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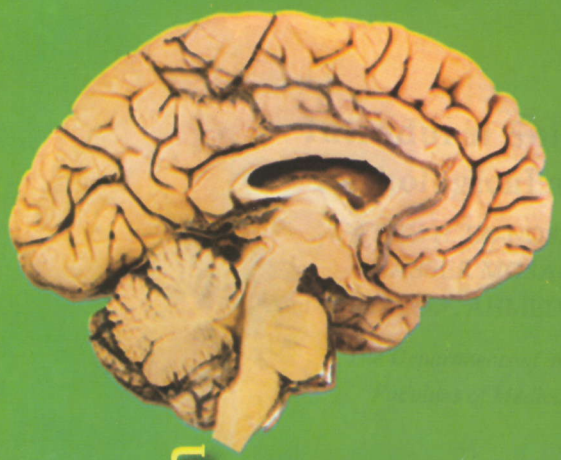

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Intradural Spinal Cord Tumors Clinico - Pathological and Therapeutic Features

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Abstract

A series of 40 patients having intradural spinal cord tumors (30 with intradural extra medullary tumors and 10 with intramedullary tumors) were managed at Neurosurgical departments of Benha and Al-Azhar University Hospitals, during the period from 1992 to 1997, were studied for clinical presentation, pathological features, treatment modalities and surgical outcome. 16 patients had meningiomas, 12 neurofibromas, 5 astrocytomas, 4 ependymomas, and one for each of heamangioma an arachnoid cyst and dermoid cyst . The patients included 28 females and 12 males with a median age of 38 years. Motor deficits were the most frequent neurological findings {85%}, back pain was present in 70%, and sphincteric dysfunction in 35% patients. Metrizamide myelography, CT scan with intrathecal metrizamide and MRI were the imaging modalities of the study. Total removal of the tumor was the aim of surgery. It was achieved in all 16 cases {100%}of meningiomas, in 11 cases {92%}of neurofibromas, in 2 cases {50%}of ependymomas and for each case of haemangioma, arachnoid cyst and dermoid cyst, and subtotal removal in 3 cases {60%} of astrocytomas, in 2 cases {50%}of ependymomas and one case {8%} of neurofibroma, whereas biopsy in 2 cases {40%} of astrocytomas. Radiotherapy was performed in malignant tumors that were subjected to subtotal removal or biopsy. The clinical outcome was influenced to a large extent by the preoperative neurological status, tumor pathology and patient's age.

We emphasise the importance of early diagnosis and appropriate management including {recent advances in neuroimaging as MRI, new technological advances in surgery as CUSA unit, and microsurgical tools} to improve the surgical outcome of these tumors.

Introduction

SPINAL cord tumors account for about 15 percent of central nervous system neoplasms [1]. The majority of these intradural tumors arise from the cellular constituents of the spinal cord and filum terminale, nerve roots, or meninges. Intradural spinal cord tumors are broadly categorized according to their relationship to the spinal cord [2]. Intramedullary tumors arise within the substance of the spinal cord, whereas extramedullary tumors are extrinsic to the cord. A small number of neoplasms may have both intramedullary and extramedullary components [3]. The most common intramedullary tumors are ependymomas {56%}and astrocytomas {49%} [6], while the most common intradural extramedullary tumors are neurofibromas{49%}and meningiomas {45%} [4]. The operative morbidity and mortality which was reported as 22% had been greatly reduced with the advent of recent advances in diagnosis such as metrizamide C.T and MRI and new facilities of treatment as laser, intraoperative evoked potential monitoring and CUSA unit [4].

Aim of the work:

The aim of this work was to study clinical presentation, pathological features, treatment modalities and surgical outcome of the patients with intradural spinal cord tumors.

Patients and Methods

This is a retrospective study that consisted of 40 patients of spinal cord tumors were managed at Benha and Al-Azhar Universities Hospitals during the period from 1992 to 1997. The same protocol of management was achieved in both departments. There were 30 patients with intradural extramedullary tumors {75%} {Group I} and 10 patients with intramedullary tumors{25%}{Group II}. There were 28 females {70%}and 12 males{30%}, their ages ranged between 9 and 67 years with a median age of 38 years. All the patients were subjected to precise history and accurate general and neurological examination.

