

FAP-RELATED DESMOID TUMORS TREATED WITH LOW-DOSE CHEMOTHERAPY

results from a multicentre retrospective analysis

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BACKGROUND

Desmoid tumours (DTs) are monoclonal neoplasms with fibroblastic-myofibroblastic differentiation and they represent the most common extra-intestinal manifestation of **familial adenomatosis polyposis (FAP)**.

DTs are often multifocal and, even in the absence of a metastatic potential, they represent the first cause of death in FAP patients after colectomy.

Data on the activity of chemotherapy in FAP-associated DTs are limited. We specifically examined the activity of chemotherapy with low-dose methotrexate (MTX) + vinca alkaloids.

METHODS

We retrospectively reviewed data from all patients treated with MTX + vinca alkaloids for FAP-associated DTs in 5 reference centres and cases included into the National rare cancer network were also reviewed and included if sufficiently informative for the study purposes (**Table 1**).

Radiological responses were assessed using both RECIST and Choi criteria.

Table 1

Patient characteristic	
Sex (M:F)	13:15
Age at diagnosis (years)	34 (17-57)
Multifocal disease	12/28 (42,8%)
Previous surgical treatment	17
Previous therapy	9
- COX2 inh	3
- HT	6

RESULTS

We identified 28 patients treated with MTX + vinca alkaloids. All patients had progressive disease before chemotherapy; 17 patients and 9 patients had previously received respectively surgery and/or systemic treatments (i.e. hormone therapy, NSAIDs).

Chemotherapy was administered for a median duration of 11 months. According to RECIST criteria (Choi evaluation is ongoing) complete response, partial response, stable disease, and progressive disease were observed in 1, 17, 10, and 0 patients, respectively.

The median progression-free survival (PFS) was 78 months (Fig 1 and Table 2); it was 124 months in responding patients (Fig 2 and Table 2).

Table 2

	Median PFS (95% CI) (months)
Responders (CR + PR)	124,0 (71,5-176,5)
Non-responders (SD)	45,0 (30,4-59,5)
Overall	78,0 (38,4-117,6)

