4 — Les radiographie simples, montrent en générale les lésions osseuse.

5 — La Myelographie, a une valeur de diagnostic plus importante dans les régions cervicales et dorsales (85%) que lombaire.

6 — La paraplegie Pottique constitue 80% des cas. Dans ces cas, les pluparts situé dans la région dorsale, la laminectomie semble être plus éfficace que costotrans. versectomie ou la decompression anterolaterale.

7 — L'intervention suivie de la radiotherapie semble être plus éfficace que la radiotherapie seule.

8 — Les chrdomes, ont tendance de recidiver, ceci semble être plutôt la consequence d'une exeres incomplete, qu'une transformation maligne.

9 — Les lesions osseuses, suspect d'être secondaires, avec l'absence apparente de foyer primitif, sont operées pour la decocpression et diagnostic.

10 — La decompession de la moelle dans les cas de l'hernie discale de la région dorsale, semble être la méthode la plus.

11 — Deux cas d'abcès extraduraux consecutifs à une fonction lombaire sont rapportés.

12 — Angiomes ou angioendotheliomes constituent 14% des cas. Leur exerse complete ou partielle, suivie de radiothérapie est satisfasante.

13 — Plus que 50% des neurofibromes extraduraux ont une expension extrayertebrale (Bisac).

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ON THE CLINICAL PICTURE AND PROGNOSIS OF MALIGNANT EXTRADURAL TUMORS OF THE SPINAL CORD

K. Nittner, M.D. ★

The malignancy fo spinal tumors is frequently a relative matter and may sometimes be ascertained only through clinical study.

In judging the degree of malignancy of tumors one is inclined to consider only the morphological point of view. The relative characteristics are frequent mitotic figures, infiltrative growth, and a tendency to metastasis.

We know that real spinal cord tumors, i.e., gliomas and ependymomas, rarely form metastases, not even in the meninges; and furthermore, that in some cases they do not even acquire infiltrative properties. So, with these tumors it depends essentially on how fast they grow.

In cases of rapid growth the clinical picture is characterized by short histories, acute or sub-acute courses, and flaccid paraparesis or paraplegia. So, their relative malignancy may be judged solely from clinical investigation.

This is, however, usually not the case with malignant extradural tumors of the spinal cord, which are mostly sarcomas and carcinomas, nor with malignant tumors of the vertebral column. But among these malignancies of the spinal cord there is one group of sarcomas and certain skeletal tumors occurring in the epidural region which are distinguished from others because of clinical and prognostic characteristics.

In the literature we find rather divergent data concerning the relative frequency of malignant extradural tumors: BALLATINE and DADIN, with 25% of their entire series give the highest rates; TON-

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NIS and NITTNER give 17.8%; ELSBERG 11.4% and WOLTMANN, KERNOHAN and ADSON 11.2%.

Figure 1 shows a synopsis of the spinal cord tumors during a 30-year observation by TONNIS. The grouping principle is that of their site of origin, that is, whether they arose from the vertebral column or in extradural, juxtamedullary, or intramedullary regions. It can be seen that of 359 tumors 10% were vertebral-column tumors, 16% had developed in extradural regions, and 6% occurred both in extradural and intradural regions at the same time.

Further subdivision, according to the different levels of the spinal canal, shows that in 66 cases (that is almost 18%) we are concerned with malignancies (figure 2). On the other hand we find 52 gliomas and ependymomas (15%), 82 meningiomas (23%), 77 neurofibromas (19%), 27 angiomas (7%), 21 embryological tumor (6%), and finally, 34 miscellaneous compressive lesions (10%).

In this summary we are interested exclusively in the malignant group, namely cases which expand primarily or secondarily in the epidural space or originate from the vertebral column. Among the epidural tumors, sarcomas and secondary tumors are usually considered ones which infiltrate into the epidural space, chiefly metastasizing adenocarcinomas.The chordoma, met with mainly in the cancer age, should likewise be considered as a malignant tumor which can metastasize. Occasionally benign tumors such as meningiomas and neurofiromas undergo malignant changes.

