despite the rarity of peripheral CHSa the consequences of these tumors are significant and they deserve a highly specialized treatment, espetialy if dominant extremity is affected. However their behavior is not as benign as proposed by Bovee and successful treatment requires careful planning. Despite high rate of recurrence after intralaesional surgery, we still would reccomend this as treatment of choice for GI laesions as it is difficult to distinguish them from benign ECH and this approach gives best functional results. However in case of a LR, more aggressive approach should be taken in consideration as the risk of developing a HG laesion raises with every reccurrence. Despite believed benign course of peripheral CHSa, fatal metastatic disease can develop in GIII laesions. The fact that feet were more likely to harbor a HG CHSa can be atributed to the the fact, that in this location they can go longer unnoticed. The peak incidence correlates with average life expectancy in Czech Republic and it is likely, that if this is to increase, the increase in incidence of peripheral CHSa will follow. Prefably those of higher grades.