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Gliomas of the conus medullaris

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Six patients with histologically verified gliomas of the conus medullaris (two astrocytomas, two ependymomas, two myxopapillary ependymomas) were diagnosed and studied. There were four men and two women ranging in age from 23 to 47 years. Predominant initial symptoms were back pain (four cases) and leg weakness (two cases). The most common findings on admission were flaccid paparesis with impaired sensation and bladder dysfunction. Postoperative MR images with more than 95% removal of a tumour were defined as 'subtotal removal' (noted in two of six cases), and less than 95% as 'partial removal' (four of six cases). All patients had postoperative radiotherapy, and the two patients with an astrocytoma underwent chemotherapy. During the follow up period ranging from 2 to 7 years, there was no tumour recurrence or regrowth on MR images except in a patient with a malignant astrocytoma, who indeed died from intracranial dissemination 2 years after surgery. Adjuvant therapy following the excision of a spinal glioma is also discussed.

Keywords: intramedullary tumors; conus medullaris gliomas; radiotherapy; chemotherapy; outcome.

Introduction

Spinal cord gliomas comprise the majority of intramedullary cord tumours.[1] At the conus medullaris, the incidence was expected to be 26% out of all spinal cord gliomas.[1] They often show characteristic clinical, radiological, and intraoperative features which differ from gliomas involving other parts of the spinal cord. The purpose of this paper is (1) to describe the clinical, radiological and surgical aspects based on six patients with gliomas of the conus medullaris; (2) to report the longest 7 years' surgical follow up results with precise determination of either the subtotal or partial tumour removal by means of pre- and postoperative magnetic resonance (MR) images; and (3) to discuss adjuvant radiotherapy and chemotherapy after the removal of the glioma of the conus medullaris of the spinal cord.

Clinical materials and methods

From January 1982 to April 1992, 57 patients with intramedullary tumours were admitted and surgically treated, and 31 gliomas were histologically verified in our department. In six patients, the gliomas were located in the conus medullaris; there were four men and two women ranging in age from 23 to 47 years (mean 37 years). These patients form the basis of this study.

Clinical summary

The clinical course of all the patients was slow but progressive (Table I). Regarding the initial symptoms, four patients presented with back pain, one with monoparesis of the right leg, and one with paraparesis. Two patients had been diagnosed as having a herniated lumbar intervertebral disc and were operated on in another hospital. The duration of the symptoms was from 1 to 17 years (median 6 years). On admission, all patients had a flaccid paraparesis, gait disturbances, and sensory disturbances below the level of the tumour. Five patients had urinary bladder dysfunction.

Radiological study

A preoperative radiological evaluation included plain spine films and computerised

Table I Clinical summary of six cases of glioma of the conus medullaris

Case no.	Age (yrs)	Sex	Onset	Duration of symptoms	Motor	Neurological dysfunction	
						Sensory	Bladder
1	36	F	Monoparesis (rt leg)	1 year	Flaccid paraparesis (right > left)	Hypaesthesia (below L4 and L5)	–
2	39	M	Lumbago	4 years	Flaccid paraparesis (right > left)	Hypaesthesia (below L1)	+
3	23	F	Lumbago	2 years	Flaccid paraparesis	Hypaesthesia (below L4)	+
4	47	M	Lumbago	6 years	Flaccid paraparesis (left > right)	Hypaesthesia (below T8)	+
5	40	M	Lumbago	17 years	Flaccid paraparesis (right > left)	Hypaesthesia (below T11)	+
6	39	M	Paraparesis	8 years	Flaccid paraparesis	Hypaesthesia (below L5)	+

tomography (CT) in six patients, myelography and CT myelography in four patients, and MR images in five patients. On the plain spine radiographs, vertebral body scalloping, median pedicle erosion, and thinning of the laminae were noted in three cases with more than six years' duration of symptoms. MR images with Gd-DTPA enhancement clearly demonstrated large cystic compartments and a long cephalo-caudal extent of the tumours (Figs 1, 2). The longest extent was 17 vertebrae from T8 down to S2 (case 5, myxopapillary ependymoma) and the shortest was three vertebrae from T12 to L2 (case 3, ependymoma). On average, the extent of the tumour was 5.2 vertebrae. Spinal cord enlargement and exophytic growth of tumours were noted in all of the patients. Tumour cysts were defined in three cases. A summary of the neuroradiological findings is presented in Table II.