Among tumors of the skeletal system are also included the clinically malignant diseases of the hematopoietic system such as different forms of lymphatic and myeloid leukemia, lymphogranulomatosis, the less frequent reticulo-cell-sarcomas, and the leukocytically related chloroma. Finally, two groups of clinically interesting tumors remain: the myeloma, or plasmocytoma, and the giant cell tumors.

In our material the 79 epidural tumors were as follows (figure 3):

1. sarcomas—23 cases, or 29%
2. secondary carcinomas—11 cases, or 14%

These form a total of 34 cases, or 43%; the malignant vertebral column tumors totaled 27 cases, or 78%.

In this large number of malignant spinal tumors there is one group of sarcomas that differs from other types of sarcomas by its clinical behavior and its favorable prognosis. This type appears to arise primarily from tissues in the epidural space. Clinically, they usually present a history of up to three years or longer. In some cases we can

Topography of the Tumors

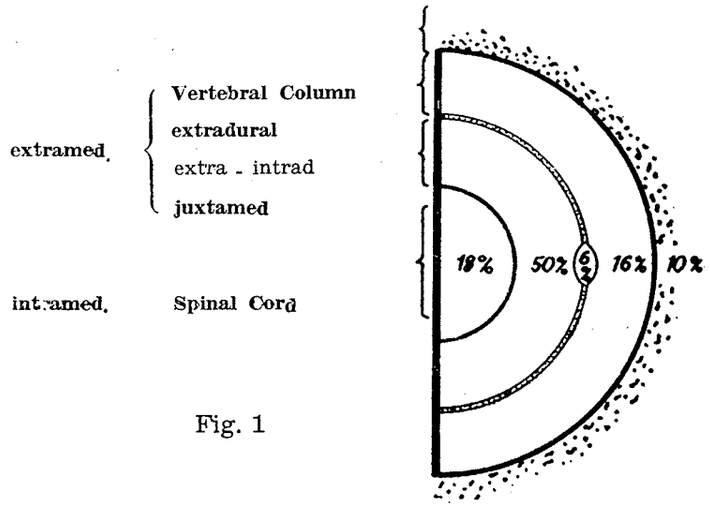

Fig. 1

359 CASES OF SPINAL CORD COMPRESSION

	total Nr.	Malignomas	Gliomas Ependymomas	Meningiomas	Neurofioramas	Angiomas	embryo-logical Tumors	Compression of other types
Medulla oblongata	13	—	9	2	—	1	—	1
Cervical	79	11	11	20	26	6	2	3
Thoracical	214	46	22	60	41	18	9	18
Lumbo-sacral	30	7	4	—	3	1	6	9
Cauda	25	2	6	—	7	1	4	3
	359	66	52	82	77	27	21	34

Fig. 2.

believe that they are secondary malignantly transformd tumors. The clinical picture is characterized by tenderness on pressure and movement of the spinal column, segmental pains, spastic paralysis which may be flaccid, accompanied by rapidly developing sensory, bladder and sphincteral disturbances. Not in every case is the history short and the paralysis flaccid, in spite of the histological signs of malignancy. However, we may sometimes find a lack of motor changes and vegetative disturbances with sensory disturbance being the only objective finding. From the radiological point of view bone manifestations may be lacking, and in about half of the cases the bone is spared.

These sarcomas are distinguished at the operation by their labile fleshy appearance. They lend themselves to easy removal from the often intact dura and allow total macroscopic dissection. Histologically they usually show a small cell sarcoma which simulates the nuclear groups of reticulo-cell-sarcomas, as well as more differentiated sarcoma types.

The follow-up period for all cases of sarcomas was between 4 weeks and 8 years (figure 4). The average survival period in our cases amounted to $4\frac{1}{2}$ months. Two thirds of the sarcomas demonstrated an unchanged clinical picture following a long interval after the operation. In half these cases the tumor had been macroscopically totally removed. Whenever a post-operative deterioration occurred, it frequently was fatal.

