Management of Pediatric Spinal Tumors

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INTRODUCTION

Primary spinal tumors account for approximately 8% of all central nervous system neoplasms.6,17,24
Spinal tumors are especially challenging in children. Moreover, about 12% of spinal tumors arise during the first year of life.4
Pediatric spine tumors entail a diverse collection of pathologic diagnoses that differ markedly based on location and age of the child. Children can be affected both by primary and metastatic tumors, thus, making the differential diagnosis and management options extensive.3

Spinal tumors are potentially incapacitating. Clinical picture of spinal cord compression is the usual presentation in about 28-76% of cases.1,2,8,14
As the spinal cord is less tolerant to radiotherapy than the brain. Surgery is usually the first line option for these patients.16
Recent advances in surgical adjuncts and operative techniques give good opportunity for achieving safe surgery and good outcome.

PATIENTS AND METHODS

Patient Population
This retrospective study included twenty five pediatric patients diagnosed with spinal tumors, of which, fourteen were males and eleven females with a mean age of 3.8 years ranging from 1.5–16 years. The
patients were treated at Children’s Cancer Hospital Egypt (CCHE-57357) between July 2007 and June 2012.

Clinical assessment
Demographic data including patient sex and the age at presentation were collected. Clinical data including the presenting symptoms and signs, tumor pathology, surgical plan, operative findings and techniques utilized, complications and neurological outcome during the follow-up period were reported and analyzed.

Neuromaging
Series of magnetic resonance imaging (MRI) studies for the whole spine without and with Gadolinium contrast were performed routinely for each case: preoperatively, early post-operatively—within twenty four hours of surgery—to assess the extent of tumor resection, and then every three months for follow-up.

Initial MRI study of the brain was obtained as baseline to exclude concomitant cranial pathology. Computed Tomography (CT) study for the brain was obtained whenever hydrocephalus is suspected especially in cervical intramedullary spinal cord tumors (IMSCTs).

Surgical management
In tumors arising in cranio-cervical, cervical or cervico-dorsal locations, patient head was fixed in three pin head holder or gel-padded holder in order to keep a reasonable degree of stability and flexion.

Intraoperative neurophysiological monitoring was utilized in twenty cases. Somatosensory evoked potential (SSEP) and Motor evoked potential (MEP) including anal electromyogram (EMG) were utilized.

It was important to insert all needles after good disinfection of skin puncture sites especially perianal area. Also, good fixation of wires and electrodes with adhesive tapes is empirical to avoid intraoperative malfunction while patient is positioned prone.

In lesions caudal to cervical cord we used lower limb muscle groups for EMG especially quadriceps, hamstrings, tibialis anterior and gastrocnemius muscles in addition to anal EMG and SSEP from posterior tibial nerve. One reference electrode was inserted in one triceps muscle to obtain a baseline EMG curve of a surely normal muscle. In cases with cervical cord lesions electrodes from both upper limbs muscle groups were added plus SSEP from median nerves.

In cases with cervicomedullary extension, monitoring of lower cranial nerves was applied especially via soft palatal and oropharyngeal electrodes which were inserted after intubation with the aid of a Magill forceps and were secured in place by adhesive tapes at the corners of the mouth and gauze packing of the oral cavity.

Anesthesiologists are informed as regards avoiding paralytic agents and minimizing volatile anesthetic gases during the procedure. All device connections were checked for integrity and security. Special care was directed towards avoiding iatrogenic injury to anatomical structures during insertion of needles especially median nerve, brachial artery, posterior tibial nerve and sural nerve.

Appropriate spinal level and skin marking were determined with the aid of intraoperative C-arm fluoroscopy

Systemic antibiotics were administered in recovery room forty five minutes before patient enters the operative room.

Attention was paid to meticulous hemostasis in each step from skin to dura with special care given to minute bony ooze to be sealed with bone wax. A clean and dry operative field was a must before dural opening.

Determination of solid part of the lesion was double checked by C-arm fluoroscopy and transdural ultrasound.

In intradural lesions, the dura was then opened under surgical microscope. Dural stay sutures to the muscles were taken. Careful arachnoid opening was done and arachnoid was stitched to the dura for closure at the end of the procedure.

