PRIMARY INTRASPINAL TUMORS IN CHILDREN AND ADOLESCENTS

A REPORT ON 12 CASES

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Tumors in children and adolescents are not so infrequent as generally believed. One is impressed by the difference between early textbooks of pediatrics, which refer very briefly to tumors, and modern works in which extensive chapters deal with this problem. Tumors of the central nervous system are prominent among neoplasms occurring in childhood. In a statistical analysis of 1770 cases of tumors in children, tumors of the central nervous system represented 21 per cent, thus sharing first place with bone tumors. The vast majority of tumors of the central nervous system in children are localized in the cerebellum, brain stem and cerebral hemispheres and only a few in the spinal canal. This accounts for the inadequate presentation of intraspinal tumors in most textbooks of pediatrics with the exception of Bucy’s excellent contribution to Nelson’s Textbook of Pediatrics although, as will be indicated later, the authors would not agree with his somewhat sweeping statement that ependymomas are the tumors encountered most frequently.

The existence of intraspinal tumors in children was demonstrated for the first time at the end of the nineteenth century by Horsley and Gowers, and then by Schlesinger, both times in postmortem studies.

Later, with the progress of clinical knowledge and therapeutic means, the study of intraspinal tumors in the young entered a new epoch and numerous reports on this subject appeared in the literature.

According to Kornianskii the number of cases published at the time of his report (including his own series) was 467. We believe that the number of primary intraspinal tumors is somewhat smaller because some authors, for instance Ford and Svien et al. included in their series metastatic tumors, extrarachidian tumors (mediastinal) with secondary intraspinal invasion, lipomas associated with malformations of the spinal column or of the spinal cord, and vascular malformations, all of which should not be included since they are not primary intraspinal tumors. It is worth mentioning that in 1935 Hamby was able to gather only 100 cases from the literature.

FREQUENCY, SEX, AND AGE

Between 1935 and 1960 we studied in the "Prof. D. Bagdasar" neurosurgical clinic in Bucharest 12 patients (under 16 years of age) harboring intraspinal tumors. Both our data and those collected from the literature confirm Bailey’s statement that whereas in adults the rate of intracranial tumors/intraspinal tumors is 6/1, in children it is 20/1 (the only exception is the rate of 5/1 given by Ingraham and Matson). As far as the sex of our patients is concerned, females predominated (2/1) while in a series of intraspinal tumors in adults the rate was roughly 1/1.

Our patients were divided in two age groups: between 0 and 12 years, and between 13 and 16 years. Only 3 patients were in the first group, while the second included 9 patients (a rate of 3/1 in favor of the older group). The youngest patient was 6½ years of age.
PATHOLOGY

More than half of the tumors (58 per cent) were localized in the thoracic segment of the spinal canal, 33 per cent in the thoracolumbar or lumbar segment and only 1 tumor was situated in the cervicothoracic segment. No tumor was purely cervical. These differ from the data gathered by Kornianskii (53 per cent thoracic, 25 per cent cervical and 18 per cent lumbar). Only 1 tumor was extradural, 8 were subdural and 3 were intramedullary (9, 66, and 25 per cent).

Our data are in agreement with those of Kornianskii for intramedullary tumors, but most authors reported a much greater frequency of extradural tumors, roughly 40 per cent in Grant and Austin’s series and 20 per cent in Kornianskii’s own cases.

The nature of the tumors met with in our patients was varied. There were 4 cases each of neurinomas and gliomata, and 2 cases each of meningiomas and sarcomas. In only 1 of the 4 gliomata could the type of glioma be ascertained (an ependymoma); in the other 3 cases, because of the intramedullary situation of the tumor, no fragment of tissue was taken for microscopic examination. Our series differs from those reported by others in that we had no case of teratoma or dysembryoma, and the percentage of meningiomas and neurinomas was high in our series. There was no difference between children and adults in the proportion of different types of intraspinal tumors observed in our clinic, a fact that is in disagreement with the observations of most authors.

CLINICAL ASPECTS

In all cases the evolution of clinical signs and symptoms was progressive. The duration from the initial symptom until admission to the clinic is shown in Fig. 1.

The shortest duration was 1 month; the longest was 7 years. The first symptom in half of our patients was pain (of radicular or cordal type) whereas in 5 patients the clinical evolution started with motor weakness, and only 1 patient presented from the very beginning a mixed algio-motor syndrome.

In only 1 of the 3 patients with intramedullary tumors were pains (of the cordal type) the first symptom.

On admission most patients presented the classical triad of spinal compression (sensory, motor and sphincteric disturbances). Motor weakness was present in all patients: of spastic type in 7, flaccid in 4, and flaccid-spastic in 1. Four of our patients showed myatrophy (2 had intramedullary tumors, 1 a caudal-equina ependymoma and 1 a meningioma in the lower thoracic region).

A dissociated hyp- or anesthesia with upper level was present in all but 1 case (in the latter a L3 neurinoma was found). It is true that in young patients it is sometimes difficult to ascertain if there is a disturbance of sensation with an upper level, but a useful trick is to test sensitivity to pin prick by mimicking a game—while the child’s attention is distracted by another person, the examiner watches his facial expression. Pain was much less frequent in the advanced stage of clinical evolution than at the beginning.

Half of our patients had sphincteric disturbances (both sphincters being affected in these cases).

One patient showed kyphosis and 1 a kyphoscoliosis.

