The treatment of desmoid tumors associated with familial adenomatous polyposis: the result of a Japanese multicenter observational study

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Abstrak

PURPOSE: Familial adenomatous polyposis (FAP)-associated desmoid tumor (DT) is sometimes life threatening. However, the optimal treatment for DTs has not been established. The aim of this study was to analyze the outcomes of surgical and pharmacological treatments for DT in Japanese FAP patients.

METHODS: We retrospectively reviewed the data of 303 patients who underwent colectomy for FAP between 2000 and 2012. We analyzed 41 patients with DTs in which the location was apparent. The selection of treatment for intra-abdominal DTs was also evaluated according to Churchs classification.

RESULTS: Surgery was frequently used to treat extraabdominal DTs. Multimodal treatments, including surgery, and the administration of non-steroidal antiinflammatory drugs, hormonal therapy, and chemotherapy were widely used for intra-abdominal DTs. The most effective pharmacological treatment was cytotoxic chemotherapy, which was associated with a response rate of 45.5% and a disease control rate of 72.7%. After a median follow-up period of 53.0 months, the 5-year DT-specific survival rate in patients with stage IV disease was 71.4%; in contrast, the rate in patients with other stages was 100%. Four-stage IV patients died of DT due to uncontrollable rapid progression. No cytotoxic chemotherapy was administered; however, incomplete resection was performed in three cases.

CONCLUSION: Our findings will provide clues that may help physicians in selecting the optimal strategy for this rare disease.