

## **Extra-abdominal Desmoid Tumor: Prognostic Factors and Clinical Outcome in a Surgical Series**

**Min Wook Joo, Yong-Koo Kang, Yang-Guk Chung, Won-Jong Bahk, Seung Han Shin, Jin-Woo Kang**

Department of Orthopaedic Surgery, College of Medicine, The Catholic University of Korea, Banpo-daero 222, Seocho-gu, Seoul 137-701, Republic of Korea

### **Background**

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 required discriminating from intra-abdominal and abdominal types.

### **Questions/Purposes**

We investigated the clinical characteristics and prognostic factors on recurrence in patients surgically treated.

### **Patients and Methods**

We retrospectively reviewed the medical records on patients who underwent surgical excision of extra-abdominal desmoid-type fibromatosis at three 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**

There were a total of 133 lesions in 89 patients, 70 lesions in females and 63 in males, with a median age of 35 years at diagnosis. Median Follow-up was 66 months in all patients and 27 in those with recurrence. Extremity lesions were seen in 81. Forty-seven lesions in 29 patients recurred, of which 28 were primary lesions. Eleven lesions recurred over 2 times. No patients reported family history or Gardner's syndrome. 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. Twelve trunk lesions showed no recurrence. 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. Univariate analyses showed site as a prognostic factor in all recurrent tumors, and girdle and extremity recurrent tumors.

### **Conclusions**

Age may be correlated to proliferative activity of the disease. Tumor site must be closely related with quality of surgery. Disease status and multifocality were not important factors in this study. The key on management for desmoid-type fibromatosis may be assessment for biological activity and decision of intervention timing rather than treatment selection itself. Multicenter collaboration must be mandatory for this extremely rare disease.