Surgical procedures

Surgery was performed with the patient in the prone position, with somatosensory evoked potential (SSEP) monitoring from the anal sphincter. Laminectomy was performed over the location of the tumour. After the laminectomy, ultrasonography was used to identify the tumour and the cyst.

With the aid of an operating microscope, a midline dural incision was made leaving the arachnoid intact. After cutting the arachnoid, a midline myelotomy was performed using sharp dissection with minimal coagulation of the small pial vessels crossing over the dorsal midline. Pial traction sutures were used to keep the myelotomy edge open. Tumour removal was carried out using the bipolar coagulator and fine dissectors.

Astrocytomas were so poorly defined from the normal cord that it was almost impossible to perform a total removal. Ependymomas were usually well circumscribed and easier than the astrocytomas to remove. However, at the level of the conus medullaris and filum terminale, the part of the exophytic enlargement of the tumour had strong adhesions with the nerve roots and the filum terminale (Figs 3, 4), which made total removal difficult. The tumour cysts were easily defined. They contained xanthochromic fluid, produced presumably from the tumour. The dura was closed hermetically.

The extent of surgical resection was determined by postoperative MRI. Subtotal removal (more than 95%) was achieved in two patients, and partial removal (less than 95%) was achieved in the remaining four patients.

Pathology

The pathological diagnosis was: two astrocytoma (grades 2 and 3 respectively), two

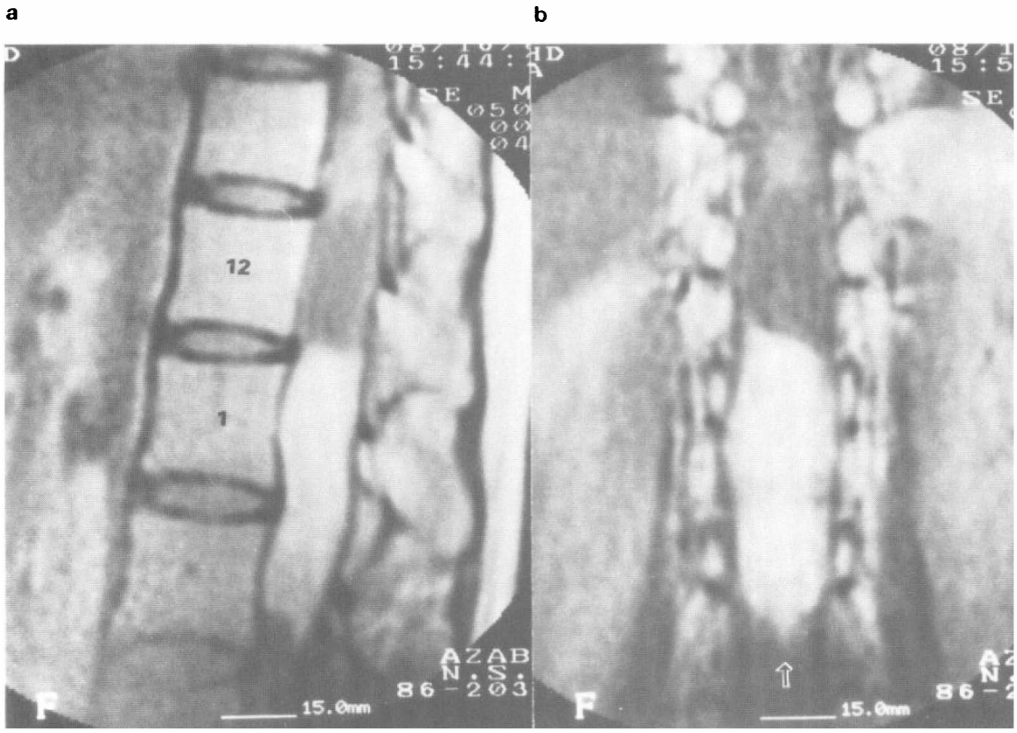

Figure 1 Case 3, ependymoma. T1-weighted MR images with Gd-DTPA enhancement showing the massive part of the tumour at the L1–2 level and the cystic part above it. (a) is sagittal and (b) is a coronal view. Note that the caudal end of the exophytic enlargement of the tumour is extending caudally in the midlongitudinal direction (arrow).

ependymoma, and two myxopapillary ependymoma. A summary of the location of the tumours, cyst, and cavities are schematically illustrated in Figure 5. Three out of four ependymomas had a cyst or cavity; one within the tumour (case 6), two in the cord cephalad to the tumour (cases 3 and 5). All six patients had exophytic growth of the tumour.

