Sacroccygeal malignant germ cell tumor (SC-MGCT) with intraspinal extension

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Abstract

Background: Neurological involvement due to intraspinal extension in sacroccygeal malignant germ cell tumors (SC-MGCTs) has rarely been reported.

Aim: To evaluate the incidence, presentation, management and the outcome of patients of SC-MGCT with intraspinal extension.

Materials and Methods: Case records of all cases of SC-MGCT from 2001 to 2008, were reviewed to identify cases with vertebral involvement and intraspinal extension. They were evaluated in terms of their presentation, response to therapy, extent of surgical resection, recovery of neurological symptoms and outcome.

Results: Of the 31 cases of SC-MGCT, 5 (16%) had intraspinal extension. Age ranged from 12 to 84 months (median 24 months). Four patients had Altman type 4 disease (stage 4) and 1 had Altman type 3 (stage 3) disease. The intraspinal extension in all patients was detected on contrast CT scan. Patients presented with neurological symptoms in the form of lower limb paresis (80%), bowel and bladder (20%) incontinence. All the tumors responded to pre-operative chemotherapy. Gross complete local resection could be achieved in 4 (80%). Neurological recovery was complete in all except for persisting neurogenic bladder in one. During follow up of 3–32 months, all were alive with no recurrence.

Conclusions: SC-MGCT presenting with neurological deficits due to intraspinal extension is usually advanced disease. These patients respond to chemotherapy and surgical resection and most have complete neurological improvement.

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range of 12–84 months (median 24 months). Four (80%) patients presented with Altman type 4 disease while one (20%) patient had Altman type 3 disease. Patients with Altman type 4 disease had stage 4 disease with bilateral lung metastasis while the one with Altman type 3 disease had stage 3 disease. While 1 (20%) had no neurological symptoms, the other 4 (80%) had severe bilateral lower limb weakness and 1 (20%), in addition, had urinary and fecal incontinence. All patients received 3 cycles of neoadjuvant chemotherapy (Table 1). All the tumors responded to neoadjuvant chemotherapy with the pulmonary metastasis resolving completely and primary tumor shrinking in size and the intraspinal extension resolving completely (except in one). Complete surgical resection of the residual sacrococcygeal mass with coccyx was possible in 4 (80%). One patient (20%), who had some intraspinal residual disease after the 3 courses of neoadjuvant chemotherapy (Picture 1), underwent partial sacral (S3,4,5) excision. However, there was still gross residue remaining in sacrum in this patient. Following resection, all patients received 3 additional cycles of adjuvant chemotherapy (Table 1). Pathology revealed endodermal sinus tumors in all patients. The lower limb paresis in all 4 patients who presented with paresis recovered completely. The lone patient with fecal and urinary incontinence showed resolution of fecal incontinence while the urinary incontinence persisted. This patient is dry on clean intermittent catheterization (CIC). The follow up has ranged from 3 to 32 months (median 24 months). All are alive with no recurrence.

3. Discussion

Intraspinal extension by SC-MGCT has never been the primary focus of a report. All reported cases of sacrococcygeal teratoma with intraspinal extension have been either mature or immature teratomas [3,8–10]. Further none of these reported patients with intraspinal extension had any neurological symptoms of bowel or bladder incontinence or lower limb paresis. All except one were operated in the neonatal period where the intraspinal extension was also resected. One patient with neonatal SCT recurred in adulthood and during this recurrence there was an intraspinal extension that was resected [3]. The cases in the present series were all endodermal sinus tumors and had neurological symptoms in the form of bowel or bladder incontinence or lower limb paresis. All these presented late beyond infancy and underwent surgical resection at an older age.

Table 1
Patient characteristics, extent of disease and treatment details of patients. POG: Pediatric Oncology Group.

<table>
<thead>
<tr>
<th>Age (months)</th>
<th>Altman type</th>
<th>POG Stage</th>
<th>Neurological symptoms at presentation</th>
<th>Chemotherapy courses</th>
<th>Gross residue at surgery</th>
<th>Residual neurological deficit</th>
<th>Follow up (months)</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Lower Limb Paresis</td>
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<td>Neoadjuvant</td>
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</tbody>
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Neoadjuvant chemotherapy achieved good response in terms of reduction in the tumor size, resolution of intraspinal component and also the metastatic disease in most patients. This renders the tumor easily resectable without the need for spinal exploration or vertebral resection. The resolution of neurological symptoms on neoadjuvant chemotherapy suggests that the symptoms were because of compression of the spinal nerves with intraspinal component rather than invasion or destruction of the nerves. The spinal exploration with partial vertebral resection was required in only one patient (20%) in the present report and this too could not achieve complete resection. However, this patient too showed complete resolution of the residual mass and also of neurological symptoms after adjuvant chemotherapy and did not have any recurrence in the follow-up during the interval. This report supports the need for neoadjuvant chemotherapy for all such patients of SC-MGCT with intraspinal extension. Coccygectomy should always be performed, even in Altman type 4 patients, to reduce the recurrence rate.

Patients of SC-MGCT with intraspinal extension with neurological symptoms have good prognosis with chemotherapy and surgery. Neoadjuvant chemotherapy obviates the need for extensive intraspinal exploration in most patients. Even patients with residual neurological symptoms can be expected to lead a near normal life with the currently available medical care.

References