PEDIATRIC INTRASPINAL TUMORS

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ABSTRACT:

Intraspinal tumors in infancy and childhood create diagnostic problems different than those of adults. Muscle weakness, back pain, gait disturbance, rigidity, and sphincter dysfunction are the most common presenting symptoms. In all intraspinal tumor cases the clinical signs and symptoms are progressive, and the neurological deficits due to cord compression are almost irreversible; but in the pediatric group, the patient has a high chance of being ambulatory with early diagnosis and appropriate treatment.

Although plain roentgenography and myelography are suggestive, CT and MRI must be performed for accurate diagnosis and follow-up of these patients. In addition to microneurosurgical dissection; intra-operative ultrasonography, SEP monitoring, CUSA, and lasers may be used to achieve maximal tumor removal with minimal neurologic deficit. When total removal of the tumor will result in additional neurological deficits, it should be avoided.

Between the years 1965-1994; 103 patients younger than 15 years of age were operated in Ankara University, Department of Neurosurgery. Most of the tumors were located extradurally, in the thoracic region. Primary spinal tumors constituted the major histopathological group. Nearly 2/3 of the patients clinical condition improved postoperatively.

To obtain the best results in the treatment of pediatric intraspinal tumors, we advocate early surgical intervention followed by radiotherapy and/or chemotheraphy combined with a physical rehabilitation programme.

Key Words: *Pediatric Intraspinal Tumors, Diagnosis, Surgical Treatment*

INTRODUCTION

Intraspinal tumors in infancy and childhood create diagnostic and theraupetic problems different than those of adults. Muscle weakness, back pain, gait disturbance, rigidity and sphincter dysfuction are the most common presenting symptoms (1, 7). Although plain roentgenography and mylography are suggestive, CT and MRI must be performed for accurate diagnosis and follow-up of these patients. In addition to microneurosurgical techniques, Intra-operative ultrasonography, SEP monitoring, CUSA and lasers may be used to achieve maximal tumor removal with minimal neurologic deficit. To obtain the best results in the treatment of pediatric intraspinal tumors, we advocate early surgical intervention followed by radiotheraphy and/or chemotheraphy combined with a physical rehabilitation programme (2, 11).

MATERIAL AND METHOD

Between the years 1965-1994; 103 patients younger than 15 years of age were operated in Ankara University Department of Neurosurgery. Of the 103 patients, 49 were boys and 54 were girls. This group comprised 8.25 % of all spinal neoplasms operated in our clinic during the same period. The incidence of intraspinal tumors in the pediatric group increased after 11 years of age in our patients.

SIGNS AND SYMPTOMS :

Muscle weakness (88/84.5 %), back pain (62/60.2 %), extremity pain (37/35.9 %), gait disturbance (91/89.3 %), rigidity (23/22.3 %), urinary dysfunction (51/49.5 %) were the most frequent symptoms. Rectal syphincter disturbance (14/13.6 %), the presence of a mass/scoliosis (8/7.8 %) and torticollis (3/2.9 %) were the other presenting symptoms. Reflex changes were seen in 85 patients (82.5 %) with paralysis in 73 (70.9 %) and sensory deficits in 71 (68.9 %) of them. The shortest duration of symptoms was 5 days; the longest was 6 years (mean: 9.5 months). In all cases the development of clinical signs and symptoms were progressive.

DIAGNOSIS

Plain roentgenogrpahy showed abnormal findings like; pedicle erosions, lytic lesions, widened spinal canals and foramina, scoliosis, exostosis and compression fractures in 78 patients (75.7 %). Myelography

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was performed on 92 patients; 77 of them having complete. 14 of them having incomplete block and one patient with normal mylogram. CT scans were available in 26 of the patients and 12 patients had MRI examinations, all demonstrating the lesions.

LOCALISATION AND DISTRIBUTION

Segmental distribution of the lesions in our series were as follows; cervical 13 (12.7 %), cervicothoracic 10 (9.7 %), thoracic 44 (42.7 %), thoracolumbar 19 (18.4 %), lumbar 14 (13.6 %), lumbosacral 3 (2.9 %). The tumors were localised intramedullary in 27, intradural-extramedullary in 18, epidurally in 53 of the patients. 5 patients had dumpbell tumors.

TREATMENT

All 103 patients were treated surgically. In 62 cases (60.22 %) total and in 39 cases (37.9 %) subtotal excision of the tumor were performed. In 2 cases only biopsy from the tumor could be taken after performing decompressive laminectomy. We avoided total excision intentionally in 16 cases because of the possibility of increasing the neurologic deficit iatrogenically.

