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Long-term outcomes of 47 patients with aggressive fibromatosis of the chest treated with surgery

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Abstract

Aims: The purpose of the study was to review a large series of chest aggressive fibromatosis (AF) cases with an emphasis on the outcomes of different degrees of resection and the value of postoperative radiotherapy.

Materials and methods: The records of patients with chest AF treated at our hospital from 1982 to 2014 were retrospectively reviewed. Recurrence rates and non-disease survival (NDS) times were compared between the R0, R1, and R2 resection groups.

Results: Forty-seven cases of chest AF were treated during the study period (21 men, 26 women), with an average age at diagnosis of 40 years (range, 9–77 years). One patient died before surgery, and 46 patients received a total of 85 resections. Forty-one patients had complete follow-up data, and the average follow-up time was 125.6 months (range, 11–524 months). Recurrence rates were 6.7%, 92.9%, and 100% for the R0, R1, and R2 resection groups, respectively, and the R0 recurrence rate was significantly lower than the R1 and R2 rates (both P values < 0.001). The NDS time of the R0, R1, and R2 groups was 80.3 ± 64.8, 23.6 ± 38.7, and 9.8 ± 10.8 months, respectively; the NDS time of the R0 group was significantly longer than that of the R1 and R2 groups (both P values < 0.01). Within each resection type, no significant differences were found in the recurrence rates of patients having surgery alone compared with those receiving surgery and radiotherapy (all P values > 0.05).

Conclusion: R0 resection is the most effective treatment for chest AF. Postoperative radiotherapy did not reduce the recurrence rate.

Keywords: Aggressive fibromatosis; Desmoid; Reconstruction; Chest wall defect

Introduction

Aggressive fibromatosis (AF), also known as desmoid tumor or desmoid-type fibromatosis, was defined by the World Health Organization (WHO) in 2002 as a cloning fibroblast proliferative tumor occurring in deep tendon membrane tissues.[1] AF is characterized by abundant collagen fibers with involvement of the skeletal muscle aponeurosis and fascia; it has a tendency towards progressive infiltration of surrounding tissues and local recurrence without the ability to metastasize.[1] Its biological behavior is classified between that of fibroblastoma and fibrosarcoma.[1] AF accounts for 3% of soft tissue tumors, and 0.03% of all malignant tumors; in the United States, the incidence is 2–4 cases/100,000 persons yearly.[2] The disease occurs primarily in younger age groups, and there is no specific gender difference. Approximately 5% of tumors are multifocal, and the mortality is about 8%.[2]

Surgery is the primary therapy for resectable cases of chest AF, and after surgery alone, local recurrence rates range between 20% and 80%.[2,3] Traditionally, the goal of surgery has been R0 resection; however, this is difficult in many cases of chest AF.[4,5] Some authors have suggested less aggressive surgery and postoperative radiotherapy to improve the local control rate; however, the group of patients for whom this is suitable therapy remains a matter of debate.[4,5] Other therapies, including irradiation, hormonal therapy, chemotherapy, and receptor tyrosine kinase

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inhibition, can be successful in stabilizing growth or shrinking lesions, and although no single therapy is effective in all cases, different therapies may be of value in certain pathological subgroups.[6]

AF is a rare disease for which there is a lack of prospective randomized studies; additionally, there are no standardized treatment guidelines,[7,8] and the literature regarding chest AF is mostly case reports. Thus, the purpose of this study is to report a large series of chest AF cases treated at our institution with an emphasis on the outcomes of different degrees of resection and the value of postoperative radiotherapy.

Materials and methods

In this study, the records of patients diagnosed with chest AF and treated at Peking Union Medical College Hospital from December 1982 to May 2014 were retrospectively reviewed. This study was approved by the Institutional Review Board of the hospital, and because of the retrospective nature of the analysis, the requirement for the patients' informed consent was waived.

