Introduction

Background: Myeloproliferative ependymoma (MPE) is a rare tumor accounting for 10-20% of pediatric intramedullary spinal cord tumors.

Management of children with MPE is controversial, typically involving gross total resection (GTR), with or without adjuvant therapy, and followed by spinal radiation (SRT) & recurrent tumors. The role of craniospinal irradiation (CSI) is not well studied, but may aid in developing brain tumors in neurocognitive deficits. Reports of pediatric patients with MPE are limited to small series & individual case studies. Treatment strategies & recommendations in children with MPE are not evidence-based and extrapolated from adult studies.

Objective: To determine optimal RT fields in pediatric MPE, specifically whether CSI is needed in the presence of tumor dissemination.

Incidence of Intracranial Dissemination

6.5% (2/33 patients from TXCCC & meta-analysis)

- 0/8 patients from TXCCC & 2/23 patients from meta-analysis had intracranial dissemination at initial diagnosis, 0/2 in histopathology & 0/2 in sequencing. Both received CSI & both did not recur in the brain (Case 10: 2012)

- 1 patient initially had disseminated spine disease & received local RT, developed a new intracranial relapse in asymptomatic 4th & 6th ventricles. He further received CSI, remained progression-free for 4 years & a median follow-up of 4.1 years (Range: 0.6-30 years).

- 5/23 (21.7%) of children received CSI in the absence of intracranial dissemination at initial diagnosis.

Abstract

Purpose: Myeloproliferative ependymoma (MPE) accounts for 10-20% of pediatric intramedullary spinal cord tumors. Management of children with this rare tumor is controversial. Treatment recommendations in children are not evidence-based but extrapolated from adult studies and are not validated in pediatric cases. We performed a meta-analysis of patients with pediatric MPE reported in the PubMed database. Results: In our study, 13 of 16 patients (81.2%) had intracranial dissemination confirmed by imaging studies. Fourteen underwent CSI & 8 received SRT, but both did not recur in the brain. Five of 23 (21.7%) children received CSI in the absence of intracranial dissemination at initial diagnosis. Two of 3 children (60.0%) had intracranial dissemination at initial diagnosis & received CSI. Conclusion: This is the largest comprehensive analysis of RT field in pediatric MPE. The pattern of dissemination suggests that the brain can be excluded from the RT field in the absence of intracranial dissemination, which is particularly important in young children with more than 10 years of age to avoid neurocognitive sequelae of CSI.

Description of study

TXCCC series: Consecutive series of 8 patients over a period of 15 years from 1999 to 2009 identified. Outcome in relation to RT fields & disease studied.

Meta-Analysis: PubMed database used to identify case series of pediatric MPE. A PubMed search of English language articles from 1997 to 2012 performed using a combination of the keywords which included MPE, children, & pediatric.

Inclusion Criteria: Presence of MPE in the spine with or without brain involvement, histologic diagnosis of MPE, age of diagnosis between 0 to 18 years old, & treatment information (including dose & field when RT is used).

Cases identified for meta-analysis: 23 cases of pediatric MPE. From a total of 27 included the inclusion criteria. 20/23 received RT either at diagnosis or recurrence; 3 cases received RT only. Dissemination was defined by presence of more than 1 tumor nodules or diffuse central nervous system involvement.

Results

Table 1. Comparison of patterns of dissemination between TXCCC series & meta-analysis

<table>
<thead>
<tr>
<th>Time Period</th>
<th>TXCCC</th>
<th>Meta-Analysis</th>
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</thead>
<tbody>
<tr>
<td>2012</td>
<td>1/16</td>
<td>1/23</td>
</tr>
<tr>
<td>2009</td>
<td>2/8</td>
<td>2/15</td>
</tr>
<tr>
<td>2000</td>
<td>0/7</td>
<td>1/11</td>
</tr>
<tr>
<td>1999</td>
<td>0/6</td>
<td>1/11</td>
</tr>
</tbody>
</table>

Table 2. Radiation field and dose in relation to site of recurrence and outcome of pediatric MPE from TXCCC.

Table 3. Meta-analysis of 23 pediatric MPE treated with radiotherapy from 6 reported series.12

Conclusions

(1) This is the largest comprehensive analysis of RT field & dose in pediatric MPE, providing evidence-based recommendation for this disease in children.

(2) We provide evidence that the brain can be excluded from the RT field in the absence of intracranial dissemination, which will spare neurocognitive sequelae of irradiating the developing brain particularly in young children less than 10 years of age.

(3) We show that the incidence of spinal dissemination is higher in children (6.5%) compared to adults. Majority of disseminated tumors (86.7%) remained stable with additional RT after surgery.

(4) We would recommend that children with disseminated MPE receive RT regardless of the degree of resection & a screening MRI brain performed at diagnosis as it can change the RT field.

References


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