HISTOPATHOLOGY

Histopathological diagnosis, typing and distribution of the tumors were as follows:

Type of the tumor:	Number
Primary spinal tumors	31
Astrocytoma	14
Meningioma	5
Neurofibroma	9
Ependymoma	3
Congenital Tumors	19
Epidermoid	10
Lipoma	3
Arachnoid cyst	4
Teratoma	2
Bone Tumors	23
Haemangioma	6
Aneurysmal bone cyst	4
Endothelial myloma	4
Immunoblastic sacroma	2
Osteoma	2
Osteochondroma	2
Ewing's sacroma	3
Lymphoid Tumors	7
Lymphosarcoma	2
Lymphoblastic lymphoma	2
Burkitt lymphoma	1
Malignant lymphoma	2
Infections - Parasites	6
Cyst hydatid	2
Tuberculous granulation tissue	4
Miscellaneous	17
Metastasis	9
Ossified fibroma	2
Fibroadipose tumor	2
Non-verified	4

RESULTS

All patients in this series have been followed-up for a minimum period of a month. The average follow-up period was 15 months post-operatively. The outcome was classified as; markedly good: when there was no evidence of clinical or radiological recurrence of the initial disease at the follow-up and the patient was neurologically intact, slightly good: when there was no evidence of recurrence but the patient had persistant neurologic deficit, no change: when there was recurrence of the tumor and the patient had persistant neurologic deficit, poor: when the patient had no improvement or detorioration of the initial clinical condition with major neurologic deficit.

At the end of the average follow-up period: 24 patients (23.4 %) were in the markedly good, 54 (52.5 %) patients were in the slightly good group. 17 patients (16.5 %) clinical conditions were unchanged and 8 (7.8 %) patients have had poor results.

In general; the patients with intra-medullary tumors were neurologically stabilised, the patients with congenital tumors were in markedly good condition, whereas the patients with bone tumors had poorer results.

POST-OPERATIVE COMPLICATIONS AND REHABILITATION

Eighteen patients (17.5 %) had kyphoscoliosis 7 (6.8 %) had paralytic dislocation of the hips, 9 (8.7 %) had leg length discrepancy and 33 patients (32 %) had contractures and deformities of the lower extremities in the post-operative follow-up period.

When a rehabilitation programme was indicated intensive physiotherapy was instituted as soon as the patients clinical condition improved and wheelchair, spinal brace or long leg walking brace was used according to the patients needs.

DISCUSSION

The child with an intraspinal tumor usually presents with weakness, pain or syphincter dysfunction. Pain is reported to be the most frequent and important symptom in the literature; that enables early diagnosis (13). In our series extremity weakness was more common (84.5 %) and urinary dysfunction which was reported to be up to 30 % was 49.5 (7, 4, 15). Scoliosis occured in 4.8 % of our series. Richardson (16) emphasized the importance of spinal rigidity as an early of an intraspinal tumor. An alternation of gait was the most frequent symptom in these patients and should be stressed as a pertinent finding (13). The percentages of the clinical signs and symptoms in our series approximately match those reported in the literature (7, 4, 15). Although in 15 % of the patients pain continued; various neurological deficits were cured in 10 months time post-operatively. The incidence of kyphoscoliosis was 6.8 % in our series; accordingly we suggest that after intraspinal tumor surgery immediate spinal brace application should be employed. We agree with Matson (13) that much of the residual disability which occur, result from the prolonged delay before a proper diagnosis is made; rather than from the effects of the operation or from the progression of the disease post-operatively.

In the literature (5) it is reported the the most frequent tumors were glial, sarcomatous and congenital in decreasing order. In our series the frequency distribution of was different; primary spinal, bone, congenital and glial tumors in decreasing order. Lymphoid tumors have an incidence of 1-3 % in other series (5, 7) whereas they were more frequently (6.8 %) encountered in our series.

Spinal roentgenography was abnormal in 75.7 % of our patients. Matson (13) had abnormal roentgenographic findings in 66 % of his patients. Myelography demonstrated the lesion in 98.7 % of the patients; De Sousa reported this to be 95.7 % (4). We advise mylography as a cheap and reliable diagnostic method. It should also be acknowledged that MRI is a diagnostic tool to determine the nature of the lesion (6). Blews (3) considers MRI to be equal to or more valuable than myelography or CT imaging for the diagnosis of these lesions. MRI can accurately localise spinal canal widening without the need for the introduction of intrathecal contrast media (9).

By means of intra-operative ultrasonography, cystic or solid components and borders of spinal tumors are better identified and localised than by CT (10). Also the intra operative use of ultrasonography shows the optimal area to open the dura and to perform myelotomy.

Somatosensory evoked potentials (SSEP) are used for monitoring intraspinal sensory pathways. This monitoring is only valuable if the information is immediately available and utilised by the surgeon to modify the operative dissection. When used in conjunction with lasers, SSEP monitoring provides time for the surgeon to stop and cool the tissue by irrigation (12). We performed SSEP monitoring in 13 of our patients and 8 of these patients findings correlated closely with the post-operative clinical condition.

The treatment of all the intraspinal tumors in our patients were surgical. Total tumor removal was reported to be possible in over one third of the cases (4), but this was 60.2 % in our series. As expected, the patients in whom total tumor removal could be performed had a chance for complete recovery, indicating a better prognosis for patients diagnosed and treated early.