Remarkably, however, we found that one third of our cases survived more than the nine-month limit; and there were also the unbelievable survival periods of 2 to 8 yars. We could prove with certainty that these cases belonged to the above described slow-growing group of sarcomas.

The carcinoma metastases had the shortest courses. More than half of these cases died during the first postoperative month. The longest survival and follow-up periods did not reach more than 3 months.

Differential diagnosis: Meningiomas, neurofibromas, and also spongioblastomas commonly occur in the cervical region. Ependymomas and spongioblastomas on the contrary are rare in the thoracic region. There was a manifest predisposition for ependymomas and neurofibromas (with exception of embryological tumors) in the conus and cauda regions.

Since it is frequently difficult to assess preoperatively the extradural sarcomas, owing to the frequent absence of vertebral changes, we can deduce from the above mentioned data the following:

359 cases of spinal cord compression

79 Extradural Compressions

23 Sarcomas 29%

11 Carcinomas 14%

43%

36 Vertelral column- tumors

27 Malignomas 78%

Fig. 3.

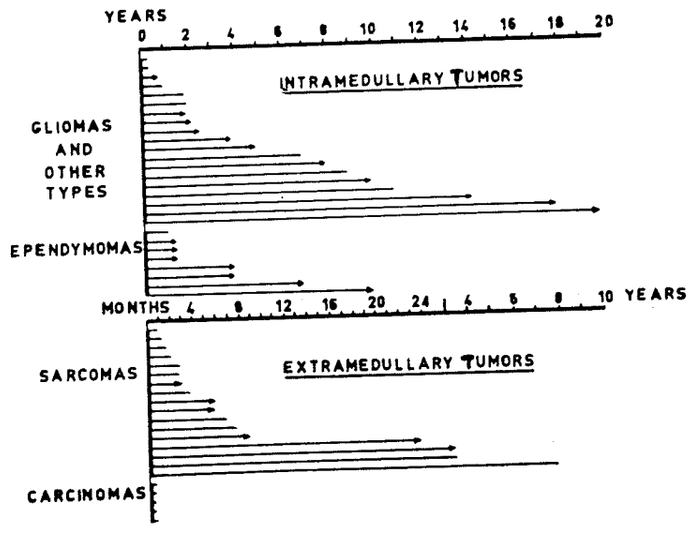

Fig. 4.

In the cervical region the probability of spinal sarcoma is quite low. In the dorsal region we must consider in addition to meningiomas and neurinomas, also sarcomas, while glial tumors (ependymomas and spongioblastomas) occur here rarely.

In cases of tumor in the regions of conus and caude we find more neurofibromas and ependymomas than meningiomas or sarcomas. We can also obtain preoperative data which helps in diagnosis from the length of the history, age, sex, and clinical picture. Short case histories are here the rule; longer case histories indicate a benign tumor. In cases with acute onset of transverse section syndrome with a short history we must exclude the possibility of epidural spinal abscess and of epidural hematoma.

Points which favor the diagnosis of sarcoma are: young patients, male sex, tenderness on pressure and movements of vertebral column, and segmental pains. Short histories with rapidly-developing spinal compression and usually early onset of flaccid paralysis favor the diagnosis of malignancy. The X-ray pictures are negative in half of the cases. If signs of bone involvement occur, we are concerned with pressure effects which extend over several segments. The cerebrospinal fluid protein content is usually markedly raised, and usually reveals a complete or partial spinal block. The sedimentation rate is usually only slightly raised and can be normal in children. Among the sarcomas of the epidural space one group is characterized by its special clinical course and is histologically very similar to reticulocell-sarcoma.

In our case material were also found tumors of the vertebral column with special clinical courses. Of these 9 were plasmocytomas and 4 were giant cell tumors. The 9 plasmocytomas were, at least at the time of operation, localized swellings; 8 were dorsal, almost all in the level of D2 to D5; only one case occurred in the upper cervical region.

Of these plasmocytomas 4 patients survived from 1 year up to 14 years, while the remainder died within the first three months after the operation.