Appropriate site for myelotomy (usually midline unless the tumor paves another clear way) was determined by intraoperative ultrasound and guidance of intraoperative neurophysiological monitoring.

Myelotomy was performed with micro dissector, micro forceps and micro scissors. Bipolar and unipolar diathermy were seldom used at low power to avoid heat production.

Care was taken to choose appropriate non-injurious suction pieces and suitable suction power at each step.

Cleavage tissue planes were searched for. One should not abruptly enter into the lesion for debulking before ascertaining that the lesion is not a rare unexpected one such as arterio-venous malformation, aneurysm or cavernoma.

If good cleavage planes were detected, trials at total excision were attempted but again, with guidance of neurophysiological monitoring. However, if no good tissue cleavage planes were detected, it's hazardous to harshly manipulate on normal cord tissue to try total excision. In these circumstances and with neurophysiological guidance, it is wise to obtain a safer near total or subtotal excision. Some IMSCTs were inoperable and even untouchable, in those, only the smallest biopsy was allowed.

Intraoperative ultrasonography was utilized to aid localization and step-wise tumor resection control.

Intraoperative loading dose of methyl prednisolone was given to fourteen patients to avoid postoperative new deficits.
RESULTS

The study included twenty-five children with different spinal tumors. There was slight male predominance. Mean age at presentation was 3.8 years (range 1.5 to 16 years). The mean duration of symptoms was six months.

The most common clinical presentation was motor deficits in twenty-two cases (88%), sensory deficits in fifteen cases (60%), and pain in twelve cases (48%) and sphincteric affection in ten cases (40%).

According to tumor location, eleven tumors were intramedullary, three tumors were intradural-extramedullary, six tumors were intracanalicular-extradural, three tumors affected the vertebral body with extradural extension and two tumors affected vertebral body only.

Cervical and dorsal spinal cord locations were the commonest in this study being nine (36%) and eight (32%) cases, respectively.

Commonest tumor pathologies encountered were ependymomas and astrocytomas in six (24%) and four (16%) cases, respectively. There were other pathology entities such dorsal primitive neuroectodermal tumor (PNET). Also, there was an interesting case of cauda equina space occupying lesion with total excision was accomplished. The lesion turned out to be a Bilharzian granuloma. Antibilharzial medical treatment was given to the patient who had gradual improvement of his motor and sphincteric deficits. Table 1 shows the different pathological entities within the studied cases.

<table>
<thead>
<tr>
<th>Location</th>
<th>Intramedullary</th>
<th>Extramedullary</th>
<th>Intracanalicular</th>
<th>Vertebral body</th>
<th>Vertebral body and extradural</th>
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<tbody>
<tr>
<td>Ependymomas</td>
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<tr>
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Intraoperative ultrasonography technology was utilized in nineteen cases (76%). Intraoperative neurophysiological monitoring techniques were utilized in twenty cases (80%). Setup for motor evoked potential (MEP) including anal EMG and somatosensory evoked potential (SSEP) was prepared after induction of anesthesia while the patient is still supine. Anal EMG was the only positive indicative parameter for intraoperative guidance in five cases.

Total excision was performed in nine cases (36%). Near total or subtotal excision was achieved in twelve cases (48%) while only biopsy was possible in four cases. Total excision was achieved in 50% of ependymomas and in 25% of astrocytomas. Tumor recurrence was encountered in two cases. One of them was anaplastic ependymoma and the other was anaplastic astrocytoma.

Postoperatively, neurological deficits improved in twelve cases, stabilized in eleven cases and worsened in two infiltrating astrocytoma cases. Four patients showed transient deterioration of the neurological functions with gradual improvement over five days to eight weeks.

Postoperative hydrocephalus occurred in three cases with cervical lesions. Cerebrospinal fluid (CSF) diversion in the form of ventriculo-peritoneal shunts was needed in these cases.

Superficial wound infections occurred in two cases. In one of them, surgical debridement was performed.

DISCUSSION

Revolution in neuroimaging especially the gold standard high quality MRI had led to significant improvement in diagnosis, assessment and followup of spinal tumors.5,13,23
Marvelous advances in microsurgical techniques and technologies made safe and effective management of spinal tumors possible.\textsuperscript{10,15,21}

Early diagnosis and early management of pediatric spinal tumors remain the most crucial elements affecting the outcome of such tumors.\textsuperscript{7}

In this study, the most common clinical presentation was motor deficits, sensory deficits, pain and sphincteric affection in 88\%, 60\%, 48\% and 40\% of cases, respectively.

