Case Report

Complete Remission of Locally Recurrent MPNST after Hypofractionated Image-Guided Radiotherapy-A Case Report

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Abstract

Background: Malignant peripheral nerve sheath tumors (MPNST) are a rare disease. The role of radiotherapy for inoperable tumors is unclear.

Case report: A 85 year old male with a symptomatic inoperable local recurrence of MPNST in the retromaxillary region 5 years after surgery was treated with hypofractionated image-guided radiotherapy. The clinical target volume of radiotherapy included the macroscopic tumor (25cm³) with a small safety margin of 2mm. The total dose was 48Gy administered in 12 fractions on consecutive working days. The patient tolerated treatment well without any side effects. One year after treatment, there was maintained complete remission on follow-up CT examination without late radiation sequelae.

Conclusion: Hypofractionated image-guided radiotherapy may have a curative potential as primary treatment in selected small to medium-sized MPNST.

INTRODUCTION

A malignant peripheral nerve sheath tumor (MPNST) is a rare subtype of sarcoma. The clinical presentation varies [1 -6]. The prognosis is worse as compared to other sarcomas. Current treatment recommendations for sarcomas are mainly based on surgical resection as main part of therapy with adjuvant or neoadjuvant radiotherapy or chemotherapy in presence of risk factors. In inoperable sarcomas, definitive radiotherapy may result in long-term cure in a subset of patients. However, there are currently no specific guidelines for the treatment of MPNST; treatment recommendations refer to the treatment of other sarcomas. Radiotherapy may play a role as adjuvant treatment in MPNST, but there are no data whether or not radiotherapy as sole treatment may have a curative potential. We report a case of recurrent MPNST treated with hypofractionated high image-guided radiotherapy.

CASE REPORT

An 85 year old Caucasian male had undergone transfacial tumor resection of a retromaxillary mass in 2010 at the age of 79 years. Histology revealed a completely resected malignant peripheral nerve sheath tumor. Adjuvant treatment was not administered. He developed a symptomatic local recurrence about 5.5 years later; the recurrent tumor was located in the retromaxillary area with direct contact to the mandible and was considered as inoperable (Figure 1). The patient’s general condition had meanwhile deteriorated. He had an implanted cardiac device; MR-imaging was therefore not performed. The tumor board decision was to offer palliative radiotherapy.

The clinical target volume of radiotherapy included the macroscopic tumor with a small safety margin of 2mm. The total dose was 48Gy administered in 12 fractions on consecutive working days. The patient tolerated treatment well without any side effects. One year after treatment, there was maintained complete remission on follow-up CT examination without late radiation sequelae.

DISCUSSION

Prospective studies on MPNST as sole entity are lacking; earlier data on MPNST mostly derived from single institutional retrospective series. Recently, multi-institutional retrospective studies, analyses of SEER-data and subgroup analyses of clinical studies in sarcomas which often comprise a small number of MPNST have increased knowledge on outcome and prognostic factors [1-7]. In 1986, a single-institution review of 120 cases with MPNST treated over a period of 71 years at...
the Mayo Clinic reported a 5-year overall survival rate of 34% [8]. 52% of the cases were associated with Recklinghausen’s disease (neurofibromatosis type 1, NF 1); patients with NF 1 were younger as compared to patients without NF 1 (median age 28.7 versus 39.7 years) and had a worse survival (5-year OS 16% versus 53%). 23 out of 120 cases were located in the head and neck region; 11% were considered as radiation-induced with a mean latency period of about 17 years. 62% of the patients were treated with complete tumor resection, 49% received radiotherapy and 21% chemotherapy. Local recurrence occurred in 45% of NF1-patients and 38% of non-NF1-patients after a median interval of 13.3 months. There was no association between histological patterns (such as grade or mitotic index) and outcome. The only prognostic factor was complete resection. A recent multi-institutional series with 179 cases from Norway, Sweden and Italy treated in the period of 1970 through 2011 reported comparable results with a 5-year survival of about 40% [9]. MPNST has been reported to be the most frequent cause of death in patients with neurofibromatosis [2].

The largest prospective series has been reported by the Italian and German pediatric sarcoma studies and comprises 167 pediatric and adolescent patients treated in a variety of studies between 1975 and 1998 [7]. The results suggest that surgery is the mainstay of treatment and that postoperative radiotherapy may improve local control in patients with minimal residual tumor after surgery. Other reports support a beneficial effect of radiotherapy [7,10,11], and radiotherapy is frequently used as adjuvant treatment [5,6,12]. Nevertheless, surgery is considered as standard and some authors recommend even aggressive mutilating surgery [13-17].

There is currently few if any evidence on radiotherapy as sole local treatment in malignant peripheral nerve sheath tumors. Our case report suggests that radiotherapy, especially if administered with contemporary high precision irradiation techniques have, may have a curative potential in MPNST. Modern radiotherapy techniques (e.g. image-guided radiotherapy) allow smooth treatment for small to medium sized tumors. In our case, the target volume of radiotherapy as restricted to the tumor itself with a small safety margin. Such treatments are well tolerated with minimal side effects and long-term sequelae, as was observed in our case. The fractionation regimen (48Gy in 12 fractions in an overall treatment time of less than three weeks) is biologically equivalent to a total dose of 70Gy or more in conventional fractionation (with single doses of 1.80Gy to 2.00Gy) over about 7 weeks.

Chemotherapy is frequently used for palliation and as neoadjuvant treatment for locally advanced tumors but its impact is not well defined [3,18]. Further future options may derive from targeted therapies. A variety of potential target structures have been recently identified [12,15,18,19].

In summary, this case report suggests that radiotherapy, especially if administered with modern techniques, can be effective and may have a curative potential as primary treatment in selected small to medium-sized primary or recurrent MPNST. The role of hypo fractionation requires further investigation.

REFERENCES


