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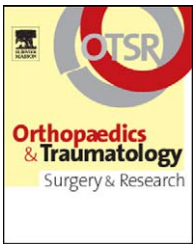

ORIGINAL ARTICLE

Primary or recurring extra-abdominal desmoid fibromatosis: Assessment of treatment by observation only

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KEYWORDS
Desmoid fibromatosis;
Soft tissue tumor;
Surgery;
Conservative treatment;
Simple observation

Summary
Introduction: Extra-abdominal desmoid fibromatosis (EADF) is a benign tumoral condition, classically managed by more or less radical and sometimes mutilating excision. This treatment strategy is associated with a recurrence rate of nearly 50% according to various reports.
Hypothesis: EADF may show spontaneous stabilization over time.
Methods: A retrospective series of 26 cases of EADF managed by simple observation was studied to assess spontaneous favorable evolution and identify possible factors impacting evolution. Eleven cases were of primary EADF with no treatment or surgery, and 15 of recurrence after surgery with no adjuvant treatment. MRI was the reference examination during follow-up.
Results: Twenty-four cases showed stabilization at a median 14 months; there were no cases of renewed evolution after stabilization. One primary tumor showed spontaneous regression, and one recurrence still showed evolution at end of follow-up (23 months). The sole factor impacting potential for evolution was prior surgery. No radiologic or pathologic criteria of evolution emerged from analysis.
Discussion: The present series, one of the largest dedicated to EADF managed by observation, confirmed recent literature findings: a conservative ''wait-and-see'' attitude is reasonable and should be considered when large-scale resection would entail significant functional or esthetic impairment.
Level of evidence: Level IV, retrospective study.

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Extra-abdominal desmoid fibromatosis (EADF) or aggressive fibromatosis is a rare tumor developing in the musculo-aponeurotic structures. It is a low-grade soft tissue tumor, which is systematically benign, without potential malignancy or remote dissemination, but showing severe local aggression and unpredictable evolution following treatment. The reference attitude is extensive surgical resection which, however, entails a risk of functional sequelae and significant morbidity with a high rate of recurrence even when resection extends to neighboring healthy tissue, due to the infiltratory character of EADF [1—5].

The literature contains certain reports of short series or occasional cases showing stabilization or regression of non-operated primary or recurrent EADF managed by simple surveillance [3,6—8]. These reports encouraged us to try simple wait-and-see surveillance in 26 of the EADF patients managed in our center between 1989 and 2009.

The study sought to assess the reality and frequency of spontaneous favorable evolution in the series, and to identify predictive factors for evolution so as to improve treatment strategy.

Material and methods

Material

The main inclusion criterion was confirmed presence of all anatomopathologic diagnostic signs of EADF on biopsy of non-operated tumors or in the exeresis specimen in case of recurrence after surgery:

- architectural criteria: tumor proliferation of fibroblastic or myofibroblastic spindle cells, without areas of necrosis, over a collagen ground rich in broad divergent bundles and a few vessels surrounded by clear space. At the periphery of the tumor, there may be small lymphoid islands. These tumors are poorly contoured, invading fat and muscle;
- cytologic criteria: myofibroblasts showing monomorphic nuclei with between one and three small nucleoli and an occasional mitosis.

Immunomarking: systematic exploration for smooth-muscle actin, beta catenin, desmin, caldesmon, AE1, AE3, EMA, PS100 and CD34 markers.

All included patients had been managed in the department for EADF between 1989 and 2009.

Data were collected by systematic retrospective harvesting of all historical, clinical and surgical records. Slides and MRI slices were systematically reassessed.

All patients with complementary medical treatment or radiochemotherapy (Glivec, Tamoxifen, anti-TNF alpha, Indocid, etc.) were excluded.

No patients were lost to follow-up.

In all, 45 patients were treated for EADF, 26 of whom (57%) underwent simple radioclinical surveillance. Two subgroups could be distinguished:

- primary EADF (11 cases), with no surgical or medical treatment;
- recurrent EADF (15 cases), undergoing surveillance after one or more surgical operations: 14 of the 34 EADF patients operated on once or more during the study period were cured; five recurrences were managed medically and 15 underwent simple radioclinical surveillance and were included in the present study.

The choice between surgery and simple surveillance was based on the feasibility of sequela-free marginal resection.

Methods

Surveillance comprised 6-monthly clinical examination and systematic MRI. MRI comprised sagittal, frontal and coronal T1, T2 and gadolinium-enhanced sequences. The evolution criteria were tumor size on the longest axis and change in tumor signal.

Events were dated according to age on the day of initial diagnosis. Surveillance of primary EADF was referenced by the date of initial diagnosis and of recurrent EADF by the date of the diagnosis of recurrence.

Exeresis quality was assessed on the Union Internationale Contre le Cancer (UICC) R classification [9].

Data submitted to analysis concerned tumor location, size and MRI signal.

Statistical analysis

Survival was analyzed using the Kaplan-Meier method. The event considered was recurrence. Mean values were compared by Fisher's F-test; the significance threshold was set at 5%.

Results

General series characteristics

The series comprised 26 cases, with an M/F sex-ratio of 1/10 for primary EADF and 1/2 for recurrent EADF, or 1/3.3 for the series as a whole. In recurrent EADF, mean age on the day of diagnosis of recurrence was 36 years (range, 14—67 years) and, in primary EADF, mean age on the day of diagnosis of primary tumor was 35.5 years (range, 21—73 years).

In primary tumor cases, discovery involved tumefaction in all cases, with associated pain in four. In previously operated patients, recurrence was diagnosed on control MRI in all cases, with associated tumefaction in seven. There were no histories of Gardner syndrome; trauma was noted in seven cases, but could not be formally linked to the tumoral pathology (shoulder, thigh or calf).

Surveillance found stabilization at a median 14 months: by month 14, tumor evolution had stabilized in half of the patients; Fig. 1 shows the cumulative incidence curve for evolution arrest.

Mean follow-up after case-by-case stabilization was 12.7 months (range, 2—27 months) in the primary EADF group (Figs. 2 and 3) and 19.1 months (range, 1—80 months) in the recurrence group. No surgery was required in any patient during surveillance.