ORIGINAL ARTICLE

# Primary spinal tumors in childhood: A single institution 15 year experience

Po-Cheng Hung, Chieh-Tsai Wu[1], Tang-Her Jaing[2], Jiunn-Ming Sheen[3], Ming-Liang Chou, Kuang-Lin Lin

Department of Pediatrics, Division of Neurology, Chang Gung Children's Hospital, [1]Department of Surgery, Division of Neurosurgery, Chang Gung Memorial Hospital, [2]Department of Pediatrics, Divisions of Hematology and Oncology, Chang Gung Children's Hospital, [3]Department of Pediatrics, Chang Gung Memorial Hospital-Kaohsiung Medical Center, Chang Gung University, Taoyuan, Taiwan

## ABSTRACT

**Background:** Pediatric primary spinal tumors (PST) are fairly uncommon, with little available data regarding incidence and outcomes.

**Materials and Methods:** We conducted a retrospective review of the 22 consecutive patients less than 18 years old diagnosed with PST between March 1997 and May 2011 and treated at Chang Gung Children Hospital. All patients had undergone magnetic resonance imaging (MRI) for pre-operative evaluation and operations for PST. The extent of tumor removal was assessed by surgical report by the neurosurgeon or by post-operative MRI if available.

**Results:** Ten of them had intramedullary tumors and 12 had intradural extramedullary tumors. All patients were treated with surgery to the primary site. A total of 15 patients underwent gross total tumor resection and seven patients received post-operative radiotherapy. Five patients received adjuvant chemotherapy for their primary tumor. Fourteen patients (64%) survived from study entry without tumor progression.

**Conclusions:** PST encompassed a diverse group of pathologic entities that differ markedly based on the location and age of the children. Total resection of pediatric PST in children could be performed with acceptable risk and satisfactory long-term results.

**Key words:** Children, histology, primary spinal tumors, surgical resection

## Introduction

Primary spinal tumors (PST) should be considered in cases of a solitary spinal lesion. A wide variety of primary neoplasms can involve the spine. Spinal tumors can be classified according to their tissue of origin.[1] Patients' symptoms may be insidious and non-specific.[2] They are either intradural, intramedullary, or intradural extramedullary.[3] PST in children remains a vexing clinical problem as its existence is often unrecognized until occurrence of spinal cord compression.

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**Address for correspondence:**
Dr. Tang-Her Jaing, Department of Pediatrics, Division of Hematology and Oncology, Chang Gung Children's Hospital, Chang Gung University, 5 Fu-Shin Street, Kwei-Shan 33305, Taoyuan, Taiwan. E-mail: jaing001@cgmh.org.tw

Due to some sufferers who may present repeatedly to physicians before an unknown diagnosis is made, spinal imaging is usually instigated once the patient has developed neurological deficits. We highlighted the study not only for the clinical presentation, diagnosis, and management of these children, but to heighten the clinician's awareness of this rare group of tumors.

## Materials and Methods

All patients with PST surgically treated at Chang Gung Children's Hospital between March 1997 and March 2012 were available for retrospective analysis and approved by the Institutional Review Board. All patients had undergone magnetic resonance imaging (MRI) for pre-operative localization and focused operations. The extent of macroscopic removal was assessed by surgical report, by the neurosurgeon, and by post-operative MRI. The patients' data on demographic and clinical characteristics, treatments, duration of symptoms prior to diagnosis, location of tumor, degree of resection, and clinical outcomes were extracted. In cases of diagnostic uncertainty, images were reviewed by a second board-certified neuroradiologist to assure unbiased interpretation.

Gross total resection (GTR) was attempted in all cases. Sensory and motor evoked potentials were used to

monitor all cases. Resection of the tumor by multi-level laminectomy or laminoplasty was performed in all cases. For hemangioblastomas and small- to moderately-sized ependymomas, the tumors were resected using an outside-in technique, with *en bloc* removal when identification of tumor-spinal cord transition plane was clear.

The primary outcome measured was survival. Patients were considered to have an event at time of death from any cause; survivors were censored at last follow-up. Research ethics board approval was obtained, with patient consent not required for this retrospective study with de-identified subjects. All patients had a pathologic diagnosis and were grossly evaluated for focal hemorrhage, cystic change, and myxoid change. No personal identifiers were included in the electronic database.

## Results

### Patient population

We retrospectively reviewed the records of 22 consecutive pediatric patients seen at a single institution from 1997 to present patients newly diagnosed with PST. There were 12 boys and 10 girls, with a male-to-female ratio of 1.2:1. The median age at diagnosis was 12.2 years, with a range of 2 weeks up to 17.9 years. The median duration of symptoms before diagnosis was 2 months (0.5-48). All patients were followed-up for a median of 80 months. The most common clinical presentation was limb weakness (52%) followed by back pain (29%) [Table 1]. Location of the tumor was intradural, intramedullary in 10 patients, and intradural extramedullary in 12 patients. Intramedullary tumors comprised predominantly of gliomas (infiltrative astrocytomas and ependymomas). There were no patients with neurofibromatosis type 2 in this series.