Myelography was performed in 16 patients, via lumbar puncture in 12 patients,via cisternal puncture in 3 patients and via combined punctures in one patient. Plain CT scan was performed in 15 patients, after intrathecal contrast in 13 patients and after IV contrast in 2 patients. MRI before and after gadolinium was performed in 9 patients.

All the patients were subjected to surgery through midline approach {laminectomy, opening the dura and removal of extramedullary tumor or posterior midline myelotomy for intramedullary tumor}.

All tumors were subjected to histopathological examination after surgical interference.

Postoperative radiotherapy was performed in malignant tumors that were subjected to subtotal removal or biopsy.

The survivals were evaluated neurologically at time of discharge from the

hospital and periodically through follow up period (6 months to 30 months).

The outcome was categorized into :

I- Favorable outcome: Patients who showed improvement of the neurological status.

II- Unfavorable outcome: Patients who showed no improvement or deterioration of the neurological status.

III- Mortality.

Results

Table {1}: Age and Sex distribution in Group I.

Age	No. of patients	%	Male		Female	
			No.	%	No.	%
1st decade	-	-	-	-	-	-
2nd decade	2	7	2	7	-	-
3rd decade	2	7	1	3	1	3
4th decade	4	12	1	3	3	10
5th decade	8	27	1	3	7	24
6th decade	11	37	1	3	10	34
7th decade	3	10	1	3	2	7
Total	30	100	7	22	23	78

This table shows that:

- The peak incidence occurred in the 5th and 6th decades of life {27%and 37% respectively}.
- Marked predominance of females{78%}
- Male: Female ratio 1:3.3

Table{11}: Age and Sex distribution in Group II.

Age	No. of patients	%	Male		Female	
			No.	%	No.	%
1st decade	1	10	1	10	-	-
2nd decade	2	20	-	-	2	20
3rd decade	3	30	2	20	1	10
4th decade	3	30	2	20	1	10
5th decade	1	10	-	-	1	10
6th decade	-	-	-	-	-	-
7th decade	-	-	-	-	-	-
Total	10	100	5	50	5	50

This table shows that:

- The peak incidence occurred in the 3rd and 4th decades of life{30%for each decade}.
- Males and Females are equally affected.

Table (III): Clinical presentation of the patients.

Clinical presentation	No.	%
I- Symptoms:		
1- Pain:		
- Back pain	28	70
- Radicular pain	16	40
2- Motor weakness*	35	87.5
3- Sensory changes	14	35
4- Sphenicteric disturbances	14	35
II- Signs:		
*** Motor deficits:**	35	87.5
- Partial weakness	30	86
- Complete weakness	5	14
* Spastic	3	60
* Flaccid	2	40
*** Muscle wasting**	11	27.5
*** Sensory deficits:**	-	-
- Superfacial hypothesia with sensory level	18	45
- Deep sensory loss	8	20
- Sacral sparing	2	5
*** Reflex changes****	30	75
*** Back signs*****	3	7.5

This table shows that:

° Motor weakness* was the most frequent clinical feature {87.5%}.

° Sensory deficits ** were present in {70%} of patients, superficial hypothesia with sensory level constituted {70%} of sensory deficits and deep sensory loss {29%}.

° Reflex changes *** whether diminished, absent, exaggerated or appearance of pathological reflexes were present in {75%} of patients.

° Back signs**** in the form of back deformity {scoliosis and kyphoscoliosis} in 2 patients and skin dimple in one patients.

NB.: Dissociated sensory loss was recorded.

Table (IV): Pathology of the tumours (40 patients).

Type of pathology	Group I		Group II	
	No.	%	No.	%
1- Meningioma	16	54	-	-
2- Neurofibroma	12	40	-	-
3- Astrocytoma	-	-	5	50
4- Ependymoma	-	-	4	40
5- Haemangioma	1	3	-	-
6- Arachnoid cyst	1	3	-	-
7- Dermoid cyst	-	-	1	10
Total	30	100	10	100

This table shows that:

° Meningioma was the most predominant intradural extramedullary tumor (54 %), whereas Neurofibroma came a close second tumor (40%).

Astrocytoma and Ependymoma were the most predominant intramedullary tumors (50 % and 40 % respectively).

Table (V): Pathology in relation to sex.

