Radiotherapy and Oncology 106 (2013) 181–185

Contents lists available at SciVerse ScienceDirect

Radiotherapy and Oncology

journal homepage: www.thegreenjournal.com

Ependymoma

Spinal cord ependymoma in children – Results of postoperative radiotherapy

Katarzyna Pędziwiatr *, Anna Skowrońska-Gardas, Marzanna Chojnacka

Department of Radiotherapy, M. Skłodowska-Curie Memorial Cancer Center – Institute, Warsaw, Poland

ARTICLE INFO

Article history:
Received 19 October 2012
Received in revised form 12 February 2013
Accepted 13 February 2013
Available online 13 March 2013

Keywords:
Ependymoma
Spinal cord
Children
Radiotherapy

ABSTRACT

Purpose: A retrospective study was performed to evaluate the results of postoperative radiation therapy of spinal cord ependymoma in children.
Methods and materials: Between 1984 and 2005, 28 children with spinal cord ependymoma were treated with radiotherapy, after surgery and in three cases after chemotherapy as well. Median age at diagnosis was 13.3 years (range from 4.7 to 16.2 years). Ependymoma myxopapillare was identified in 13, ependymoma in 12 and anaplastic ependymoma in 3 cases.
Results: With a median follow-up of 8.7 years (range from 3 to 25 years) 22 patients were alive. The overall survival rate of 2, 5 and 10 years was 93%, 85% and 77% respectively, whereas progression free survival rate was 82%, 74% and 74% respectively. Patients with myxopapillary ependymoma had significantly better 5-year overall survival rate 100% than those with other histopathological types 60% ($p = 0.016$). There were 2 relapse incidences observed among 13 patients with myxopapillary ependymoma, both underwent repeated surgery and reirradiation. In the group of 20 patients with gross total resection the overall 5-year survival rate was 100% in comparison with 62.5% with partial surgery, but it did not achieve statistical significance.
Conclusions: The histological type of ependymoma myxopapillary was a statistical significant favourable prognostic factor. The gross total resection with adjuvant radiotherapy allows obtaining a high total survival rate.
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Primary neoplasms of the spinal cord occur rarely, constituting only 5–15% of all the tumours of the central nervous system. Ependymomas are diagnosed second after meningiomas and constitute about 23% of spinal cord tumours in adult patients [1]. In paediatric patients under 14 years of age, about half of all the tumours, localised in the spinal cord constitute astrocytomas, with ependymomas accounting for 15% [2]. Reports of spinal cord tumours in paediatric patients are rare, more frequent being those presenting collective results of the therapy in both adults and paediatric patients [3]. Furthermore, there is lack of unanimity in therapeutic procedures, in particular those regarding radiotherapy and chemotherapy subsequent to a primary surgical treatment. Various authors present diverse opinions [4].

In particular, the issue that remains the most controversial is the adjuvant radiotherapy in ependymoma myxopapillare, both in adults and in paediatric patients [4]. Owing to the infrequent occurrences of spinal cord ependymomas in paediatric patients, the purpose of this analysis is to evaluate the clinical course, the administered treatment and its results observed in a group of 28 paediatric patients diagnosed and treated in the Department of Radiation Oncology of the Cancer Center and Institute of Oncology in Warsaw, between 1984 and 2005.

Materials and methods

Between 1984 and 2005 in the Department of Radiation Oncology of Maria Skłodowska-Curie Memorial Cancer Center and Institute of Oncology in Warsaw, 28 patients with the spinal cord tumours were treated, which constituted 19.6% of all paediatric patients with ependymoma. The age ranged from 4.7 to 16.2 years, the median age was 13.3 years. In 17 (60.7%) patients the tumour was located in the cauda equina region, in 8 patients (28.6%) in other parts of the spinal cord, while in yet 3 other cases (10.7%) the multifocal process was diagnosed. In the majority of cases, the symptoms of disease included pains, limb paresis or paralysis, dysfunction of sphincters and, less frequently, sensory disturbances. The symptoms lasted longer in the spinal than in the typical intracranial localisation, and sustained in 12 patients less than 3 months, in 11 from 3 months to one year, and in 5 – a year or longer. In 13 of them (46.4%), the neurological condition was good or slightly impaired, while 15 patients (53.6%) sustained grave neurological disorders.

Histological diagnosis of ependymoma myxopapillare type was found in 13 patients (46.4%), in 3 cases (10.7%) anaplastic ependy-

* Corresponding author. Address: Department of Radiotherapy, M.Skłodowska-Curie Memorial Cancer Center – Institute, Wawelska 15, 02-034 Warsaw, Poland.
E-mail addresses: 3kasia@wp.pl (K. Pędziwiatr), agardas@coi.pl (A. Skowrońska-Gardas), marzanna.ch1@wp.pl (M. Chojnacka).

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http://dx.doi.org/10.1016/j.radonc.2013.02.007

moma occurred, while in the remaining 12 the classical type of ependymoma was diagnosed.

Gross total resection was conducted in 20 patients (71.5%), in 2 (7.1%) subtotal resection, in 4 (14.3%) partial and in 2 (7.1%) only biopsy.