Especially during the operations on intromedullary tumors the use of bipolar coagulation. CUSA and lasers (CO2, Argon, Nd: YAG) helped the surgeon in total removal of the tumor. CUSA enabled us to excise the tumor without spinal cord manipulation and trauma in operations where therewas no definite tumor-cord cleavage plane: like those in glial tumors. CO2 laser provides the vaporisation of solid tumors and removal of the residual tumor after the use of CUSA. Argon laser is usually less traumatic and often more hemostatic being used for the excision of strategically placed, small and moderately vascular spinal cord tumors. Nd: YAG laser can be used in the vicinity of spinal cord and brain stem with the help of SSEP monitoring (12).

In a study by Raffel (15), it is stated that pediatric intraspinal tumors are best treated by a combination of surgical decompression and tumor removal followed by adjuvant theraphy regardless of the tumor type. We applied a similar protocol in our series. Radiotheraphy and/or chemotheraphy was given to all patients following the maximal tumor removal by surgery.

Deformities may develop in patients who had multilevel laminectomies (8) or assymetric irradiation (14); so we strongly believe in and recommend the early institution of the proper rehabilitation programme.

CONCLUSION

It is almost impossible to lessen the neurological deficits caused by spinal cord compression in adults despite every effort for treatment. But in case of children; inspite of total motor and sensory deficit, the patient has a 50 % chance of being ambulatory. Due to above mentioned reasons, the combined useage of early diagnostic methods together with radical surgery and medical theraphy modalities become more important. Modern diagnostic tools enables early and accurate diagnosis of the lesion; whereas microsurgical techniques and instruments offer the surgeon great advantages and lower the incidence of post-operative complications. Chemotheraphy and/or radiotheraphy should always be employed as an adjunct to surgery. Preventive physical theraphy and rehabilitation measures when taken properly may lessen post-operative disability.

REFERENCES:

1. Arseni, C., Horvath, L., Iliescu D.: Intraspinal tumour in children. Psychiat Neurol Neurochir 1967; 70: 123 - 133.
2. Banna, M., Gryspeerdt L.: Intraspinal tumour in children. Clin. Radiol 1971; 22: 17 - 32.
3. Blews, D.E., Wano, H., Kumar, A.J., Robb, P.A., Phillips, P.C., Bryan, R.N.: Intradural spinal metastases in pediatric patients with primary intracranial neoplasms: Gd-DTPA enhanced MR vs CT mylography. J. Comput Assist Tomogr. 1990; 14: 730 - 735.
4. De Sousa, A., Kalsbeek, J. Mealey, J. Campbell, R.L., Hockey, A.: Intraspinal tumors in children. A review of 81 cases. J. Neurosurg 1979; 51: 437-445.
5. Di Lorenzo, Giuffre, R., Fortuna, A.: Primary spinal neoplasm in childhood. Neurochirgia 1982; 25: 153-164.
6. Duthoy, M.J., Lung, G.: MR imaging of the spine in children. Eur J. Radiol 1988; 8: 188-195.
7. Farwel, J.R., Dohrmann, J.: Intraspinal neoplasm in children. Paraplegia 1977-78; 15: 262 - 273.
8. Fisher, G.G., Hall, J.G.: Spinal deformities following neurosurgical procedure in children. In: Schmidek, H.H., Sweet, W.H., editors. Operative Neurosurgical Techniques. Intications. Methods and Results. New York: Grune and Stratton, Vol. 1; 1982; 63 - 69.
9. Frederics, B.J.: Diseases of the spinal canal in children: Diagnosis with non-contrast CT scans. AJNR 1989: 10: 1233 - 1238.
10. Grundy, B.L.: Monitoring of SEP during neurosurgical operations: Methods and applications. Neurosurgery 1987; 20: 169 - 182.
11. Hoffman, H.J. Griebel, R.W., Hendrick, E.B.: Congenital spinal cord tumors in children. In: Symon L. editor. Advances and Technical Standarts in Neurosurgery. Wien: Springer Verlag. 1986: 175 - 200.
12. Levy, W.J.: Clinical experience with motor and cerebellar evoked potential monitoring. Neurosurgery 1987; 20: 169 - 182.
13. Matson, D., Tachdjian, P. Intraspinal tumors in infants and children. Postgrad Med. 1963; 34: 279-285.
14. Mayfield, J.K., et al.: Spinal deformity in children treated for neuroblastoma. The effect of the radiation and other forms of treatment. J. Bone and Joint Surg. 1981; 63 (A) : 188 - 193.
15. Raffel, J., Neave, V.C., Lavine, S., McComb, J.G.: Treatment of spinal cord compression by epidural malignancy in chilhood. Neurosurgery 1991; 28: 349 - 356.
16. Richardson, F.: A report of 16 tumors of the spinal cord in children. J. Pediatr. 1960; 57: 42 - 54.