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Results and prognostic factors in 15 patients with peripheral dedifferentiated chondrosarcoma

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Background: Dedifferentiated chondrosarcoma is an uncommon tumor that is known to arise from pre-existing, low-grade cartilage lesions. Peripheral dedifferentiated chondrosarcomas (PDC) arise from pre-existing exostoses, or extracortically, and may appear as a peripheral chondrosarcoma without the features of its dedifferentiated counterpart. Dedifferentiated chondrosarcoma has a very poor prognosis. Aim of this study was to evaluate the survival of patients with peripheral PDC and to evaluate possible prognostic factors.

Methods: Between 1980 and 2006, 15 patients were treated for PDC: 11 males and 4 females, mean age of 42 years. In 1 case tumor was located in the humerus, in 3 in distal femur, in 1 in emi-anterior chest, in 5 cases in ileums, in 2 in scapula, in 2 proximal femur, in 1 proximal fibula.

The dedifferentiation was in malignant fibrous histiocytoma in 9 cases, osteosarcoma in 5 cases and spindle cell sarcoma in 1 cases. 14 patients received surgery (one patient was not operable for multiple distant metastases): tumor resection in 9 cases, amputation in 5. Chemotherapy was given to 8 patients.

Results: 4 patients (26.6%) were Ned at a mean followup of 14.7 yrs and 11 patients DWD at a mean time of 2.6 yrs. The overall survival of patients was 34% at 10 years. There was not significant difference in survival between patients with D.C. of the trunk and those with D.C. of the extremities ($p = 0.2397$).

There was no significant difference in survival with chemotherapy and surgery or with surgery only ($p = 0.6269$).

Conclusion: The prognosis for patients with D.C. remains dismal. Surgery with wide margins remains the principal treatment for this condition. There was no statistical evidence of any beneficial effect from chemotherapy.

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