Postoperative therapy

All patients underwent spinal irradiation with a linear accelerator using a localised P–A field, covering the field of plus one to two vertebral levels above and below the lesion. Doses varied from 30 to 50 Gy (median 44 Gy) (Table III). The usual fractionation was 2.5 Gy/day, and all patients were irradiated 3 days a week.

Two patients with astrocytomas underwent chemotherapy. ACNU (1-(4-Amino -2-methyl -5-pyridinyl) methyl-3- (2-chloroethyl)-3-nitrosourea hydrochloride) was administered intravenously at a dose of 2 mg per kg of body weight. The first dose was given on the day when radiotherapy was started, and the maintenance dose of 1 mg per kg was given every 8 weeks after the first administration.

Results

The immediate postoperative results of the six cases were as follows: one improved, two unchanged, one worsened temporarily, and one worsened permanently. With a follow up from 2 to 7 years (Table III), five patients have all improved without recurrence or regrowth of a tumour in the MR images. In three cases (cases 2, 3, 4), both motor and bladder functions recovered fully and these patients returned to their previous work. Two patients (cases 5, 6) use a self

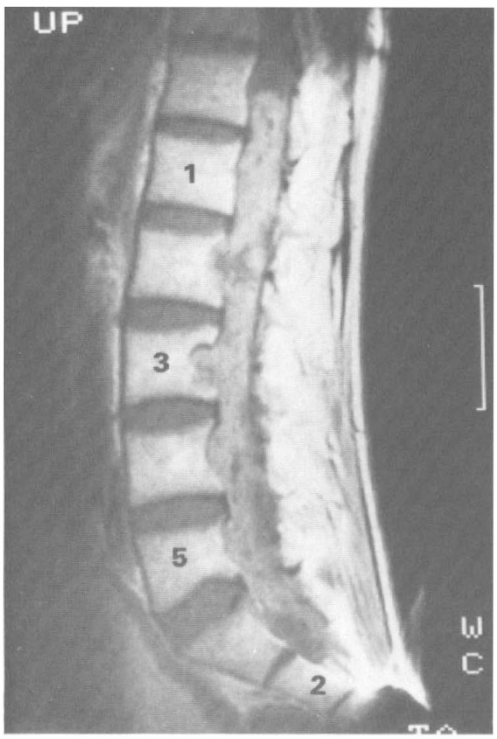

Figure 2 Case 6, myxopapillary ependymoma. T1-weighted MR images with Gd-DTPA enhancement showing a large tumour from T12 down to S2 with erosion of the vertebral bodies. This is the second largest in this series.

catheterisation technique for urination; one patient (case 5), who had been using crutches for 6 months prior to surgery, showed an improvement in walking with crutches, and returned to his work as a physician; the other patient (case 6), can walk slowly and is doing rehabilitation exercise. A patient with an astrocytoma grade 3 died due to intracranial dissemination 2 years after operation.

Discussion

The complete removal of an intramedullary astrocytoma is often difficult due to infiltration into the cord tissue.[2–5] However, excellent results have been reported following a radical excision of ependymomas, which are often non infiltrative.[3,6–9]

In our series of conus medullaris gliomas, a complete removal of tumour, both of astrocytomas and ependymomas, was rather difficult. This was due to the fact that the majority of the tumours were large (the largest was 17 vertebrae in the cephalocaudal extent (case 5), the mean was 5.2 vertebrae) producing longstanding mechanical compression, stretching and dislodgement of the nerve roots (Fig 3), with subsequent tight fibrous adhesions between the tumour and the nerve roots. Exophytic growth may be enhanced by these processes.

Interestingly, the exophytic growth differed between the two types of neoplasms: the ependymomas (cases 3, 4, 5, 6) showed predominantly vertical downward enlargement documented by MR images (Fig 1) and operative findings (Fig 5); the astrocytomas, although there were only two cases (cases 1, 2) showed predominantly not vertical but lateral enlargement (Fig 5).

