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ABDOMINAL DESMOID TUMORS: A SERIES FROM A SINGLE INSTITUTION.

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Desmoid tumour (DT) is a rare disease, accounting 5-6 cases per million of the population / year which is locally aggressive with no potential for metastasis or dedifferentiation.

Women are slightly more affected than men with a peak age of 30-40 years.

Active surveillance is now considered the best first approach for stable, asymptomatic primary or recurrent desmoids, particularly if resection would entail major morbidity. Treatments should be considered only in case of persistent progression or increase of symptoms.

Intraabdominal desmoids are often less resectable than extra-abdominal and abdominal wall ones because they diffusely infiltrate the mesenteric roots and are often multiple, leading to unacceptable morbidity, thus discouraging surgery in favour of medical therapy.

We report a retrospective review of our single centre series of intra-abdominal DT treated at the European Institute of Oncology (IRCCS IEO Milan) from January 2003 to March 2019. Twenty-two patients affected by DT were identified, all histologically proven by a biopsy, whose 13 were males. Median age was 41,5 years. 19/22 underwent to surgery, 17/18 (94%) with negative or positive microscopic margins (R0-R1) and only one was intralesional (in one case data about margins were not available). RT was never used postoperatively, instead medical treatment (Tamoxifen 200 mg in combination with Celecoxib 20 mg) was administered to the only patient with intralesional margin at surgery.

Median follow up was 80 months (range: 3-209 months) and no death related disease was recorded, neither major or minor morbidity surgery-related. Four patients locally recurred, whose 2 presented R1 margins at first surgery. Relapses were treated in 1 case conservatively and in one case with a further marginal intervention, while two patients received respectively RT and CT obtaining stable disease. Unfortunately, the patient who underwent to RT died for cause different from DT.

DT treatment changed in the last years moving from a mainly surgical approach to a more conservative one, due to the observation that non operative management produces comparable outcomes to upfront surgery. Thus, watchful waiting has been advocated for asymptomatic patients but surgery remains the mainstay of treatment in case of symptoms or progression. In our series of abdominal desmoids very few patients (4/22) have been treated in the last four years. One out of these 4 underwent to surgery, to further underline the changed therapeutic approach to DT.

Of interest, when a surgery has been the treatment of choice, relapse rate was low, justifying surgical resection with R0/R1 margins in symptomatic or progressive disease.

Tables

Table 1: Epidemiologic data

EPIDEMIOLOGIC DATA	
Number of patients	22
Mean age	41
Median age	41,5
Sex (male:female)	13:9
Mean age male	41,5
Mean age female	43,4
Biopsy	22 (86%)
Surgery	19 (1 lost at f-up)

Table 2: Surgery

SURGERY	
Pts undergone to surgery	19
Surgery outside IEO	8
Quality of surgery	18 (1 data not reported)
radical	13
marginal	4
intralesional	1

Table 3: Adjuvant treatment

ADJUVANT TREATMENT	
RT	0, (3 data not reported)
CT	1 intralesional (3 data not reported)

Table 4: Recurrences

LOCAL RELAPSE	
Patients who relapsed	4
Surgery at IEO	2

Quality of first surgery	
radical	1
marginal	2
unknown	1
Time from surgery	4 mos, (IEO) 8 mos (IEO), 5 yrs, 1 yrs
To treat relapse	
Surgery	1 (first surgery outside IEO) but not effective
RT to treat relapse	1 (dead for other disease)
CT to treat relapse	1 Tamoxifene-Celebrex with SD
State at F-UP	
Alive with disease	3
Dead	1 (for other disease)

Mos: months

SD: stable disease

Table 5: Follow up

FOLLOW UP	
Mean (months)	73,2
Median	80
Range (months)	3-209
Lost at fup	1