Age	No. of patients	%	Male		Female	
			No.	%	No.	%
I- Group I:	30	75	7	23	23	77
1- Meningioma	16	53	2	12.5	14	87.5
2- Neurofibroma	12	40	3	25	9	75
3- Haemangioma	1	3	1	100	-	-
4- Arachnoid cyst	1	3	1	100	-	-
II- Group II:	10	25	5	50	5	50
- Astrocytoma	5	50	3	60	2	40
- Ependymoma	4	40	1	25	3	75
- Dermoid cyst	1	10	1	100	-	-
Total	40		12	100	28	100

This table shows that:

- In Group I: Marked predominance of meningioma in females (77%).
- In Group II: Slight predominance of astrocytoma in male (60%) and ependymoma in females (75%) .

Table (VI): Level of the tumours.

Type of pathology	Group I		Group II	
	No.	%	No.	%
1- Foramen magnum	1	3		-
2- Cervical	5	17	4	40
3- Cervicodorsal			2	20
4- Dorsal	23	77	1	10
5- Dorsolumbar	1	3	1	10
6- Conus medullaris	-	-	2	20
Total	30	100	10	100

This table shows that:

- Dorsal region was the predominant site in Group I (77 %), whereas cervical region was involved in 40 % in Group II.

- Site of predilection of the tumors:

- In Group I.

- Meningiomas were distributed in dorsal region in 13 patients (82%), in cervical region in 2 patients (12%) and in foramen magnum in one patient (6%).

Neurofibromas were distributed in Dorsal region in 8 patients (67%) (Figure II & III), in cervical region in 3 patients (25%) (Figure V) and in dosrolumbar region in one patient (8%).

Haemangioma and arachnoid cyst were located in dorsal region (Figure I & VII).

-In Group 11:

Astrocytomas were distributed in cervical region in 3 patients (60%), cervicodorsal region and dorsal region were each involved in one patient (20%).

-Ependymomas were distributed in conus medullaris in 2 patients (50%) (Figure IV), cervical region and cervico-dorsal region were each involved in one patient (25%).

Whereas the dorsolumbar region was involved in the single patient with dermoid cyst.

Surgical interference:

Total removal was achieved in all 16 cases (100%) of meningiomas, in 11 cases (92%) of neurofibromas, in 2 cases (50%) of ependymomas, in one case (20%) of astrocytoma (where a plane of cleavage was clear between the cord and tumor) and in each case of hemagnioma, arachnoid cyst and dermoid cyst.

Subtotal removal was achieved in 2 cases (40%) of astrocytomas, in 2 cases (50%) of ependymomas, (where the plane of cleavage was indistinct) and in one case (8%) of neurofibroma due to it's

large extension into chest through the neural foramen. Biopsy was achieved in 2 cases (40%) of astrocytomas (where the tumors were infiltrating the cord).

Adjuvant therapy:

Postoperative radiotherapy was indicated in the all 5 cases (100%) of astrocytomas, and in the 2 cases (50%) of ependymomas that were removed subtotally.

Table (VII): Postoperative complications.

Complications	No.	%
- Wound infection.	5	12.5
- CSF leakage	4	10
- Pseudomeningocele	2	5
- Bed sores	5	12.5
- Deterioration of neurological status*	9	22.5
- Deep vein thrombosis	3	7.5
- Meningitis	2	5
- Mortality**	2	5

This table shows that:

-Neurological status showed deterioration in the early postoperative period in 9 patients (22.5%). Improvement had gradually occurred in 6 patients, whereas the remaining 3 patients did not improve.

- Mortality** had occurred in 2 cases (5%) .These 2 cases occurred in Group II, the 1st mortality was due to bronchopneumonia during the 1st postoperative week and the 2nd mortality was due to pulmonary embolism during the 2nd postoperative week.

Table (VIII): Correlation between preoperative neurological status (motor power) and surgical outcome.

Preoperative motor power	Total No. of the patients	%	Outcome					
			Favourable		Unfavourable		Mortality	
			No.	%	No.	%	No.	%
- No motor wakness.	5	125	5	100	-	-	-	-
- Partial paralysis*	30	75	22	73	6	20	2	7
- Complete paralysis	5	125	1	20	4	80	-	-
- Spastic paralysis**	3	60	1	33	2	67	-	-
- Flaccid paralysis	2	40	-	-	2	100	-	-
Total	40	100	28	70	10	25	2	5

This table shows that:

Favorable outcome was (100 %) in patients who had no motor weakness and (73%) in those who had partial paralysis, whereas it was 20% in patients who had complete paralysis.

