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Outcome of non-surgical management of extra-abdominal and abdominal desmoid-type fibromatosis: a population based study.

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Background

Non-surgical management of patients with desmoid-type fibromatosis (DF) is increasing. The present study reports type and outcome of first-line non-surgical management. Aim is to provide insight on which treatment is applied and outcome.

Methods

In the Dutch Pathology Registry (PALGA) patients diagnosed between 1993 and 2013 having extra-abdominal or abdominal wall DF were identified. Medical records were retrieved. Only first-line treatment was analysed. Best response, using RECIST-criteria and time till progression (TTP) were analysed.

Results

1134 patients were identified, 96 patients had first-line non-surgical management and were included. 40 patients had surveillance (42%), 36 patients received radiotherapy (37%), 20 patients received systemic treatment (21%). Surveillance resulted in spontaneous complete response (CR) in 3/40 patients, partial response (PR) in 6/40 patients, stable disease (SD) in 14/40 patients, and progressive disease (PD) in 15/40 patients. Median TTP was 7 months (interquartile range (IQR) 5-27). Radiotherapy resulted in CR in 3/36 patients, PR in 15/36 patients, SD in 12/36 patients and PD in 2/36 patients. TTP was 31 and 47 months. Outcome of systemic treatment was CR in 1/20 patient, PR in 1/20 patient, SD in 9/20 patients and PD in 4/20 patients. Median TTP was 7 months (IQR 6-106). Overall, 66% of patients had CR, PR or SD.

Conclusion

In the present 20-year nationwide population-based study, 1134 patients were diagnosed with DF. 8.5% primarily had non-surgical management, resulting in distinct outcome parameters. These outcomes should be taken into account when a non-surgical management is discussed with an individual patient.