PARACLINICAL INVESTIGATIONS

In three-quarters of our patients radiography of the spinal column disclosed no abnormality. In 1 patient it showed widening of the spinal canal, in 1 a scoliosis, and in 1 a kyphoscoliosis. Lefèvre and coworkers gave a much higher percentage of radiological changes. In 14 out of their 18 cases there were changes in the statics of the spinal column and in 8 out of 18 the spinal canal was widened. Myelography (with Lipiodol)
was performed in 9 of our cases, showing a complete block of the opaque oil in 3 and a partial one in 6, whereas Lefèbvre et al.\textsuperscript{12} in 16 cases in which myelography was performed found a complete block in 11, and a partial block in 5.

The Queckenstedt-Stookey test was used in 4 cases; in 2 of these there was blockage of the subarachnoidal space. Whenever it was performed, the cerebrospinal fluid showed a sublesional increase of protein varying between 1.32 gm. and 17.60 gm. per cent (Heller method), the number of cells being normal or slightly increased. Occasionally the fluid was xanthochromic.

**DIAGNOSIS**

The diagnosis of an intraspinal tumor does not present any difficulty provided that one is "tumor-conscious." In most cases the clinical evolution gives a clue to the correct diagnosis which can be ascertained by ancillary investigations, the most valuable being the radiologic, as has been shown by Lefèbvre et al. and demonstrated by practice. During the "pure" algetic phase of the clinical evolution a differential diagnosis is to be made from non-neurological diseases (thoracic, abdominal and rheumatic) which can give rise to pseudoradicicular pain, as well as from radiculitis and neuritis. The signs and symptoms which later are associated with the algetic disturbances as well as some of their peculiarities—for instance, their being more accentuated during the night—eventually enable one to make the correct diagnosis.

In patients presenting an algionmotor syndrome—this is the type of complaint with which the patient is usually brought in consultation—the essential problem is to eliminate the presence of Pott's tuberculous spondylitis giving rise to radiculomedullary phenomena. The absence of tuberculosis of any type in the past history, the negative intradermic test and the normal radiography of the spine make it possible to rule out this diagnosis. Although bone tumors are very frequent in children, tumors of the spine are extremely uncommon. Arseni et al.\textsuperscript{3} in a series of 350 tumors of the spine reported only 2 patients under 10 years and 16 under 20 years of age. Of course there is always a characteristic radiological image present. Despite the fact that vascular malformations of the spinal cord are congenital, they almost never become clinically manifest in patients under the age of 20 years,\textsuperscript{2} although Ford\textsuperscript{6} mentioned a case of arteriovenous fistula in a child. Spinal arachnitis is not frequent in children and has clinical peculiarities which do not make the differential diagnosis very difficult. Syringomyelia is quite frequent in adolescents but the disturbances of sensibility have characteristics not usually found with intraspinal tumors, although theoretically an intramedullary tumor at the beginning of its clinical evolution might give rise to a similar aspect. Moreover, motor disturbances in syringomyelia are discrete and trophic disturbances are usually present. In infants with discrete motor weakness and hypotonia in whom no disturbances of sensibility can be demonstrated, an Oppenheim's amyotonia congenita and a Werdnig-Hoffmann syndrome have to be taken into consideration. The rapid increase of motor weakness, the want of sensory disturbances and the familial character of the latter disease as well as the negative ancillary investigations make it possible to establish a correct diagnosis. In patients with flaccid motor weakness and myatrophic disorders, sequelae of poliomyelitis have to be taken into consideration but these are never evolutionary and the finding of sensory disturbances and ancillary investigations point to the correct diagnosis. Finally there are rare cases of juvenile multiple sclerosis, but almost never is spasticity present alone, and the course is not steady but undulating. The essential problem is to make the correct diagnosis early enough in order not to jeopardize the results, which can be good in cases of extra- and subdural tumors.

**TREATMENT AND RESULTS**

The treatment of intraspinal tumors is either surgical, or surgical and roentgenologic.

In cases of extramedullary tumors the tumor is always extirpated. Roentgenotherapy is given only in cases of sarcomas
and ependymomas. In cases of intramedullary tumors, after a decompressive laminectomy with opening of the dura mater, sometimes the spinal cord is split caudorostral over the tumor to lessen the distention of the spinal cord by the tumor. In some instances after this maneuver, part of the tumor extrudes and can be removed for histological examination. In our opinion it is not advisable—if this does not happen—to try removing fragments of tumor because of the difficult hemostasis and subsequent danger of ischemia of the spinal cord. The dura mater is never closed. After postoperative recovery roentgen-ray therapy is given in the usual doses. The operative mortality in our series was nil. The immediate and late functional results in cases of intramedullary tumors are disappointing although, with adequate nursing, the survival rate is not too bad. In none of our 3 patients with intramedullary tumors could any postoperative improvement be observed.

With extramedullary tumors the results as far as function is concerned are much better, provided that the diagnosis is made soon enough. It is only an apparent paradox to say that good results are attributable to the neurologically minded pediatrician and not to the neurosurgeon. This is especially important because, at least in our series, many if not most extra- or subdural tumors are benign and the prognosis—quo ad vitam—is excellent, and there are few situations more dreadful both for the patient and for his parents than to remain disabled for a lifetime.

In 5 out of 19 cases of extramedullary tumors we obtained more or less important functional recoveries. It should be pointed out that, with one exception, recoveries occurred only in patients with a preoperative evolution of less than 9 months and almost exclusively in those with spastic motor weakness (a single patient with flaccid motor weakness showed some improvement). Important factors in functional recovery are long kinestherapy and adequate nursing.

One can conclude that in cases of benign extramedullary tumors the patient’s future is not too gloomy if he gets adequate neurosurgical treatment soon enough.

REFERENCES