Of the 3 patients presented with complete spastic paralysis, only one of them showed favorable outcome (33 %), and the other 2 showed unfavorable outcome (67 %), these 2 patients were at 6th and 7th decade of life. Whereas the 2 patients who had complete flaccid paralysis showed unfavorable outcome (100 %).

Table (IX): Correlation between tumour pathology and surgical outcome.

Tumour pathology	Total No. of the patients	%	Outcome					
			Favourable		Unfavourable		Mortality	
			No.	%	No.	%	No.	%
I- Group I	30	75	27	90	3	10	-	-
1- Meningiomas	16	54	15	94	1	6	-	-
2- Neuorfibromas	12	40	10	83	2	17	-	-
3- Haemangioma	1	3	1	100	-	-	-	-
4- Arachnoid cyst	1	3	1	100	-	-	-	-
II- Group II:	10	25	1	10	7	70	2	20
1- Astrocytomas	5	50	-	-	4		1	20
2- Ependymomas	4	40	1	25	2	80	1	25
3- Dermoid cyst	1	10	-	-	1	50	-	-
						100		
Total	40	100	28	70	10	25	2	5

This table shows that:

- Favorable outcome was 90% in Group I, whereas it was 10% in Group II.
- Favorable outcome in patients with meningiomas and neurofibromas was 94% and 83%, respectively.
- Unfavorable outcome was present in patients with astrocytomas, whereas 25% of patients with ependymomas had a favorable outcome.
- Mortality was 20% in Group II, whereas there was no mortality in Group I.

Table (X): Final outcome of the patients.

Outcome	Group I		Group II	
	No.	%	No.	%
I- Favourable outcome	27	90	1	10
II- Unfavourable outcome	3	10	7	70
III- Mortality	-	-	2	20
Total	30	100	10	100

This table shows that:

- Favorable outcome was 70% of all the patients, whereas unfavorable outcome was 25% and mortality was 5%.
- Favorable outcome was 90% in Group I, whereas it was 10% in Group II.
- No operative mortality among Group I, whereas there were 2 mortalities in Group II, due to pneumomia in one patient and pulmonary embolism in the other.

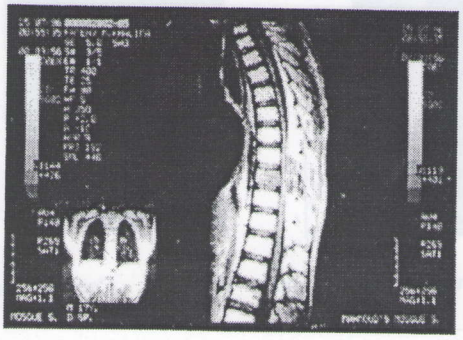


Fig. 1: Dorsal haemangioma

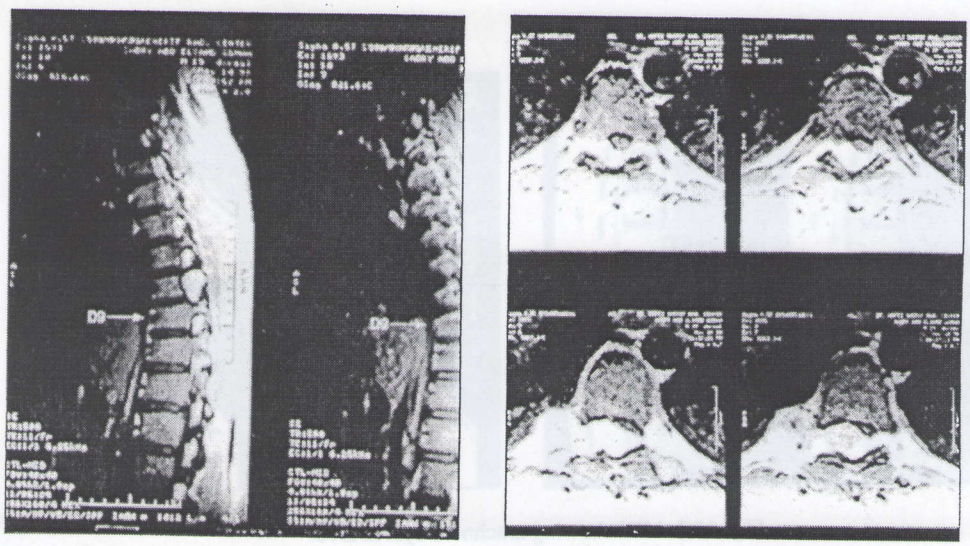


Fig. 2&3: Dumbell shaped Neurofibroma at D 8 & 9.