3 patients in the postoperative period received chemotherapy: in one due to the multifocal, inoperable tumour, and in 2 due to the traces of anaplasia in histopathological diagnosis. The following medications were administered: iphosphamide, adriamycin, VP-16, carboplatin, according to the treatment regimen of malignant glioma [5].

Radiotherapy was applied in 26 (92.9%) patients as postoperative adjuvant treatment and in 2 (7.1%) patients with ependymoma GII after the second resection due to the recurrence. In 21 children (75.0%) only the area of the tumour was irradiated, with the margin of uninvolved tissues, whereas in 3 (10.7%) radiotherapy was administered to the whole spinal cord with boost to the tumour. One child (3.6%) with anaplastic tumour received total irradiation on the spinal cord and posterior cranial fossa. Three patients treated between 1984 and 1988 received a whole CNS radiotherapy, in accordance with the currently applicable procedure of treatment. Elective doses ranged between 30 and 36 Gy, the dose administered to the tumour varied depending on its localisation, the length of the irradiated spinal section and the age of the patient, and ranged from 43.4 Gy to 55.5 Gy (median 49.5 Gy).

By 1997 the adopted technique was 2D radiotherapy planning, with 1–2 straight fields and 3 oblique ones. Since 1997 the conformal radiotherapy with 3D planning has been adopted. Characteristics of the patients were presented in Table 1.

Table 1
Characteristics of patients.

Feature	Description	No. of patients	%
Age	4,7 – 16,2 (median 13,3)	28	100
Sex	Male	15	54
	Female	13	46
Clinical symptoms	Pain	19	68
	Extremity weakness/paresis	17	61
	Sensory disturbances (paraesthesia)	2	7
	Urinary/faecal incontinence	9	32
	Signs of increased intracranial pressure	2	7
	Without symptoms	1	4
Duration of symptoms	<3 months	12	43
	3–12 months	11	39
	>12 months	5	18
LQS neurological scale	I	6	21
	II	7	25
	III	8	29
	IV	7	25
	V	0	0
Histology	Myxopapillary ependymoma	13	46
	Ependymoma	12	43
	Anaplastic ependymoma	3	11
Site	Upper spinal cord	8	29
	Cauda equina	17	61
	Primary multifocal	3	11
Type of surgery	Total resection	20	71
	Subtotal	2	7
	Partial	4	14
	Biopsy	2	7
Radiotherapy field	Local	21	75
	Whole spinal cord + local boost	3	11
	Posterior fossa and spinal cord + local boost	1	4
	Craniospinal + local boost	3	11
Radiotherapy technique	2D years 1984–1996	16	57
	3D years 1997–2005	12	43

Statistical methods

The retrospective analysis of 28 patients treated between 1984 and 2005 has been conducted. The information on survival rate and present health status was collected on the basis of medical documentation of the Children's Memorial Health Institute, information obtained by means of letters and phone calls from parents or already adult patients, follow-up examination in Department of Radiation Oncology and from the files of Warsaw Cancer Registry. The observation ceased at the end of 2010.

For the purpose of describing the material and the analysis of post-treatment complications, the standard methods of descriptive statistics were applied, frequency tables for categorical variables and the mean, as well as the standard deviation or median and quadrille for continuous variables.

The criterion for the treatment efficacy was the survival rate from the beginning of the oncological treatment until the death or the last information that the patient was alive, with the time of living without disease progression. In order to analyse the time of survival, the survival curves were calculated by means of the Kaplan–Meier method. Due to the small number of analysed patients, the univariate method was applied.

Results

At the end of the observation, out of the group of 28 paediatric patients, 22 were alive, with the observation period ranging from 3 to 25 years (median 8.7 years). 6 patients died within the period ranging from 3 months to 14 years after the end of the radiotherapy (median 5.3 years). In 5 patients death resulted from primary disease: failure to cure (1), localised recurrence (2), and recurrence with associated dissemination (3). In any of them there was no confirmation of dissemination outside the CNS. In one case, the death could not be accounted for (the parents informed about the death of a 22-year-old daughter without providing any reason for that, as she died without traces of the primary disease 14 years after having completed the treatment). This woman had a tumour located in the lumbar spine (Th12-L3).

The overall survival rate of 2, 5 and 10 years was 93%, 85% and 77%, respectively, whereas the progression free survival rate was 82%, 74% and 74%, respectively. Figs. 1 and 2 present the curves of the overall survival rate and progression free survival.

In the group of 20 patients with gross total resection, the overall 5-year survival rate was 100% (one patient died 14 years after treatment due to other reasons), within 8 cases after partial surgery, 5 children (62.5%) survived more than 5-years. Similarly, a 5-year survival without disease progression was 95% and 42.9%, respectively. Due to a small number of cases, the data did not reach statistical significance.

The diagnosis of ependymoma myxopapillare was connected with a significantly more favourable prognosis: a total 5-year survival was 100%, in comparison with 60% for other histopathological cases ($p = 0.016$). However, in this group, 2 patients suffered the recurrence, in one case it was localised next to the border of the irradiation field, while in the other it was inside the irradiation area in a child with primary multifocal disease. Both of them underwent the second line of treatment with the repeated surgical procedure and radiotherapy. The first child lived for eight years without evidence of disease. The second patient lived with recurrence of the disease, for two years after retreatment.

No statistically significant difference was observed in the survival rate with recurrence depending on the histopathological