RESULTS: Recurrence occurred in seven patients (33%) and six of them relapsed within the first 18 months. No significant difference was observed between conservative and surgical treatment. However, a significant difference was observed among patients undergoing wide resection and who experienced improved local control.

CONCLUSION: The recurrence rate of desmoid tumor was 33.3%. There was no difference in recurrence between conservative and surgical treatment. In surgical treatment, wide margins showed better results for recurrence control. Level of Evidence III. Retrospective Observational Study.

Keywords: Fibromatosis, aggressive. Medical oncology. Recurrence.
minimum and maximum follow-up periods were 16 and 127 months, respectively. With respect to tumor location, 14 (66.7%) occurred in the lower limbs, four (19.0%) in the upper limbs, and three (14.3%) in the trunk. The most frequent specific location was the popliteal fossa, observed in four (19.0%) patients. The average tumor size was 12.7 ± 7.5 cm, varying between 4.5 cm and 36.4 cm. Epidemiological data are summarized in Table 1. According to the initial treatment, 14 (66.7%) patients received no previous treatment and seven (33.3%) experienced local recurrence of previously manipulated tumor. Among the 21 patients, 16 (76.2%) underwent surgical treatment and five (23.8%) underwent conservative treatment. As for tumor margin in patients subjected to surgery, eight (50%) patients had wide margins, seven (43.7%) were classified as marginal, and only one (6.3%) was intralesional. Adjuvant radiotherapy was performed in one (6.3%) patient.

Among the five patients subjected to conservative treatment, three underwent hormone therapy. The hormone of choice was tamoxifen for two (40.0%) patients and progesterone for one (20.0%) patient. The other two (40.0%) patients were treated with non-steroidal anti-inflammatory drugs and were under observation (expectant management).

Statistical analysis was performed using SPSS® 17.0 software (Chicago, USA). The descriptive study was conducted using frequencies and percentages for the categorical variables, and the measures of central tendency (mean and median) and dispersion (standard deviation) were used for quantitative variables. The response variable analyzed was local recurrence (surgery) or lesion progression (conservative treatment). This variable was compared to age, gender, type of treatment, prior manipulation, surgical margins, tumor location, tumor size, and number of previous recurrences.

For the comparative study, an univariate analysis was performed using the Kaplan–Meier method associated with the results of the log-rank test. A multivariate analysis was performed using the goodness-of-fit test for a Cox’s regression model. The variables with p-values lower than 0.25 in the univariate analysis were used in the selection of covariates for adjustment of the final model. A value of p≤0.05 was considered statistically significant.

RESULTS

Local recurrence/progression was observed in seven (33.3%) patients during follow-up and the average recurrence period was 14.1 ± 10.2 months. Six (85.7%) cases involving recurrence/progression occurred before 18 months. The survival curve is depicted in Figure 1. Among the relapsed patients, only one (14.3%) developed multiple recurrences. The comparative analysis showed that gender (p = 0.253), age (p=0.660), tumor location (p=0.839), tumor size (p=0.990), and previous recurrence (p=0.930) did not significantly correlate with recurrence/progression after treatment. The data are summarized in Table 1.

With regard to the disease treatment, the recurrence rate in the surgical treatment did not significantly differ from that in the conservative treatment. However, in patients subjected to surgery, the presence of wide margins was better and with a significant results (p=0.020) when compared with marginal and intralesional excision. (Figure 2) The relative risk of recurrence among patients with intralesional margins was 24.6 times higher than that of patients with wide margins (95% CI: 1.2–508.3).

DISCUSSION

Desmoid tumor or aggressive fibromatosis was first described by MacFarlane in 1832. It is characterized by rare injuries with an incidence of 2-4 cases per 1,000,000 and it is predominant among young adults and females.° Its etiology remains unknown but is believed to be a monoclonal fibroblast proliferation associated with increased production of β-catenin protein, which stimulates fibroblast activity.° It is typically characterized by firm and painless
lesions, which harden and adhere to surrounding tissues. The growth is insidious but during its progression, it may cause significant functional limitation.