Seven tumors were located primarily in the cervical cord (2 hemangiomas, 2 neurofibromas, 1 ependymoma, 1 Langerhans cell histiocytosis, and 1 astrocytoma), 10 were predominantly thoracic (3 astrocytomas, 2 mesenchymal chondrosarcomas, 4 neurilemomas, 1 hemangioma), 4 lumbar (2 myxopapillary ependymoma, 1 teratoma, 1 primitive neuroectodermal tumor (PNET)), and 1 sacral teratoma.

**Table 1: Frequency of symptoms of primary spinal cord tumors of childhood**

| Clinical presentation | No. of patients | % |
|---|---|---|
| Extremity pain/weakness | 12 | 55 |
| Back pain | 6 | 27 |
| Neck pain/stiffness | 5 | 23 |
| Urinary/GI dysfunction | 2 | 9 |
| Spinal deformity | 1 | 5 |
| Tetraplegia | 1 | 5 |
| Sensory abnormality | 1 | 5 |

GI – Gastrointestinal

### Surgical results

All patients underwent laminectomy for removal of tumor. GTR of tumors was achieved in 15 of 22 patients (68%), subtotal resection in 2 patients (9%), partial resection in 3 patients (14%), and biopsy in 2 patients (9%). A GTR was more frequently achieved in cases of teratoma, mesecnhymal chondrosarcoma, and neurillemoma. An example of GTR is shown in Figure 1. Table 2 shows the location and extent of tumors, and their pathologic classification.

### Complications

There were no operative deaths. One patient with astrocytoma succumbed to tumor progression and one with plexiform neurofibroma subsequently died of malignant peripheral nerve sheath tumor, although chemotherapy and radiotherapy had been attempted (patients 6, 13).

### Post-operative adjuvant therapy

Most patients (59%) did not receive post-operative radiotherapy or chemotherapy. Seven patients received adjunctive spinal irradiation as part of the initial treatment, with doses ranging from 4600 to 6120 cGy. Five patients received adjuvant chemotherapy for their primary tumor. Salvage chemotherapy was administered after radiotherapy in two patients.

Five patients (patients 8, 9, 10, 13, 22) were treated with six courses of cisplatin and etoposide. One patient (patient 22) developed lung metastasis 2 years after the diagnosis of PNET of the sacrum and died of progressive disease despite receiving high dose chemotherapy with autologous stem cell rescue. At the time of analysis, the median progression-free survival in these patients was 105 months, and the median survival was 99 months.

## Discussion

Owing to the relative rarity of PST, optimal therapy has yet to be determined.[4,5] Extramedullary intradural tumors are the most common spinal cord tumors in adult,[6] whereas intramedullary tumors constitute about 35% of the spinal tumors found in children.[7,8] Low-grade gliomas such as ependymomas and

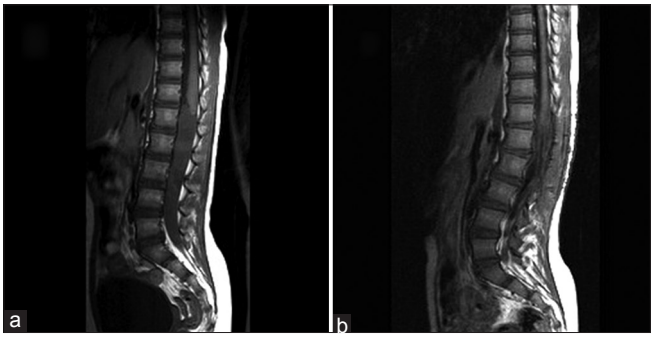


**Figure 1:** (a) Sagittal T1-weighted Gd-enhanced magnetic resonance images of myxopapillary ependymoma obtained preoperatively; (b) 48 h after gross total removal