The treatment of choice appeared to be an extensive laminectomy together with post-operative deep X-ray therapy and anti-cancer therapy. In the cases that were followed up by us, one was unique in that the plasmocytoma was lying exclusively extradural - such an occurrence has only once before been recorded in the world literature by STRATEMEYER (1950).

Of our 4 cases of giant cell tumors 2 were semimalignant, and 2 absolutely benign. According to the literature these occur only in the third and fourth decade. Our patients were afflicted at the age of 5, 15, 16, and 31 years respectively.

It was always found that the upper and middle thoracic segments were affected. The length of history varied from 1 month to 1 year. Clinically. there existed in our cases in addition to paravertebral soft tissue infiltration also signs of spinal compression, though mostly in an incomplete form. The neurological symptoms occurred usually on one side or more on one side than the other. In all our cases we found at the site of the tumor local tenderness together with a significant gibbus formation. The changes in cerebrospinal fluid and Queckenstedt were dependent on how much narrowing of the spinal canal had already occurred. Radiologically we usually found unilateral local destruction of the vertebra, which first affected the arch. The border was in every case sharply defined, but without sclerosis.

It was also found that the method of choice for treatment would be removal of as much of the swelling as possible. During our rather short periods of observation and follow-ups we found that the neurological manifestations had all regressed.

A differentiation between the semimalignant and the benign forms is radiologically not possible. Only through pathological examination and clinical course can that be ascertained.

All our malignant spinal tumors, including extradural and vertebral column tumors, were found mainly in the upper dorsal region (figure 5). The ratio in our case material between the 3 main spinal segments is as follows: sarcomas 1 : 4 : 1; carcinomas 1 : 5 : 0; other malignant tumor 0 : 2 : 1; and only intramedullary gliomas 3 : 2 : 1.

What was striking in all these malignant tumors was the fact that the most common site for these tumors was in D3 to D5. Therefore, we may accept the fact that this segment of the spinal cord (figure 6) cerresponding to the confines of medullary blood supply from the hemodynamic aspect is especially endangered in regard to tumor manifestations.

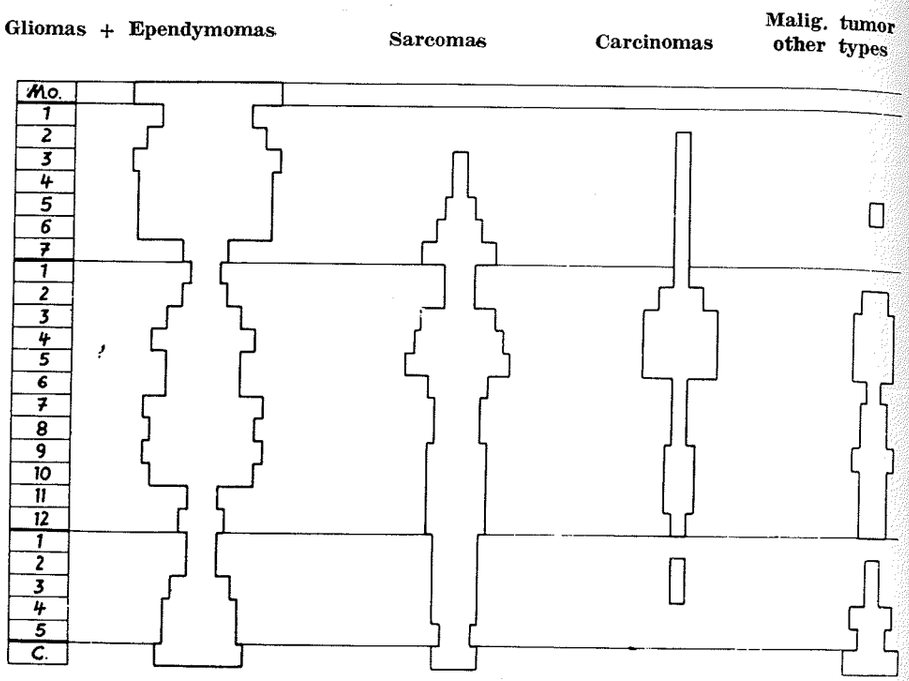
Gliomas + Ependymomas
Sarcomas
Carcinomas
Malig. tumor other types
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Fig. 5.

