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Title:

Diagnostic and therapeutic strategies for extra-abdominal desmoid-type fibromatosis: a longitudinal questionnaire survey to specialized centers in Japan

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Abstract

The mainstay of treatment modality for extra-abdominal desmoid-type fibromatosis (DF) has shifted from surgery, which often reduces ADL/QOL, to conservative treatment, active surveillance. In the present study, we conducted a longitudinal survey on the diagnosis and treatment of DF at facilities belonging to the Japanese Musculoskeletal Oncology Group, which is a research group of facilities specializing in the treatment of bone and soft tissue tumors in Japan. The same questionnaire was administered in 2015 and 2018, and responses were obtained from 46 (69%) of 67 facilities and 42 (53%) of 80 facilities in 2015 and 2018, respectively. Although immunostaining for β -catenin was often used for pathological diagnosis in both 2015 and 2018, CTNNB1 mutation analysis was not performed not only in 2015, also in 2018. As for the treatment strategy for resectable cases, surgical treatment including wide resection was selected at 11 centers (24% of respondents) in 2015, and further

decreased to 5 centers (12%) in 2018. Conservative treatment with active surveillance or medical treatment were the most common treatments for both resectable and difficult-to-resect cases. COX-2 inhibitors and tranilast were often used as drug treatments for both resectable and difficult -to-resect cases. There were few facilities that provided radiotherapy, methotrexate and vinblastine, and DOX-based chemotherapy for refractory cases in both 2015 and 2018. It will be further necessary to disseminate clinical practice guidelines to physicians more widely, and to have them understand and implement medical practice strategies for this rare disease, DF.