Desmoid Tumours: Experience of the Medical Oncology Department of Pierre and Marie Curie Center Algiers

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Abstract

Introduction

Desmoid tumours, also known as aggressive deep fibromatoses, are soft tissue tumours. Histologically benign, they are characterised by a slow and infiltrative evolution with a high rate of local recurrence.

Patients and methods: We report a retrospective series of 10 patients treated in the medical oncology department of the Pierre & Marie Curie Centre over a period of 4 years. 6 were women and 4 men, with a sex ratio of 1.5 in favour of women.

The average age was 36 [23-53]. 20% (n=2) of patients had familial adenomatous polyposis. 90% of patients had an extra-abdominal desmoid tumour compared with only 10% in the abdominal wall. 50% of patients underwent initial resection, 60% of whom recurred after 1 year. Only one patient underwent additional radiotherapy.

Initial surveillance was proposed in 50% of patients. 60% progressed after 2 years. Only one patient underwent surgical treatment combined with radiotherapy after an initial monitoring period of 9 years. Treatment with IMATINIB 400mg/d was initiated in 60% (n=6) of patients after progression or local recurrence. One patient received tamoxifen for one year.

Results: Lesion stability was observed in 6 patients (60%), a partial response (left scapular) in 1 patient and local progression in 1 patient. Tolerability: Grade 3 neutropenia in 1 patient and Grade 2 liver toxicity.

Conclusion: Desmoid tumours are rare tumours which progress slowly and are locally aggressive. Their prognosis is closely linked to their recurrence. The current therapeutic approach advocates a "WAIT AND SEE" strategy, particularly in asymptomatic, non-progressive forms. Surgery is only indicated if the immediate vital or functional prognosis is at risk.

Keywords: Desmoid tumour, Recurrence, imatinib

Introduction:

Desmoid tumours or deep fibromatoses are rare tumours that develop from musculoaponeurotic structures. Developing from musculoaponeurotic structures. These are rare rare tumours affecting young adults and are characterised by a polymorphism of clinical polymorphism Histologically benign, they are characterised by a slow and infiltrative evolution with a high rate of local recurrence. Their
management is currently debated, and the old standards have been widely questioned in recent years.

**Methods**

This was a retrospective, single-centre study of 10 patients treated at the medical oncology department of the Pierre and Marie Curie Centre from August 2014- December 2020. Characteristics of the population:

1. Sex: sex ratio= 1.5 for women
2. Age: mean age= 36 years
3. 20% of patients (n=2) had familial adenomatous polyposis.
4. Tumour location (Figure 1): 90% were extra-abdominal (thorax and limbs), abdominal site (mainly thorax and limbs) and 10% are of abdominal

**Results:**

50% of patients underwent primary resection compared with 50% who underwent initial monitoring (Figure 2) 60% (n=3) progressed after 2 years of surveillance while 60% (n=3) recurred 1 year after initial surgery.

1. 2 patients underwent additional radiotherapy.
2. Revision surgery was proposed in 3 patients
3. Medical treatment was proposed in 6 patients (n=6) after local recurrence or progression of the disease.
4. Tamoxifene -based hormone therapy was prescribed in one patient for patient for 1 year.
5. Targeted therapy such as IMATINIB 400mg/d was proposed in in 08 patients.
6. Response to treatment was judged on the basis of clinical and radiological RECIST radiological criteria. The lesion was stable in 6 patients (60%), partial response (left scapular) in 1 patient and local progression in in 1 patient Grade 3 neutropenia and grade 2 hepatic toxicity were noted with IMATINIB with IMATINIB

**Discussion**

Desmoid tumours are rare tumours representing 3% of soft tissue soft tissue tumours with an annual incidence rate of 2-4 nv cases/ year/100000 population (1).They occur sporadically (n=8) or associated with a PAF (2% in the literature compared with 20% in our series). Their management has evolved considerably as a result of the knowledge acquired in recent years. recent years. Currently, initial monitoring of the WAIT and SEE type is preferred. SEE "is preferred to a first surgical procedure as several studies have studies have demonstrated comparable disease progression with a similar rate of recurrence rate (2) (3) (4); our study found a similar recurrence rate of rate of 60% in both attitudes. Radiotherapy alone or as remains controversial despite a demonstrated benefit in local local control of the disease in the case of positive margins (5).

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in the case of inoperable or recurrent lesions. The efficacy is modest and varies greatly from one individual to another from one individual to another (6). TKIs, and in particular IMATINIB, have been used in 3 phase II trials (7)(8)(9) to achieve objective response rates of 15.7%, 12% and and 6% and 1-
year recurrence-free survival rates of 36.8%, 67% and 66%. In our study, lesion stability was achieved in 67% of cases. No deaths were reported in our series.

Conclusion
Desmoid tumours never metastasise but their prognosis is essentially linked to their recurrence, which may jeopardise functional prognosis. Appropriate management requires knowledge of the progression of these tumours. The current attitude is to refrain from treatment in the absence of prognostic or predictive factors. Prognostic or predictive factors for response to any proposed treatment.

References
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Figure 1: Tumors desmoid location
Figure 2: initial treatment

Figure 3: Therapeutic Response