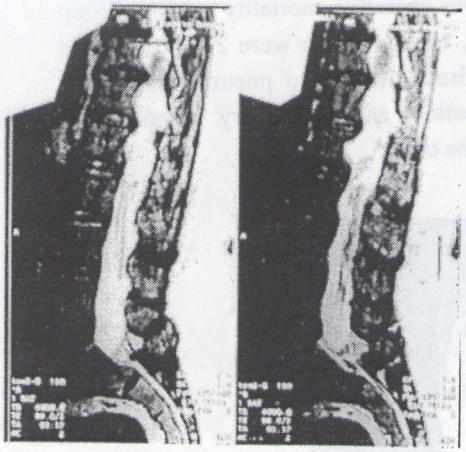


Fig. 4: Ependymoma of conus medullaris and filum terminale.

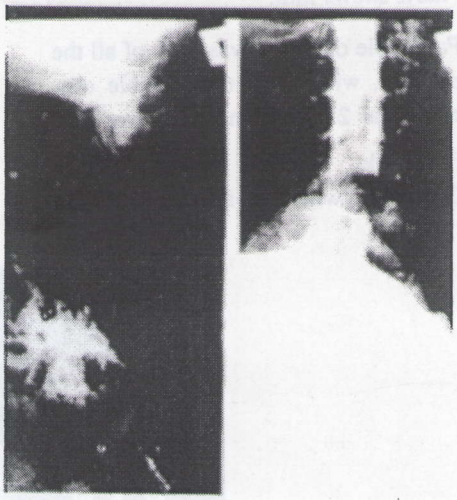
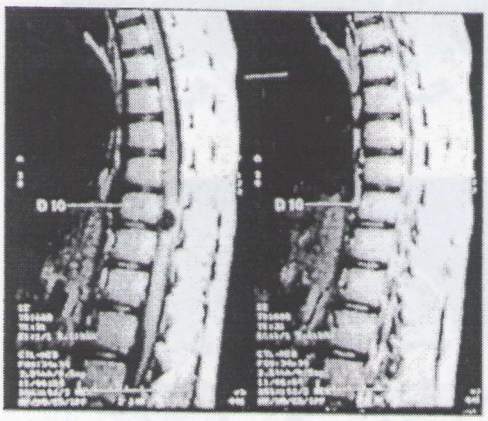


Fig. 5: Cervical myelography showed neurofibroma.



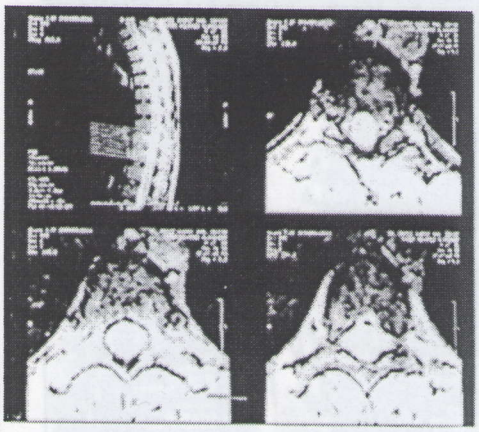


Fig. 6&7: MRI showing arachnoid cyst at D10.

Discussion

The first successful removal of an intradural extramedullary spinal tumor was performed more than 100 years ago by Victor Horsley [8].

Cushing and Eisenhardt [9] stated that a successful operation for a spinal tumor represents one of the most gratifying of all operated procedures. Even profound neurological deficit that would be hopeless, in a case of gradual damage to the spinal cord, can recover with careful surgical intervention [10].

In the present study, we reviewed 40 patients presented with spinal cord tumors (30 intradural extramedullary and 10 intramedullary tumors). Although analysis of the reported results was difficult to interpret because of the small number of the patients, numerous previous epidemiological reports [2, 10,15,30] are nearly parallel to this study.

Meningiomas and neurofibromas were the most common benign tumors in our series, (40 % and 30% respectively). The peak incidence occurred in the 5th and 6th decades of life (27% and 37% respectively), with predominance of females (77%).

