Neurofibrosarcoma of the cauda equina

Case report

RAPHAEL T. W. M. THOMEER, M.D., GERARD TH. A. M. BOTS, M.D.,
HANS VAN DULKEN, M.D., WILLEM LUYENDIJK, M.D., AND PETER HELLE, M.D.

Departments of Neurosurgery, Neuropathology, and Radiotherapy, University Medical Center, Leiden, The Netherlands

A case of a malignant neurofibroma of the cauda equina is described. Its location seems to be rare. An extensive surgical resection including the adjacent neural tissue and the enveloping dural sac was carried out, followed by radiotherapy.

Key Words: intrathecal neurofibrosarcoma • malignant schwannoma • cauda equina tumor • spinal cord resection

Malignant deterioration of a neurofibroma is well known, especially in the course of von Recklinghausen's disease. This malignant degeneration is estimated to occur in about 3% of cases. The figure of 13% given by Hosoi is generally considered as being too high. Half or possibly more of the malignant neurofibromas are associated with von Recklinghausen's disease. These figures pertain to tumors arising from peripheral nerves. Malignant neurofibromas localized within the spine are rare, and we have not found such a case reported in the literature. For this reason it seems appropriate to present the following case history.

Case Report

This 42-year-old hospital night porter was admitted to the University Hospital, Leiden, in March, 1977. For more than 9 years he had suffered from low-back pain, sometimes radiating into the left calf. This forced him to give up his job as a construction worker and to become a night porter. Four months before admission, he had to completely stop even this work because of unremitting pain. Complete impotence had developed, and in the 6 weeks before admission he had to strain while voiding. Weakness of the legs was noted for 3 weeks, culminating in inability to walk 1 day before admission.

Examination. The lumbar lordosis was flattened and mobility was severely restricted. A hypotonic paraparesis existed, with fasciculations in both calves. Hypesthesis was noted in the S3–S5 dermatomes on the left. Tendon reflexes of the legs and the anal reflex were absent. Bilateral flexor plantar responses were found. Catheterization of the bladder yielded 1700 cc residual urine. There were no superficial stigmata of von Recklinghausen's disease.

Laboratory data were within normal limits. Radiographic studies revealed no anomalies except for a transitional L-5 vertebra. Cerebrospinal fluid (CSF) obtained at the vertebral level of L5–S1 showed Froin's syndrome. After the injection of metrizamide (Amipaque), an irregularly shaped total block was found immediately above the L5–S1 level. Subocipitally administered contrast medium showed the upper border of the block at the T12–L1 level, also with an irregular configuration. No tumor cells were seen in the cisternal CSF.

First Operation. On the day of admission, laminectomy of T-12 through L-4 was carried out. After the dura mater was opened, a gray-blue, soft, vascular tumor protruded over the whole area. The conus medullaris and most of the caudal roots could not be separated from the tumor mass. Exploration was confined to biopsies, and a decompression was achieved.
Histopathology. In most areas, the tumor showed a high cellular density (Fig. 1). The cells were pleomorphic and arranged without a specific pattern. Most cell nuclei were swollen and showed a moderate amount of chromatin. Dispersed pyknosis and karyorrhexis were seen. In each of the 50 high-power fields, 10 mitoses were observed. Some necrotic areas were seen here and there. In other areas, the tumor was less cellular, showing the typical features of a neurofibroma, with a wavy pattern of fibrillary cells and elongated, somewhat cigar-shaped nuclei (Fig. 2). No mitotic figures were present in these parts. The histopathology of this tumor was consistent with a Grade II neurofibrosarcoma.15

First Postoperative Course. Once malignancy was established, a thorough search for metastases was made. These investigations (including blood chemistry and tomography of the lungs) were negative. It was decided to subject the patient to a second operation with the hope of achieving a complete removal as possible of the malignant tumor. This decision was based on the following considerations which were reviewed with the patient: 1) the residual functions of the medullary conus and the cauda equina were threatened in any event; 2) the increasing pain might be relieved; 3) unavoidable spilling of tumor cells might be reduced; and 4) after removal of the mass of tumor, it was possible that radiotherapy would be more efficacious.67

Second Operation. Twelve days after the first operation, an additional laminectomy of T-11 and L5-S4 was performed. The spinal cord was transected at the epiconus level, where the distal end of the dural sac was carefully closed. Caudal to this level, a complete exenteration en bloc of the entire contents of the spinal canal was carried out, including the dural sac which had previously been ligated at its upper end. The lumbar and sacral roots were cut through at the level of the intervertebral foramina and marked with silver clips. The tumor showed exactly the same histopathological features as described before. No tumor was found in the removed part of the spinal cord.

Second Postoperative Course. A few days after the operation the pain had almost totally disappeared. Neurological examination revealed complete motor and sensory loss below the T-12 level. Radiographic studies showed no vertebral instability. The patient could be mobilized in a wheelchair 12 days after the operation. Radiotherapy was given, with an 8-MV photon linear accelerator.† The target area encompassed the spinal canal from vertebral level T-11 to S-4. A tumor dose of 6000 rads was given. The target area was irradiated by two dorsal wedge fields. The maximal skin dose was 3000 rads. The daily dose was 250 rads, four times a week, with an overall time of 6 weeks.

For 3 years the patient did well, but then he started to complain of increasing pain in the dorsal region. Neurological examination revealed anesthesia below the T-8 level. X-ray films showed a paravertebral tumor mass from T-9 to T-11, with partial destruction of the T-10 and T-11 vertebrae. On myelography, a total block with the configuration of an intra-

†Accelerator manufactured by Toshiba Co. Ltd., Japan.
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medullary lesion was found below the T6-7 level. Furthermore, the lower lobe of the right lung was clouded by a pleural effusion, which was evacuated by puncture. No tumor cells were found in this material. A total body scan also showed the paravertebral tumor mass without signs of pulmonary parenchymal metastasis. Laboratory data were still within normal limits, with the exception of a raised erythrocyte sedimentation rate.

The patient has been treated with infusions of 120 mg adriamycin at 3-week intervals, with a total dose of 1080 mg. His condition has not changed up to now, and the total body scan shows an unchanged picture. Small doses of simple analgesic drugs are efficacious.

Discussion

Neurofibrosarcoma is a rare tumor, especially when localized within the spinal canal. The literature does not provide us with reports on cases of (neuro-) fibrosarcoma of the cauda equina. However, it is hard to believe that our case should represent the only one with this rare localization. Other cases may have been incorporated in larger series of fibrosarcomas. On the basis of the expected progressive loss of neurological functions, as well as the intractable pain in this patient, a radical extirpation of the tumor, including the adjacent neural tissue and the dural sac, was attempted. It was followed by high-dose radiation therapy.

Although the operative procedure was as radical as possible, the tumor recurred 3 years later. It was localized just above the original operative field, and involved the lower dorsal vertebrae (T-10 and T-11). Myelography revealed an intramedullary lesion below the T6-7 level. This was surprising and disappointing in that no indication of tumor was found in the part of the spinal cord removed at surgery. This experience demonstrates that apparent radical surgery of an intrathecal neurofibrosarcoma has led to a disappointing result. The surgery may not have been sufficiently radical, and/or spillage of highly malignant cells may have been the cause of recurrence.

References


Address reprint requests to: Raphael T. W. M. Thomeer, M.D., Department of Neurosurgery, University Medical Centre Leiden, Leiden, The Netherlands.