Aggressive fibromatosis is a benign, locally aggressive disease that has been shown to have an unacceptably high local recurrence rate after surgical resection alone. Definitive management of aggressive fibromatosis is controversial, with many advocating for surgical resection. However, radiation therapy (RT) has been shown to improve local control and decrease the risk for recurrence when compared to surgical resection. The literature suggests that radiation alone, or a combination of radiation with surgical resection provides the best treatment option to achieve local control in this disease.

The purpose of our study was to determine outcomes with RT in the treatment of extra-abdominal aggressive fibromatosis at our institution in patients treated with radiation as the primary treatment modality, or those treated after failure of surgical management with subsequent RT. Functional outcomes were also assessed in this same patient population.

Subjects were retrospectively identified from our institution’s Orthopaedic Oncology Database as well as our Radiation Oncology Database, RADTRAC: Radiation Oncology Outcomes Tracking Project. Inclusion criteria: patients 18 and older who were diagnosed with aggressive fibromatosis and treated with RT as their first line of treatment or for recurrence after surgical resection. Exclusion criteria: patients under 18, patients with abdominal wall or intra-abdominal/intra-pelvic tumors, and those treated with radiation as part of a planned combined treatment with surgery. One hundred fifteen patients were identified who had undergone radiation therapy in their treatment of aggressive fibromatosis from 1975 to the present. Twenty nine patients fit the inclusion criteria with an average follow up of 8 years. A chart review was performed to determine success of treatment. A questionnaire was mailed to these patients to determine if they have ongoing clinical symptoms or if they have received further treatment elsewhere. This included questions from the modified Musculoskeletal Tumor Society Functional Assessment. Twelve patients were able to be contacted for follow up. Three patients were deceased prior to our follow up and 14 patients were otherwise lost to follow up. For those patients who were deceased or lost to follow up, results from their most recent clinical evaluation were used.

Eighteen patients were treated initially with RT. Average age was 41 at the time of treatment. Radiation dosage was available for 13 of the patients with an average of 52.2 Gy per patient. The majority of the tumors were located in the shoulder girdle and chest wall. Of the 15 patients successfully treated with radiation alone, the average MSTS score at the time of follow up was 92.1%. Three patients developed recurrence at an average of 26 months and went on to surgery. Only one was felt to have had successful treatment. MSTS score for one of the remaining 2 patients was 25%.

Ten patients were treated with surgical resection, recurred and were treated definitively with RT. They averaged 45 years old at the time of radiation treatment. They had undergone an average of 2.2 surgeries (1-6). Average
radiation dosage was 52.8 Gy per patient. Tumors were distributed equally between upper and lower extremities. Nine of the patients had successful treatment with radiation. Follow up MSTS score was available for two of these patients with an average of 90%. There was only one recurrence after RT. She was placed on Tamoxifen but continued to have slow growth of her tumor noted at her last follow up.

One patient failed a series of medical treatments before successful treatment with definitive radiation. Satisfaction with final outcome was rated as high with an MSTS score of 83.3%.

Conclusion:

Our data showed a success rate of 83.3% after initial treatment with RT which is in line with data in the literature. We also had successful treatment of patients who received RT after failed surgical management with a treatment success rate of 90%. MSTS scores were high for patients successfully treated with RT. Our data is limited by the number of patients in our treatment groups as well as the retrospective nature of the study. True statistical relevance is unable to be calculated. However, the data does support RT as a first line therapy for patients and may be an improvement over surgical resection when compared to historical control rates with surgery. A prospective randomized trial would be of benefit in further clarifying treatment approach.