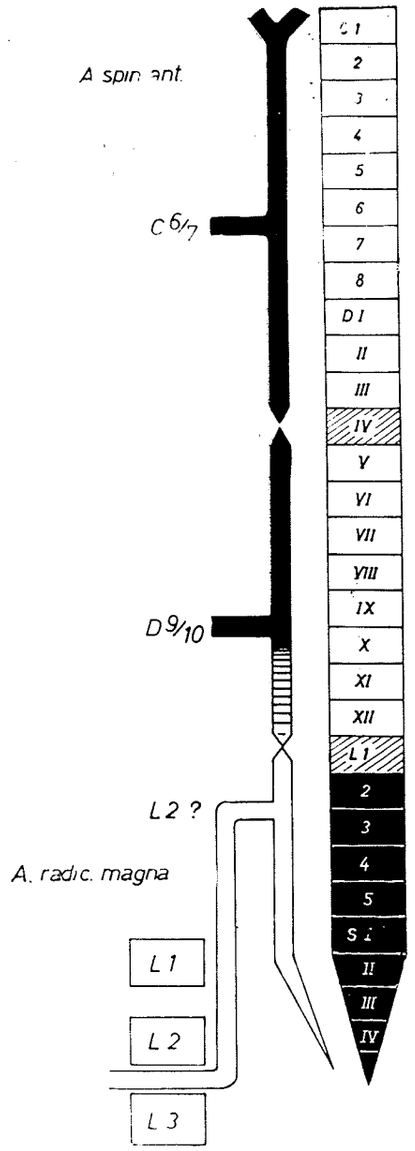
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Fig 6

Résumé

L'auteur après avoir fait un tour d'ensemble sur les caracteristiques de 359 cas personnels des compressions medullaires d'origine tumeurale, se penche sur 79 cas de tumeurs epidurales et 37 cas de tumeurs extradurales.

Après avoir discuté avec detail sur leur nature, localisations et leur traitements, en individualise plusieurs groupes qui sont caracteristiquespar leur lenteur d'évolution et les possibilités therapeutiques.

Ce qui frappe le plus l'auteur est le lieu d'élection des tumeurs malignes qui se situe au niveau de D3 — D5. Il en conclut que ce segment de la moelle, se trouvant dans la confine de la vascularisation medullaire est plus disposés au developpement des tumeurs malignes.—

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EXTRADURAL COMPRESSION OF THE SPINAL CORD AND CAUDA EQUINA

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The following is a brief survey of extradural compression cases admitted to the Pahlavi Hospital, Neurosurgical Service (Prof. Ameli) during a period of five years (1958-1963).

There were 124 cases during this period, and the etiological factors are shown in Table 1. In 35% of the cases the central disk was responsible for the compression; in 26% Pott's disease; 15% fracture and dislocation of vertebrae; 8.9% hydatid cyst; 6% malignant tumor (Primary and metastatic); 3.9% hemangioma of vertebrae; 1.9% neurofibroma; 1.9% epidural abscess and 0.9% chondroma. In addition, there was one case of compression by an osseous band.

Central Disk

There were 44 cases of central disk protrusion. (Lateral disk cases are not included in this series.) The age of the patients varied from 20 to 60 years. 60% of these patients were farmers and 20% were truck drivers.

As to location of the lesions, there were 28 lumbar ,14 cervical, and 2 thoracic disks.

In two cases, one in the lower dorsal and one in the upper lumbar region, the patients had sudden, severe pain in the spine and paraplegia in which the sequestrated disk had moved posteriorly. As soon as the laminae were removed, there was a large, loose cartilaginous mass. Inspection of the anterior part of the dura showed a fairly large opening in the annulus fibrosis. Both these patients had a partial recovery of function following the operation.

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