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Sameer Rastogi, Kaushal Kalra, Adarsh Barwad, Ekta Dhamija, Sandeep boriwal

AIIMS, New Delhi

Background

Data for desmoids tumors from India is sparse limited to case series and case reports. In last few years the emphasis on medical management has increased so it would be pertinent to see the outcomes of desmoid tumor from dedicated sarcoma / desmoid medical oncology clinic from India.

Method

This was a retrospective study from prospectively maintained database from January 2016 to July 2020 of desmoids tumors from sarcoma medical oncology clinic from All India Institute of Medical Sciences (AIIMS, New Delhi). SPSS 24 ver was used for statistical calculations. Kaplan Meyer curves were used to calculate survivals and log rank test was used for univariate analysis.

Results

There were total of 107 patients during this time with females marginally higher than males (62:45, 1.3:1). Median age was 26 years (age group ranging from 2 to 80 years). Seventy seven patients had outside reported biopsy and 23% of these patients had discrepant diagnostic report from outside. Most common site was upper limb (52%) and lower limb (15%) and abdomen (15%). Previous surgery was done in 65 patients (61%) while previous medical lines were given in 27%. Ninety percent of previous medical therapies were tamoxifen based regimen. Ninety eitght patients had disease had presentation while 9 patients had no disease (referred post surgery). For patients with disease the pattern of treatment for first line, second line and third line is shown in the table 1. For first line patients on sorafenib, tamoxifen and observation there was no statistically significant difference. Though the progressive disease was higher in tamoxifen and observation arm. Chemotherapy had dismal outcome in first line probably due to the fact it was used in aggressive disease (all patients were non extremities). Median PFS in chemotherapy was 15 months. In all lines sorafenib was given to 53 patients. Of all patients in which response were available partial response was there in 31% of patients. For all patients who received sorafenib, 2 year progression free rate was close to 80%. Only 28 percent patients could tolerate 400mg od dose. Overall 4 patients died due to disease while 4 patients had amputation.

Conclusion

Our patients had younger age as compared to western data and could be attributed to inclusion of pediatric patients as well. Majority of our patients had upper limb fibromatosis as compared to western literature. Misdiagnosis of desmoids outside tertiary care centre like us is quite high and in our case it was 23%. We couldn't find statistically significant benefit between various groups and it could be

explained by small numbers. Secondly we gave sorafenib and chemotherapy to rapidly progressive or symptomatic tumors. Chemotherapy was given mostly to abdominal and neck fibromatosis in which urgent response was needed. While observation was done for seemingly indolent tumors. Outcome on sorafenib is similar to previously documented data. 400mg dose seems to be poorly tolerated dose and needs dose reduction in majority. Though it is benign disease yet 4 patients died due to disease while 4 patients needed amputation as final resort.

Table 1 Pattern of first line treatment in sarcoma medical oncology clinic AIIMS during 2016 to 2020

	sorafenib	Tam based	Observation	Chemotherapy	Others
Treatment in our hospital (first line) N=98	34 (35%)	31 (32%)	21(21 %)	10(10%)	2(2%)
Second line in our hospital (n=32)	16 (50%)	2 (6%)	5 (15%)	5 (15%)	Surgery -3 Pazopanib =1
Third line	4			1	Pazopanib =2 Surgery =1