International Journal for Multidisciplinary Research (IJFMR)
E-ISSN: 2582-2160 ● Website: www.ijfmr.com ● Email: editor@ijfmr.com

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

E.Kerboua[1] , R.Nemmar[2]

[1]Medical oncology, Pierre and Marie Curie Center, Algiers University -Algeria
[2]Orthopaedic surgery and traumatology, M Algiers University Mustapha Hospital

Abstract

Introduction

Desmoid tumours, also known as aggressive deep fibromatoses, are soft tissue tumours. Histologically benign, they are characterised by a slow infiltrative course 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. 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 nitial 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).

Medical

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 their prognosis is essentially linked to their recurrence, which may jeopardise functional prognosis. Appropriate management requires knowledge of the 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. treatment.

References

1. Jean Pierre Droz, Isabelle Ray-Coquard, Jean-Louis Peix: rare malignant tumours
2. Bonvalot, S. (2008) Actualisation des indications chirurgicales des tumeurs stromales gastro-intestinal stromal tumours. Journal de Chirurgie, 145, 6S8-6S11
3. Marco Fiore et al. Ann Surg Oncol . September 2009.
4. Honeyman, J., Theilen, TM, Knowles, ME et al. (2013) Desmoid fibromatosis in children and adolescents: a conservative approach to management. Journal of Paediatric Pediatrics, 48, 62-66. http://dx.doi.org/10.1016/j.jpedsurg.2012.10.017
5. 5. Nuyttens JJ, Rust PF, Thomas CR, Jr, et al. Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: A comparative review of 22 articles. Cancer 2000:88(7):1517-1523
6. Armelle Dufresne: A study of the signalling pathways involved in the carcinogenesis and treatment of aggressive fibromatoses
7. Keith M Skubitz et al.Response of aggressive extra-abdominal imatinib-resistant fibromatosis to sunitinib: a case report and review of the literature on response to tyrosine kinas inhibitors. Pharmacol of cancer chemotherapy . August 2009 .
8. N Penel et al . Imatinib in the treatment of aggressive progressive and recurrent fibromatosis (desmoid desmoid tumours): a phase II FNCLCC/French Sarcoma Group trial with long-term follow-up. Ann Oncol . February 2011 .
9. Bernd Kasper et al Imatinib induces sustained progression arrest in RECIST progressive desmoid tumours : Final results of a phase II study of the German Interdisciplinary Sarcoma Group (GISG). Eur J Cancer. 2017 May.

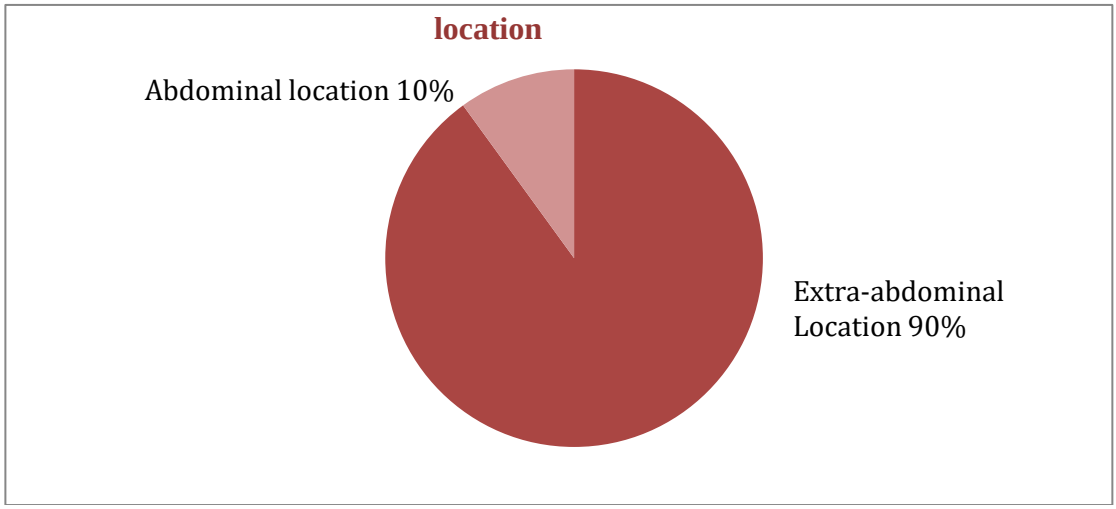

Figure 1 : Tumors desmoid location

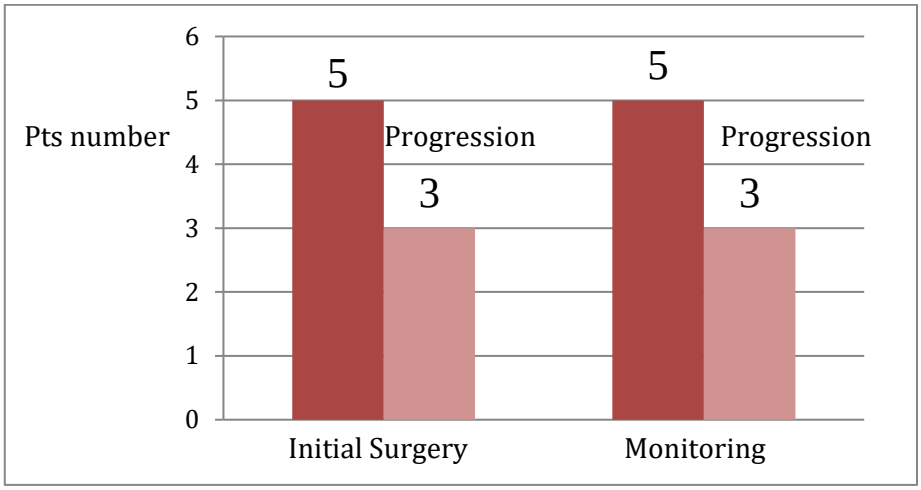

Figure 2 : initial treatment

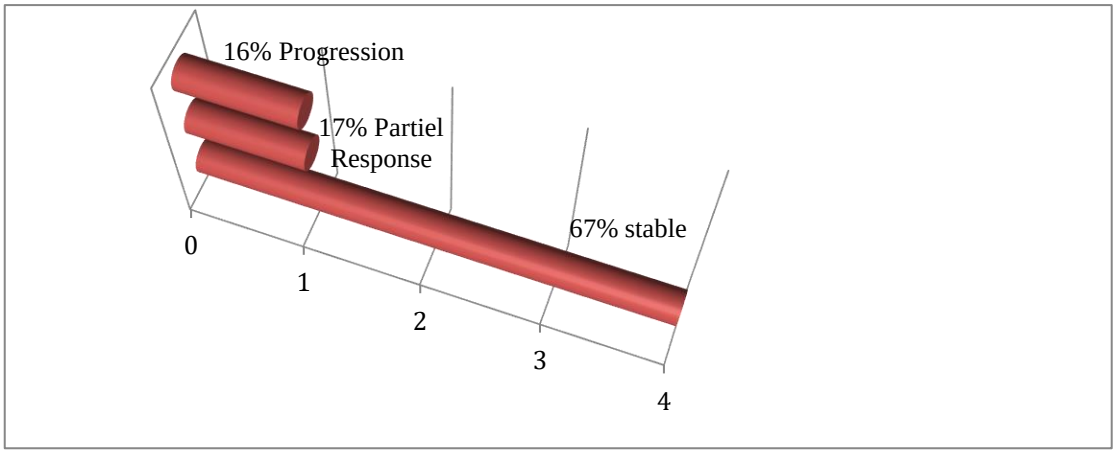

Figure 3 : Therapeutic Response