

**PP-143****Aggressive fibromatosis in children – Single institution experience****V. Ilic**, Z. Bekic, J. Bokun, M. Pudrlja Slovic, I. Bekic, L. Paripovic*Institute for Oncology and Radiology of Serbia, Belgrade, Serbia*

Introduction: Aggressive fibromatosis (AF) is a rare tumor of intermediate malignancy that has the strong potential for local invasiveness and recurrence.

Methods: The objective of the study was to present the treatment modalities in children who had extra-abdominal AF treated over an 11 year period at the Pediatric Department of the Institute for Oncology and Radiology of Serbia.

Results: The patients were drawn from a period of time between 2004 and 2014. There were 9 children (7 male and 2 female) with a median age of 12,5 years (age range from 10 to 14 years). The primary tumor sites in patients were found as follows: extremities (7 patients) and head and neck region (2 patients).

All the patients studied had surgery as the first line of treatment. Only one patient had microscopically negative margins and received no further treatment. Five out of eight patients with marginal surgery received chemotherapy and radiotherapy along with surgery as the primary treatment. Chemotherapy was administered to patients who had surgery with positive margins and macroscopically incomplete surgery and those with recurrent disease. Regimens administered were VACA, VAIA, VAC, IVA and low dose MTX/VBL. Two patients with tumor progression received second line therapy followed by a second surgery. One patient with macroscopically incomplete surgery was on watchful-waiting strategy and had a 6 year follow up that showed stable disease. One patient did not respond to any of the administered lines of chemotherapy and underwent amputation. One patient with localized head and neck tumor who underwent 5 marginal surgical interventions received radiotherapy and low dose MTX/VBL and is currently still in a 6 year follow-up. Median follow-up was 3 years.

Conclusion: Adequate surgical procedure with negative margins is a successful primary treatment modality for children with AF. The aim of chemotherapy is tumor reduction that can permit surgical resection. Mutilating surgery and highly toxic chemotherapy regimens should be avoided. Positive margins after surgery indicates high risk for local recurrence but local recurrence didn't affect patients' chance to respond to chemotherapy.