Data extracted from the medical records included patient age, gender, clinical history, surgical procedure, extent of resection, histopathological and immunohistochemical results, the use of radiotherapy, and outcome data. Patients were followed up every 6 months for the first 5 years after surgery and yearly thereafter. Postoperative follow-up consisted of outpatient examinations and annual ultrasound or magnetic resonance imaging (MRI) examinations.

All pathological specimens were fixed in 10% neutral-buffered formalin, embedded in paraffin wax, stained with hematoxylin and eosin (H&E), and examined under a fluorescence microscopy (Olympus BX51, Japan). For cases in which a diagnosis of AF was unclear, immunohistochemical staining was performed to distinguish a variety of tumors, such as smooth muscle tumors, neurogenic tumors, synovial sarcoma, fibrosarcoma, mesothelioma, solitary fibrous tumors, nodular fasciitis, malignant fibrous histiocytoma, and gastrointestinal stromal tumors.

Statistical analysis

Continuous data were reported as mean ± standard deviation (SD) or range, while categorical data were presented as number and percentage. Three surgical groups were examined and compared: R0, R1, and R2 resection groups. One-way ANOVA with Tukey's honest significant difference post-hoc test was used to compare continuous means when there were more than two groups, while a t-test was used for two-group comparisons. Chi-square tests and Fisher's exact tests, as appropriate, were used for comparisons of categorical data. Kaplan–Meier survival analysis and log-rank tests were used to compare the recurrence rate and non-disease survival time (NDS; time from surgery to first relapse) among the three resection groups. Statistical analyses were done with SPSS, version 18, with an alpha level of 0.05.

Results

Patient clinical data and treatment

A total of 47 cases of chest AF were included in the analysis, and the clinical features are summarized in Table 1. These 47 cases accounted for 4.7% of chest soft tissue tumors and 18.7% of all AF cases diagnosed at our center during the study period. Twenty-six patients were diagnosed at our hospital, while 21 were relapses that were initially diagnosed and treated at other hospitals. There were 21 men (44.7%) and 26 women (55.3%), and the average age at diagnosis was 40 years (range, 9–77 years). There were 4 patients (8.5%) with multiple lesions, 2 of whom were females with familial adenomatous polyposis, and the other patients had single lesions.

A 77-year-old woman with giant chest-abdominal AF died before surgery due to respiratory failure. The other 46 patients received a total of 85 chest tumor resections: 28 R0, 24 R1, and 33 R2. There were 23 patients who had their initial surgery at our hospital; 14 of them had R0 resections. Twenty-five patients received single resections, and 21 required multiple resections. Artificial materials were used intraoperatively to repair the chest defects

Table 1
Clinical features of chest aggressive fibromatosis.

Presenting symptoms	Pain and numbness gradually developed in over half of the patients Upper limb swelling and numbness if the subclavian axillary region was involved, and craniofacial swelling if the upper mediastinum was involved
Examination	Typically, a non-tender mass with an unclear boundary. The larger the mass, the worse the mobility Approximately 38% of recurrent cases were a hard mass beneath the scar and were associated with pain and rapid growth
Imaging studies	Ultrasound, computed tomography (CT) and/or magnetic resonance imaging (MRI) revealed a soft tissue mass on the chest wall with uniform density and an unclear boundary. The average diameter was 6.9 cm (maximum diameter, 27 cm) In 17 patients who received a bone scan, radionuclide concentration in the lesion was observed in 8 patients, and bone invasion was pathologically confirmed in two cases
Involved areas	Chest wall muscles and ribs, 29 cases (61.7%); scapular area, 10 cases (21.3%); clavicular area and thoracic inlet, 10 cases (21.3%); thoracic outlet, 8 cases (17%); breast, 6 cases (12.8%); intrathoracic area, 5 cases (10.6%); paravertebral area, 5 cases (10.6%); mediastinum, 4 cases (8.5%); diaphragm, 3 cases (6.4%)

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