In the present study, we evaluated a series of patients with the epidemiological profile observed in most studies, with a mean age of 33.0 years and predominantly females. In contrast to other authors, who observed a higher frequency of lesions on the trunk and on the pectoral and pelvic girdle, we found greater involvement of the lower limbs.

The tumor’s biological behavior remains undefined and variable. Some lesions are aggressive, with multiple local recurrences and resistance to treatment. Tumors located in the head, neck, and chest can cause death by direct compression of vital structures.

However, cases of spontaneous regression or regression after biopsy have been described. Frequent recurrence of these lesions has been widely reported, and the estimated recurrence rate varies between 15% and 75%. In our study, the recurrence rate was 33.3%, and in 85.7% of those cases, it occurred within 18 months after treatment.

The treatment options included surgery, chemotherapy, hormonal therapy, and radiation therapy, either alone or in combination. Each of these treatments has demonstrated some degree of success. However, local recurrence remains as a problem. Surgical treatment with wide margins remains the method of choice for most patients with desmoid tumor. However, frequent recurrence and the functional and cosmetic sequelae of this procedure enables conservative treatment as a good option in cases of multiple recurrences, unresectable tumors, and in cases in which it is difficult to determine tumor location.

We observed that wide margins are essential for the adequate control of fibromatosis, as previously reported by Duggal et al. Most authors acknowledge the importance of the surgical margin. However, Rock et al. did not find any benefit of extended margins for the control of desmoid tumors.

Among the conservative therapies, radiation therapy has been the most studied. Its application is usually described as a surgical adjuvant for large, unresectable tumors or in cases of contaminated margins. Jelinek et al. reported a local control rate of 53% in patients subjected to surgery and 81% in those patients subjected to surgery and radiotherapy.

The influence of hormones and the presence of estrogen receptors in these tumors has stimulated local control using hormone therapy, either as adjuvant or as main therapy. Virtually, all tumors express the nuclear estrogen receptor beta, but not all patients respond to anti-hormonal therapy.

The best studied anti-estrogen is tamoxifen; however, most studies are based on a small sample size of patients, and no prospective studies using a sufficient number of patients are available to indicate the ideal dosage. Other anti-estrogen drugs used include progesterone, medroxyprogesterone, and testolactone.

Recently, Briand et al. reported their experience with patient follow-up without treatment of desmoid tumor. Based on the findings by Gouin et al. and Barbier et al., who indicated a high rate of spontaneous interruption of tumor growth, expectant management was adopted as the initial treatment, and the percentage of tumors without progression reached 90%.

Our study showed that the surgical and conservative treatment options are useful for local control of desmoid tumor. We believe that surgical intervention is the best option for primary lesions, located where wide margins can be reached with low functional and cosmetic morbidity. Conservative treatment is an alternative for large lesions after multiple recurrences and in cases in which surgery will result in significant functional limitation.

CONCLUSIONS

The recurrence rate of desmoid tumor observed in our sample was 33.3%. No difference in recurrence was observed between patients submitted to the surgical and conservative treatments. The wide margins showed better results for local control of the disease after the surgical treatment.
AUTORS’ CONTRIBUTION: LEMT (0000-0003-1276-5679)*, ACE (0000-0003-0265-1547) * and RFV (0000-0002-7025-0457)* were the main contributors in the drafting the manuscript. LEMT and RBC (0000-0002-1430-6006)* performed the surgeries, followed up the patients and collected clinical data. ACE and RFV evaluated data from the statistical analysis. ACE, RFV, CBG (0000-0002-0772-0700)* and MAP (0000-0001-6284-213X)* carried out the literature search, reviewed of the manuscript and contributed to the intellectual concept of the study. * ORCID (Open Researcher and Contributor ID).

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