Log Rank Mantel-Cox test
 $p = 0.089$

Exemplificative response of a partial response is shown in Fig. 3

Fig. 1

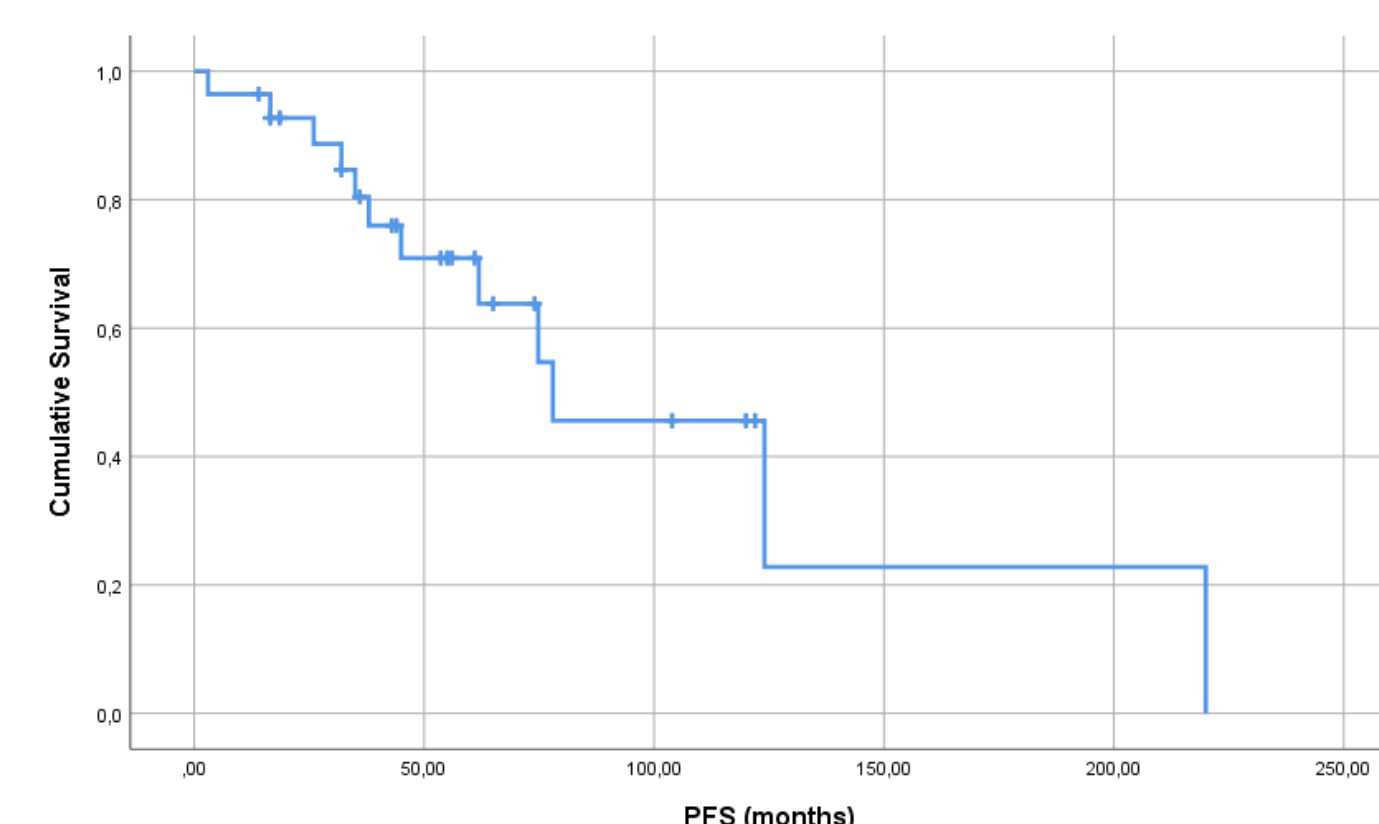


Fig. 2

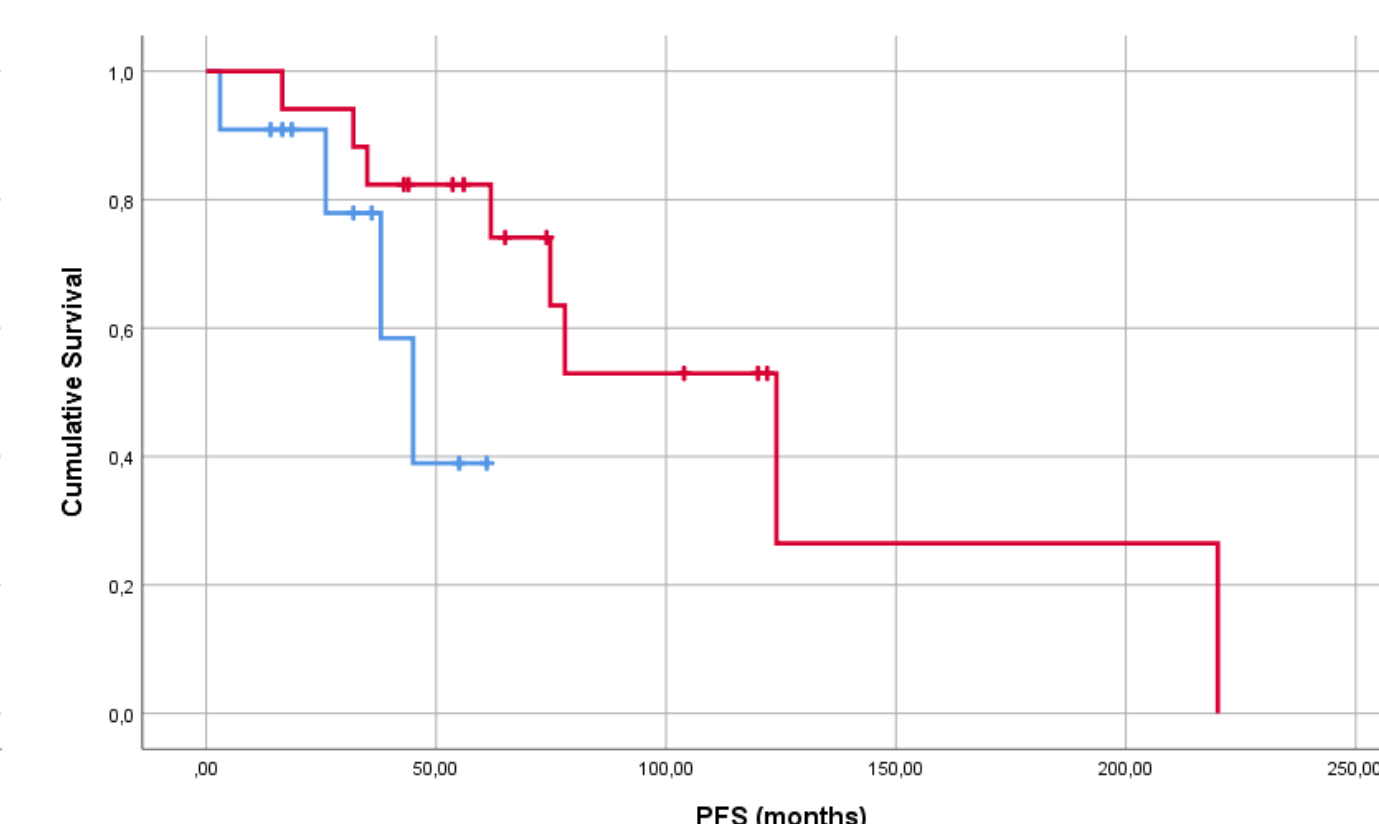


Fig. 3



After chemotherapy withdrawal, MTX + vinca alkaloids rechallenge was offered to 11 patients with progressive disease. In these patients, we obtained a control rate of 100%, resulting in a median second PFS of 64 months.

CONCLUSIONS

To the best of our knowledge, this is the largest series on the activity of low dose chemotherapy in FAP-related DTs. Our data suggest a tremendous activity of low dose chemotherapy in this very rare subset of patients.