Rhabdomyosarcoma of the extremities; analysis of 20 patients.

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Abstract:

Background: In children, rhabdomyosarcoma occurs with a frequency that is equal to or greater than that of all the other forms of soft-tissue sarcoma combined. Although the responsiveness of rhabdomyosarcoma to chemotherapy varies greatly among tumors with different primary sites and with different histologic features, the overall results of chemotherapy have been more impressive in rhabdomyosarcoma than in any other form of soft-tissue sarcoma in childhood. On the other hand, rhabdomyosarcoma occurring in the adulthood needs further assessment to know which factors influence its outcome.

Methods: Twenty patients diagnosed with intramuscular rhabdomyosarcoma were treated between 1987 to 2008. They constitute 0.8 % of soft-tissue tumors (Benign and Malignant) and 3.3% of malignant soft-tissue tumors at our files. They were 13 males and 7 females, their age ranged from one year to 72 years with an average of 26 years. Six patients were aged less than ten years while 14 above this age. They were followed up from 3 months to 20 years with an average of 53 months. The site of the tumors were as follow, two in the back, nine in the upper limb, eight in the lower limb, and one in the abdominal wall. Eleven patients were presented with localized disease (M0N0), five patients had distant metastasis without nodal involvement (M1N0), three patients had nodal involvement without distant metastasis (M0N1), and one patient was presented with both distant metastasis and nodal metastasis (M1N1).

Results: All nine patients who presented with metastasis died of disease (DOD), while six of the eleven patients presented with a localized disease died of disease (DOD), and the remaining five patients are alive. Among the long survivors (Five patients), four were aged below 10 years and four of them also had had an embryonal rhabdomyosarcoma, and all of them had a wide margin surgical procedure. The six patients who presented with a localized disease and died of disease, are aged above 10 years, five had embryonal rhabdomyosarcoma, five had preoperative chemotherapy and radiotherapy, four had a wide margin surgical procedure and two had interalesional margins where all had postoperative chemotherapy and three had postoperative radiotherapy. Local recurrence developed in four of the seventeen surgically treated patients. Ten patients had wide margins, one of them recurred, three had marginal margin, and one of them recurred, where four had interalesional margins, two of them recurred. At the end of follow up, the survival rate of patients was as follow 64 % for M0N0, 0% for M1N0,M0N1 and M1N1, 66% for patients aged younger than 10 years, and 25 % for all the twenty patients.

Conclusion: The age of the patient, the status at presentation are the most influencing factors regarding the prognosis followed by the surgical margin. The histological subtype, the use of chemotherapy or radiotherapy did not influence the prognosis of our adult patients.