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Spontaneous regression in desmoid fibromatosis (DF)

Desmoid fibromatosis (DF) is a locally aggressive rare tumor with an unpredictable clinical course. Active surveillance (AS) is the strategy of choice for almost all DF patients. Recent publications, including three independent European prospective observational studies and the latest consensus update, have stated that AS in patients with extra-abdominal desmoid-type fibromatosis should be the frontline approach for almost all patients with DF, with few exceptions carefully described in the JAMA Oncology consensus paper. Moreover, in all prospective studies, spontaneous regression has been described as a common clinical event occurring in approximately 25-30% of patients as a primary event, but also after RECIST progression in 30% of patients. Spontaneous regression seems to be related to clinical and pathological variables, including tumor size, anatomical site, and specific mutation. However, the biological mechanism has not yet been described. A potential role of the immune system and inflammation has been proposed, but further studies are needed. The probability of tumor regression should be taken into account when considering initiating active treatment. In fact, if regression is expected, a longer period can be waited in the absence of any other symptoms.