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TREATMENT MODALITIES AND FOLLOW UP OF CHILDREN WITH PERIFERAL NERVE SHEET TUMOR 15 YEARS SINGLE INSTITUTION EXPERIENCE

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Background : The objective of the study was to analyze the treatment modalities in cases of malignant peripheral nerve sheet tumor (MPNST) in children treated during a 15 year period in the Pediatric Department of the Institute for Oncology and Radiology,Serbia.
Methods: During the period of 1996-2011 there were 9 children (6 male and 3 female) with a median age of 14 years (age range of 4years to 17 years) treated using a multimodal therapeutic approach that included surgery, chemotherapy and radiotherapy. The patients presented with primary tumor sites as follows: 5 on the extremities, 1 in the sacrum, 2 in the head and neck and one localized in the shoulder. The majority of the t patients (8 out of the 9 patients) had large tumors (>5cm). Two of the patients were affected by neurofibromatosis 1.
Results: Treatment commenced in 5 patients with a gross total tumor resection and they were classified as IRS group I (hisologicaly free margins found on biopsy). Another 4 patients were classified as IRS group III and surgery was delayed til after a neoadjuvant chemotherapy treatment (of between 3 and 5 cycles).Two patients had amputations and 2 conservative resections with macroscopic residual tumor.
Chemotherapy was administered to all patients, neodjuvant in 4 of the patients. A response to chemotherapy was seen in 2 patients in the IRS group III and 2 patients experienced a progression of disease.
The chemotherapy regimens used were VACA in 4 patients, CEVAIE in 3 patients, 1 patient was treated according to EE99 protocol and 1 according to EpSSG NRSTS 2005 protocol.
Seven of the patients had radiotherapy concomitantly with the chemotherapy. The total dose ranged from 55 to 60 Gy.
Eight of the patients are now in the follow-up period with no evidence of disease for between 1 and 14 years.
Conclusion: Multimodal treatment is effective in children with MPNST. According to our study the time of diagnosis, IRS group, site and absence of metastasis are important predictors of survival.

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