Females constituted 78% of patients with intradural extramedullary tumors, whereas it was 80% in the series that report by Levy et al. [10], and 85% in the series of Bull [11]. In this study, 77% of the patients with intradural extramedullary tumors were located in dorsal region and only 17%in cervical region. These finding are nearly similar to that reported by Levy et al. [19] where 73% of their cases were located in dorsal region and 17% in cervical region. Other author [2,20] reported 80% in dorsal region of their cases.

Intramedullary tumors represented 25% of our series, which are higher than those reported by Simeone [4] which was 16%. In this study most of them were astrocytomas and ependymomas (50% and 40% respectively). Some authors [12,13, 14,15] reported this similar predominance of astrocytomas. Stein [16] reported the equal frequency of these tumors, whereas Cooper and Epstein [12] reported an incidence of 48% of ependymomas and 37% astrocytomas in a series of intramedullary tumors.

In this study both sexes were of equal ratio in patients with intramedullary tumors. El-Hefny et al. [38] reported the similar ratio, whereas Cooper and Epstein [12] reported 62% males and 38% females in their study.

In this study, 40% of intramedullary tumors were in cervical region, and 60% of astrocytomas were located in that region. Cooper and Epstein [12] reported 69% of their intramedullary tumors were located in the same region.

Cohen [17] reported a nearly similar results that 12 of 19 cases of astrocytomas (63%) were located in cervical region. In this series 50% of ependymomas were present in the conus medullaris whereas Simeone [4] and Stein [16] reported that over 50% of ependymomas were reported to be in the conus medullaris and filum terminale, whereas McCormick et al. [15] reported that most cases of intramedullary ependymomas in their series were located in the cervical and cervico-dorsal region.

In the present study motor deficits were present in 87.5 %, partial weakness in 75% and complete paralysis in 12.5%, back pain in 70% and sphincteric disturbances in 35%. Levy et al. [10] found nearly similar percentage in their series (motor deficits in 82%, back pain in 72% and sphincteric disturbances in 40%), whereas Sloof [1] reported that sensory deficits were more frequent than motor deficits (69% and 17% respectively), and sphinecteric disturbances in only 3%.

Because most intradural extramedullary tumors are benign, usually the surgical goal is complete resection [18].

Cushing and Eisenhardt [9] wrote that the removal of these frequently benign tumors was one of the most gratifying of all operative procedures.

In this series complete removal of meningiomas was achieved in all cases (100%) and complete removal was achieved in 11 (92%) of cases of neurofibromas, whereas in the remaining one case incomplete removal was performed due to extraspinal extension. In Soleros series [20], complete removal of meningiomas was achieved in 168 patients (97%), and Levy et al. [10] reported that complete tumor removal was achieved in 82% in their series of spinal meningiomas. As for neurofibromas, complete removal could be achieved without persisting neurological deficits even if the affected nerve root is resected [21].

Intramedullary spinal cord tumors remain a particulary difficult entity to treat surgically. Radical removal of these tumors can be achieved by experienced surgeons with minimal morbidity. However, most spinal cord astrocytomas in adults are infilterating, not totally resectable, and recur frequently. For many ependeymomas, surgical cure is possible, but for infiltrating ependymomas and other spinal cord tumors of glial origin, improved management awaits a better understanding of the biology of these lesions [34].

In cases of intramedullary tumors, radical removal can usually be achieved in ependymomas and low grade astrocytomas. Recently, many authors have been enthusiastic about radical removal of intramedullary tumors [15].

In cases of Extensive astrocytomas gross total removal is not possible and postoperative treatment is recommended [22]. In such cases it is advisable to per-

form a posterior myelotomy extending over the full length of the tumor, remove the tumor tissue that is clearly demarcated from the healthy tissue and leave the duramater open [23]. Only 2 out of 53 cases of astrocytomas in the series of Guidetti [23] had complete tumor removal. Epstein and Epstein [24] reported that in 3 out of 19 patients of astrocytomas, the tumor was soft and easily succable and there was a very clear plane of cleavage, thus the tumor was completely removed. Cooper and Epstein [12] achieved total removal in 7 of 11 cases of astrocytomas, whereas Cohen [17] achieved radical tumor removal in all their 19 cases of intramedullary astrocytomas with the aid of CUSA and laser. Laser was not available for use in this study, where total excision was achieved in one case (20%) out of 5 cases of astrocytomas, a plane of cleavage was visible between the cord and tumor, while in the other 4 cases, subtotal removal was achieved in 2 cases (40%) where a plane of cleavage was indistinct and biopsy in the remaining 2 cases (40%), where the tumor was infiltrating the cord.