Postoperatively, neurological deficits improved in twelve cases (six of which had total excision of their lesions), stabilized in ten cases and worsened in two cases.

It appears that neurologic improvement after surgery is more likely in patients undergoing total resection than in patients undergoing partial resection.\textsuperscript{19}

Hence, gross total resection should be the aim, but if the cleavage plane between tumor and cord is unclear, then near total/subtotal resection would still be beneficial.\textsuperscript{6}

In this study, ependymomas and astrocytomas are the most frequent intramedullary tumors. Ependymomas of the spinal canal may be located intramedullary, attached to the filum terminale, or even extradurally originating from heterotopic ependymal cells.\textsuperscript{14-15} In this study, ependymomas and astrocytomas constituted collectively 40\% of the study cases.

In the literature, overall rates for complete resections vary between 23\% and 81\%.\textsuperscript{8,10,22,19} In the current study; overall total excision rate was achieved in nine cases (36\%). Near total or subtotal excision was achieved in twelve cases (48\%) while only biopsy was possible in four cases. Total excision was achieved in 50\% of ependymomas and in 25\% of astrocytomas.

The extent of resection varied according to the nature of the lesion and whether good tumor-normal cord interface was clear. Excision was safe when there had been good cleavage planes around the tumor. In many cases that was not the situation. Tumor cleavage planes are more evident in ependymomas than in astrocytomas which are more infiltrative. We had gross total resection in 50\% of ependymomas as compared to 25\% in astrocytomas due to lack of intraoperative clear dissection planes and in many astrocytoma cases MEP or SSEP gave us warning signals that hindered safe total excision.

Three astrocytoma cases had non-total surgical excision. Those cases received adjuvant chemotherapy according to the low grade glioma chemotherapy protocol adopted in our center.

On the other hand, three cases of ependymoma with postoperative residual were subjected to adjuvant conformal radiotherapy. Figure 1 \& 2 show two cases of spinal cord ependymoma with different extents of surgical resection.

Spinal intradural extramedullary tumors constitute about 25\% of spinal tumors in children.\textsuperscript{16} In the current study; three cases (12\%) had their lesions intradural and extramedullary. One case was an arachnoid cyst. Opening of cyst and marsupialization of its walls was performed followed by insertion of simple cystoperitoneal tube. Another case was a dermoid cyst which was surgically excised. The third case was a schwannoma which was totally excised (Fig. 3 a-d).
Fig. 3 a-d: Fourteen years old male patient presented with low back pain and sciatic pains. a & b: Preoperative sagittal T1W and T2W MRI showing lumbar intradural space occupying lesion at the level of second and third lumbar vertebrae. c & d: Early postoperative T1W and T2 weighted (T2W) MRI showing total excision of the lesion. The lesion proved to be Schwannoma of the filum terminale.

Six cases had purely intracanalicular extradural lesions. Among which there had been a case with left hip synovial sarcoma which showed progression and extradural metastasis for which decompressive laminectomy was performed together with excision of the extradural lesion.

Also, two cases presented with affection of vertebral bodies with intracanalicular extradural extension. One of which was a female with osteosarcoma of the humerus that showed metastasis to the lung and the fifth lumbar vertebra with extradural extension (Fig. 4 a-d). The other case was a craniocervical chordoma for which total surgical excision was performed (Fig. 5 a-b).

Fig. 4 a-d: Twelve years old female patient who had an osteosarcoma of the left humerus. She presented later on with agonizing low back pain and mild paresis of her lower extremities. a & b: Preoperative sagittal T1W and T2W MRI showing uniform abnormal hyper intense signals of the body of the fifth lumbar vertebra together with extradural mass compressing the thecal sac at the same level. c & d: Early postoperative T1W and T2W MRI showing decompression of the spinal canal and excision of the extradural soft tissue component. The lesion proved to be metastatic osteosarcoma.

The lesion proved to be Schwannoma of the filum terminale.
Binning et al reported that about 3-5% of children with systemic cancer present with spinal cord compression. However, we encountered spinal cord or thecal sac compression in three cases in this study. Interestingly, the tumors of origin were all skeletal namely osteosarcoma, synovial sarcoma and Ewing's sarcoma.