Table II Pathology and neuroradiological summary of six cases of glioma of the conus medullaris

Case no.	Pathology	Location of tumour	Spinal cord swelling	Exophytic growth	Tumour cyst	Scalloping of vertebrae
1	Astrocytoma (grade III)	T10–L1	+	+	−	−
2	Astrocytoma (grade II)	T10–L1	+	+	−	−
3	Ependymoma	T12–L2	+	+	+	−
4	Ependymoma	T12–L3	+	+	−	+
5	Myxopapillary ependymoma	T8–S1	+	+	+	+
6	Myxopapillary ependymoma	T12–S2	+	+	+	+

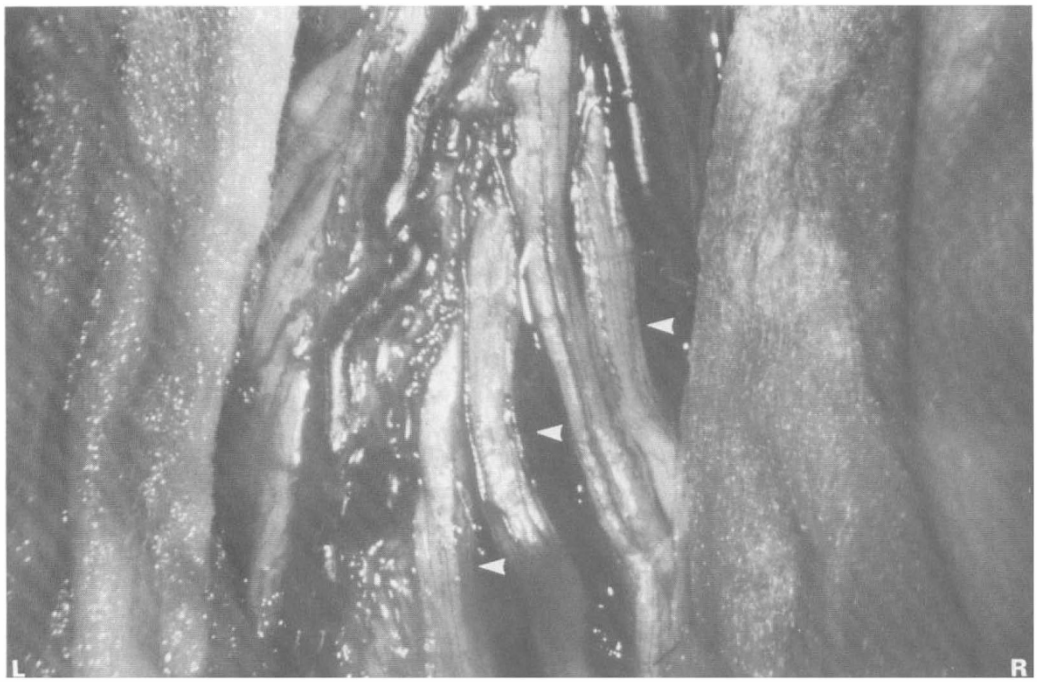

Figure 3 Case 2, grade 2 astrocytoma from T10 to L1. An intraoperative photograph showing a tumour with exophytic growth to the right lateral direction (arrow heads). Nerve roots were dislodged and adherent with the tumour.

The ependymoma arising from the ependymal or subependymal cells of the central canal,[10,11] may explain why an exophytic enlargement of the ependymoma develops on the midvertical direction below the level of the conus medullaris.

Prior to the advent of microsurgical techniques, radiation therapy was considered to be the major treatment of intramedullary glioma.[12,13]

Some authors reported an excellent survival rate after surgery alone,[3,6–9] while others stress the benefit of the irradiation for patients with residual tumours.[10,11,14] Guidetti reported 19 patients with astrocytomas who received radiation after incomplete surgical excision: only one patient died as a result of tumour growth.[8] Sonneland described 45 patients with myxopapillary

Figure 4 Case 3, ependymoma from T12 to L2. An intraoperative photograph showing a tumour with exophitic growth downward (arrow heads). The nerve roots were tightly adherent with the tumour.