Some authors [12,26] believed that radiation therapy should have a major role in the management of intramedullary tumors of glial origin. Kopelson et al. [27] believed that operation should consist of the minimum surgical procedure necessary to establish a diagnosis, and that radiation therapy is most effective treatment modality. Fischer et al. [28] stated that the efficacy of radiation therapy has not been proved, and Cooper and Epstein [12] believed that biopsy and irradiation of intramedullary tumors represent suboptimal therapy. Epstein and Epstein [29] and Post and stein [30] believed that radiation therapy is unnecessary in cases of astrocytomas where radical excision is accomplished and that radiation therapy may be indicated in cases of subtotal excision or following a biopsy.

In our study, the cases of astrocytomas which were subjected to subtotal excision or biopsy [4 cases (80%)], had postoperative, radiotherapy, while it was unnecessary in one case (20%) where total removal of the tumor was achieved.

Epstein et al. [25] stated that ependymomas do not infilterate adjacent neural tissues and surgical interference is invariably possible, so gross total removal may be accomplished in the overwhelming majority of patients. They achieved a gross total removal of ependymomas in 37 of 38 patients (97.4%). In this study, total removal was achieved in 2 (50%) out of 4 cases, where a plane of cleavage was clear between the cord and the tumor, whereas subtotal removal was achieved in the other 2 cases (50%) where a plane of cleavage was indistinct followed by postoperative radiotherapy.

In the present study 9 patients (22.5%) showed postoperative deterioration in their neurological status, different authors sush as Stein [16] and Hermann et al. [31] reported nearly similar percentage (22.6%) and 20% respectively), whereas Cooper and Epstein [12] reported different percentage (28%).

12.5% of our cases had wound infection and 10% had CSF leakage. Hermann et al. [31] reported that 26.6% of their patients had postoperative C.S.F leak while Cooper and Epstein [12] had wound breakdown and C.S.F leak only in 10% of their patients.

In this study there were 2 postoperative mortalities (5%), that occurred in Group II due to pneumonia in one patient and pulmonary embolism in the other.

Levy et al. [10] reported 3% operative mortality in cases of intradural extramedullary tumors.

Hermann et al. [31] reported 6.7% mortality in the early postoperative period due to pneumonia and heart failure.

The immediate results and future prognosis in the common intradural - extramedullary tumors, including meningioma and nerve sheath tumors, have been well established. These tumors are benign, and if they are carefully and thoroughly removed, the patients should be cured with excellent prognosis [30]. The outcome is related primarily to the patients preoperative condition, as well as patients age [12,30].

In this study the clinical outcome was influenced to a large extent by the preoperative neurological status, tumor pathology and patient's age. As favorable outcome was 100% in patients who had no motor weakness and 37% in those who had partial motor weakness, whereas it was 20% in patients who had complete paralysis (5 patients). Of the 3 of them who presented with complete spastic paralysis, only one showed partial improvement, and the other 2 patients deteriorated, these 2 patients were at 6th and 7th decade of life. Whearas the other 2 patients that had complete flaccid paralysis did not improve. The results of the other previous studies showed a similar issue [3,33].

Levy et al. [10] reported that 20% of patients who could walk preoperatively were walking at long-term follow-up review, 2 of their 6 paralysis were walking with assistance and three others had recovered some movement . Iraci et al. [39] reported that one of three preopratively paraplegic patients was able to work. Only 10% of intramedullary tumors in our series showed Favorable outcome.

Cooper and Epstein [12] reported that in 72% of patients with intramedullary tumors, the lower extremity motor function was stabilized or improved postoperatively. Hermann et al. [31] reported 73% of

patients with unchanged neurological condition and only 6.6% with improvement.

CONCLUSION:

It is apparent, that the clinical outcome of the patients of intradural cord tumors is influenced to a large extent by the preoperative neurological status, tumor pathology and patients age.

We emphasise the importance of using MRI in diagnosis, CUSA unit, and microneurosurgical tools in surgery.

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