In this study there had been one case of conus medullaris lesion that had undergone surgery for excision of the lesion and the lesion turned out to be a Bilharzioma. This highlights the fact to deal with any IMSCT cautiously. Neat and careful surgical approach to such subtype of lesions is important as those lesions may turn out to be vascular malformations or rare non-neoplastic lesion.

In every case, whenever possible, the extent of resection should be monitored by intraoperative neurophysiological monitoring. Due to unique anatomical locations within spinal cord, when SSEPs are still intact, surely motor tracts are still good while when SSEPs become abnormal, motor tracts may or may not be affected yet. Thus, MEPs are more representative to motor function however SSEPs are good positive but not good negative. Although intraoperative neurophysiological monitoring is very beneficial in surgery of IMSCTs, it’s of little help in cases of spinal extradural lesions.

Intraoperative loading dose of methyl prednisolone was given to fourteen patients whenever there was suspicion or a will to prevent spinal cord edema after intraoperative finding of highly infiltrative lesions making fine surgical manipulations potentially fearful. In these circumstances, usually a maintenance dose is also given for twenty three hours postoperatively. No cases showed any systemic complication of methyl prednisolone.

Intraoperative ultrasonography technology was utilized in nineteen cases (76%). It was especially helpful in lesions with cystic and solid components. In cases where spinal cord was not significantly distended by the tumor, it helped in accurate tumor localization, and with the aid of intraoperative neurophysiological monitoring, precise safest location of presumed myelotomy was determined. Also, ultrasonography was beneficial in real-time monitoring the extent of resection and assuring total excision in cases where good cleavage planes were found.

Intraoperative neurophysiological monitoring techniques were utilized in twenty cases (80%). Anal EMG was the only positive indicative parameter for intraoperative guidance in five cases. This highlights the importance and usefulness of utilizing anal EMG especially in lesions affecting the conus medullaris and cauda equina because it may be the only positive indicative parameter and thus, unnecessary postoperative deficits are avoidable.

In our experience, it is important to highlight the fact that postoperative hydrocephalus should be always suspected in all cases of cervical and cervicomedullary formation of pseudomeningocele. However, such an effect has not been proven in adults and could only be demonstrated for children.

Overall tumor recurrence rates of 24% were reported. We experienced tumor recurrence in two cases one of which was anaplastic astrocytoma and the other one was anaplastic ependymoma. The significant variation in recurrences between benign and malignant spinal tumors is self-explanatory.

Reinsertion of the vertebral lamina is supposed to preserve normal anatomical planes and minimize formation of pseudomeningocele. However, such an effect has not been proven in adults and could only be demonstrated for children.

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**Fig. 5 a-b:** 7 years old male patient presented with neck pain and abnormal gait. **a:** Preoperative sagittal TIW MRI with contrast showing anterior extradural cervical lesion opposite the upper four cervical vertebral bodies and exerting a mass effect on the opposing cervical spinal cord. **b:** Early postoperative TIW MRI with contrast showing total excision of the lesion which proved to be chordoma.
lesions whether the cisterna magna was opened or not. Urgent CT study of the brain should be asked for any patient with postoperative signs or symptoms of increased intracranial pressure or with significant wound collection or cerebrospinal fluid (CSF) leak. Three cases in this series had postoperative hydrocephalus as compared to preoperative baseline images. Ventriculo-peritoneal shunt systems were inserted for CSF diversion in those cases.

CONCLUSION

Management of pediatric spinal tumors is challenging. Our management strategy relies mainly on optimizing safe and maximal tumor excision in cases when there are clear cleavage planes and avoiding total excision in infiltrating tumors. Frozen section pathology is mandatory to decide further surgical attitude. Advanced surgical adjuncts are mandatory for safe and effective surgery of spinal tumors especially in infiltrating tumors. Frozen section pathology is mandatory to decide further surgical attitude. Advanced surgical adjuncts are mandatory for safe and effective surgery of spinal tumors especially intraoperative neurophysiological monitoring and ultrasonography. Anal EMG is of special importance in intraoperative neurophysiological monitoring and effective surgery of spinal tumors especially.

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