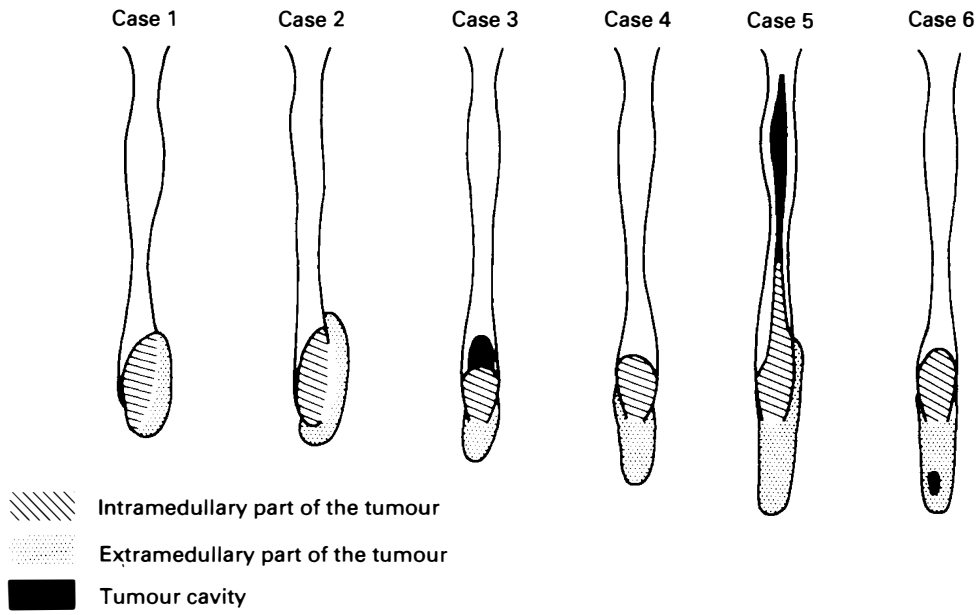

Figure 5 A schematic drawing to show a tumour within the cord (diagonal line area), outside the cord = exophytic part (dotted area) and cyst or cavity (black) of the individual cases. These drawings were reconstructed from operative findings and MR images.

Table III Treatment and outcome in six cases of glioma of the conus medullaris

Case no.	Removal	Radiation	Chemotherapy	Outcome
1	Partial	45 Gy	ACNU	Died 2 years later[a]
2	Subtotal	50 Gy	ACNU	No recurrence (for 7 years)
3	Subtotal	40 Gy	—	No recurrence (for 7 years)
4	Partial	30 Gy	—	No recurrence (for 6 years)
5	Partial	50 Gy	—	No recurrence (for 2 years)
6	Partial	50 Gy	—	No recurrence (for 2 years)

Subtotal more than 95% removal. Partial less than 95% removal. [a]Due to intracranial dissemination.

ependymomas who underwent total removal: seven (16%) recurred, and six had postoperative irradiation.[14] In our six cases, all had postoperative spinal irradiation and no recurrence was demonstrated on postoperative MR images with the shortest 2 years and longest 7 years follow up except for one case, who with a malignant grade 3 astrocytoma, died of intracranial dissemination 2 years after surgery. The remaining five tumours (one grade 2 astrocytoma, two ependymomas, and two myxopapillary ependymomas) were not removed completely, four were removed partially (less than 95%), and two were removed subtotally (more than 95%). Regardless of the extent of surgical excision, no recurrence was noted in this follow up. Although the role of postoperative irradiation is still controversial, it may be said that radiation therapy provided a beneficial effect on preventing tumour regrowth from the results obtained in this series. For radiotherapy, a localised field with a dose in the range of 40–50 Gy is recommended.[15–20] Kopelson recommended that irradiation be

given following either biopsy or subtotal removal and that a dose level of 45–50 Gy be given with the goal of long term tumour control.[16]

Garcia reported that an increasing dose of radiation correlated with an increase in tumour control and survival. Of patients who received 40 Gy or more, 75% showed apparent control of the tumour.[15] Risk of radiation-induced injury to the spinal cord or to the cauda equina is rare with a fractionated administration of 40 or 50 Gy.[18,19]

To our knowledge, no established method of chemotherapy for the spinal cord astrocytoma has been hitherto reported.[21,22] We applied ACNU (2 mg/kg i.v. as an initial dose, and 1 mg/kg i.v. as a maintenance dose every 8 weeks) in our clinic.[23,24] Further investigation is mandatory.

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