**Table 2: Information about patients enrolled in the study**

| Patient | Age at diagnosis | Gender | Symptom duration (months) | Chief complaint and pertinent physical findings | % | Pathology diagnosis | Treatment | Progressive disease | Survival duration (in months) |
|---|---|---|---|---|---|---|---|---|---|
| 1 | 9 years 4 months | F | 1 | Upper back pain, neck stiffness | C7-T1 | LCH | B | No | 69 |
| 2 | 16 years 2 months | F | 6 | Tetraplegia, LLE weakness | C1-C4 | Hemangioblastoma | PR | Yes | 70 |
| 3 | 17 years 11 months | M | 0.5 | LLE weakness | C2-C4 | Hemangioma | GTR | No | 100 |
| 4 | 17 years 11 months | F | 3 | Left extremity weakness, neck pain | C2-T5 | Ependymoma | GTR, RT | No | 135 |
| 5 | 13 years 2 months | F | 1 | Back pain, bilateral LE weakness | T10-L1 | Grade II astrocytoma | GT | No | 97 |
| 6 | 14 years 5 months | F | 0.5 | LLE weakness | T4-T5 | Astrocytoma | PR, RT | Yes | 84 |
| 7 | At birth | F | 0.5 | Presacral mass | Sacrum | Immature teratoma | GTR | Yes | 77 |
| 8 | 10 years 7 months | M | 1 | LLE weakness | C7-T2 | Mesecnhymal chondrosarcoma | GTR, RT, CT | No | 104 |
| 9 | 1 years 10 months | F | 3 | Neck stiffness | T2-T6 | Grade II astrocytoma | B, CT | No | 37 |
| 10 | 10 months | M | <1 | Neck stiffness | C1-T8 | Astrocytoma | PR, CT | Yes | 67 |
| 11 | 2 years months | M | <1 | Back pain | T11-L2 | Hemangioma | STR | No | 82 |
| 12 | 8 years 6 months | F | 2 | Bilateral LE weakness | L1-L2 | Myxopapillary ependymoma | GTR | No | 36 |
| 13 | 2 years 10 months | M | 6 | RUE weakness | C1-C7 | Plexiform neurofibroma | GTR, RT, CT | Yes | 72 |
| 14 | 9 years 1 months | F | 10 | Back pain | L1-L4 | Mature teratoms | GTR | No | 163 |
| 15 | 13 years 11 months | M | 2 | Right wrist hypoesthesia | T8-T12 | Neurillemoma | GTR | No | 178 |
| 16 | 16 years 7 months | M | 9 | Back pain | T8 | Mesenchymal chondrosarcoma | GTR, RT | No | 180 |
| 17 | 15 years 11 months | M | <1 | Bilateral LE weakness | T8-T10 | Neurilemmoma | GTR | No | 147 |
| 18 | 12 years 10 months | M | 48 | Neck pain | C1-C2 | Neurofibromatosis | GTR | No | 105 |
| 19 | 11 years 10 months | M | 5 | Back pain | L1-L5 | Myxopapillary ependymoma | STR, RT | No | 126 |
| 20 | 17 years 4 months | F | 24 | RLE weakness | T10-T12 | Neurillemoma | GTR | No | 102 |
| 21 | 12 years 9 months | M | 2 | Bilateral LE weakness | T7-T10 | Neurillemoma | GTR | No | 114 |
| 22 | 9 years 11 months | M | 2 | RLE pain | L5-sacrum | PNET | GTR, RT, CT | Yes | 40 |

LLE – Left lower extremity; RUE – Right upper extremity; RLE – Right lower extremity; B – Biopsy; PR – Partial removal; STR – Subtotal removal; GTR – Gross total removal; RT – Radiotherapy; CT – Chemotherapy; PNET – Primitive neuroectodermal tumor; LCH – Langerhans cell histiocytosis

astrocytomas account for vast majority.[9] Intramedullary tumors infrequently progress slowly and for a long time often with rather mild symptoms and ill-defined pain.

PSTs produce symptoms due to compression of nerve root or cord, and ischemia vascular compression. The clinical symptoms are invariably non-specific. Limb weakness is often the leading symptom. Tethering of the cord by the dentate ligaments and filum terminale may result when expanding lesions oppose this resistance. The main symptoms are pain, weakness, sensory disturbance, and autonomic disturbances. In addition, there may be a vertebral deformity, especially in children.

Intramedullary tumors are predominantly intrinsic gliomas (astrocytomas and ependymomas). Spinal ependymomas are more common and often can be completely removed by separating the tumor from the spinal cord and when complete no further therapy is required. However, astrocytomas infiltrate the myelon and complete resection is rare. Intradural extramedullary tumors include schwannomas, neurofibromas, and meningiomas and are usually amenable to surgical resection.

The thoracic spine is more commonly involved than the lumbar or cervical spine. In our study, the most common clinical presentation is limb weakness which is usually splastic. The treatment of PST in children is primarily surgical.[10-12] A GTR was possible in 15 of our patients. Highly malignant aggressive neoplasms of PST are rare in children.[2,13] GTR should be the preferred treatment in symptomatic patients. Serial imaging is recommended to guide subsequent resection for tumor recurrence and stabilization of progressive spinal deformity.

## Conclusions

This is a retrospective study of a moderate size of pediatric PST with a wide range of follow-up time. Being a rare disease entity with a discrete pathology and symptomology, it is obviously premature to draw an astounding statistical conclusion from this study. As such, one must be vigilant to possible signs of

spinal cord compression. According to our analytical study, it is concluded that total resection of pediatric PST can be achieved with low incidence of neurological injury.

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