FC-107

Extra-abdominal desmoid tumor: prognostic factors and clinical outcome in a surgical series

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Purpose: Extra-abdominal desmoid-type fibromatosis is an extremely rare disease and presents unpredictable and enigmatic nature. Optimal management has not been established ever, and different consideration may be needed discriminating from intra-abdominal and abdominal types. We investigated the clinical characteristics, clinical outcome, and prognostic factors on recurrence in patients surgically treated.
Methods: We retrospectively reviewed the medical records on 133 lesions in 89 patients who underwent surgical excision of extra-abdominal desmoid-type fibromatosis at three tertiary institutions from 1990 to 2013. Patients with R2 margin were excluded. Gender, age, size, multifocality, site, surgical margin, and adjuvant treatments were regarded as potential factors on recurrence-free survival and statistically analyzed.
Results: Median Follow-up was 66 months in all patients and 27 months in those with recurrence. Forty-seven lesions in 29 patients recurred, 28 lesions of which were primary lesions. Twelve lesions recurred despite negative surgical margin. No patients reported family history or Gardner's syndrome. Trunk lesion showed no recurrence. Eleven lesions recurred over 2 times. Median age in patients with primary lesion was significantly higher than in those with recurrent lesion. Median size was larger in patients with recurrence over 2 times than in those without relapse. Age and site were documented as independent prognostic factors in all tumors, all primary tumors, girdle and extremity tumors, and girdle and extremity primary tumors. Any independent prognostic factor was not identified in all recurrent tumors, and girdle and extremity recurrent tumors.
Conclusion: Age may be correlated to proliferative activity of the disease. Tumor site must be closely related with surgical margin. If it is impossible to achieve R0 margin, the key on management for desmoid-type fibromatosis may be assessment for biological activity rather than treatment selection itself. Functional imaging modalities could be of help to predict the activity, and multicenter collaboration must be mandatory for future study.