Intramedullary Ependymoma of the Spinal Cord

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We present 22 patients (10 men and 12 women), mean age 32 years (range, 11–65) who underwent the operative removal of intramedullary spinal cord ependymoma in 1980–1994. They experienced the symptoms for a mean of 16 months before initial diagnosis. At admission to clinic radiculopathy was present in 5, paraparesis in 8, paraplegia in 5 and tetraparesis in 4 patients. The location of tumor: cervical 6, thoracic 9, and cauda equina 7. Complete removal of tumor was achieved in 13, subtotal in 9 patients, among them there were 5 malignancies and one recurrence.

Recent neurological evaluation showed functional improvement in 8, recovered 5, no significant change in 7, died 2.

Key words: intramedullary spinal neoplasm, ependymoma, diagnosis, surgery, outcome

INTRODUCTION

Ependymoma accounts for 4–6% of primary central nervous system tumors, and about one-third arise within spinal canal as intramedullary neoplasm (1–6). Though ependymoma is a non-encapsulated glial tumor, histologicaly it is mostly benign, shows a little infiltrative potential, slow growth because patients present with a prolonged symptom duration prior to diagnosis and preservation of neurological function in patients with spinal cord compression. Long survival times after total tumor removal are possible (3, 4, 7–9). The aim of the work was to evaluate the clinical features and outcome after operative treatment of intramedullary spinal cord ependymoma.

MATERIALS AND METHODS

From 265 patients with spinal cord tumors, 22 patients with spinal cord ependymoma were operated on in the Neurosurgical Clinic of Kaunas Medical University Clinics in 1980–1994. The data were obtained from hospital records, out-patient inquiry and questionnaire. Tumor was diagnosed after myelography with In^{111}, RISA J^{131}, myelography with majodyl, omnipaque and CT + myelography. In 2 cases cranial CT was performed and intracranial ependymoma was revealed. At that time MRI was not available in our clinic.

Epiinfo and other statistic programmes were used in data analysis.

RESULTS

There were 10 men and 12 women, mean age 32 years (range, 11–65).

Clinical presentation consisted of neck, back or radicular pain, paresthesias, sensory loss, motor deficit and pelvic disorders. The mean duration of symptoms prior to operation was 16 (range, 6–48) months. These figures agree with the data of other authors (6, 10, 11, 13).

At admission 5 patients had only back and leg pain, mild motor disturbance as radiculopathy. Eight patients were walking with the aid of brace and cane and had a mild / middle weakness of lower extremities, unconstant pelvic disorders; these were patients with paraparesis. Five patients were paraplegic and 4 had tetraparesis.

Lumbar puncture revealed CSF protein/cell dissotiation in 16 cases with protein content 1190–35000 mmol/l, especially in cauda equina tumors (14). We have seen two deteriorations after lumbar puncture: in one patient it was leg weakness, in the second patient the weakness of upper extremities and spasticity increased. On myelography the lesions had a partial or complete block of contrast, often with a "meniscus" sign above and below the tumor (14, 15). In two cases cranial CT was performed; ependymoma of posterior fossa was malignant and metastases in thoracic spinal cord were found.

The pattern and progression of neurological deficit were related to tumor location. Six cases of

tumor location in cervical region manifested with tetraparesis and two patients had a mild leg deficit. Of 9 patients with thoracic ependymoma location, 5 patients had paraplegia and 4 paraparesis. Seven patients noted bowel and bladder problems. All patients with cauda equina ependymoma presented the pain as initial complaint: 5 patients have symptoms of radiculopathy and two patients a progressive weakness of one or both legs.

Indication for operation included the presence of lesion on myelography or computed tomography in patients with an objective and usually progressive neurological deficit. Ependymoma was removed totaly in 13 cases and subtotally in 9 cases. Histological examination revealed benign tumors in 17 cases and 5 were malign ependymoblastoma.

Benign ependymoma was classified into the following histological types: cellular (14 cases), epithelial (two cases) and myxopapillary (one case). In two cases of ependymoblastoma the patients had metastases after craniotomy in posterior fossa and radiation therapy after tumor removal. Two years later metastases in the thoracic region of spinal cord developed, and both patients with paraplegia were operated on and thoracic ependymoblastoma was subtotally removed. One patient had decubitus and thrombotic complications and in the second patient CSF leak and purulent meningitis developed. Two patients with total tumor removal and five patients after subtotal removal received local radiotherapy of 40 Gy.

Of 9 patients with radiculopathy and tetraparesis 8 (88.9%) recovered and improved, of 13 patients with paraplegia and paraparesis 5 (38.5%) improved and recovered. The risk of unimprovement in patients with paraplegia and paraparesis OR = 11.3 [1.28, 318.5] p = 0.024. Tumors in patients with paraplegia and paraparesis were of thoracic localisation. The outcome relation to tumor location is shown in Table 2.

Table 1. **Outcome related to preoperative neurologic condition**

Presentation	Outcome			
	recovered	improved	unimproved	died
Radiculopathy	5	–	–	–
Paraparesis	1	3	3	1
Paraplegia	–	1	3	1
Tetraparesis	–	3	1	–
Total	6	7	7	2

Table 2. **Outcome related to tumor localisation**

Tumor level	Outcome			
	recovered	improved	unimproved	died
Cervical	–	5	1	–
Thoracic	–	2	5	2
Cauda equina	5	1	1	–

Most (83.3%) patients with cervical localisation and with cauda equina tumors (85.7%) and only 2 (22.2%) patients with thoracic location improved and recovered.

Outcome related to initial therapy (Table 3) is as follows: after total removal of tumor recovered and improved 11, unimproved 2 patients.

After subtotal removal of tumor two patients improved, 6 unimproved and 2 patients died. One patient died 2 months after reoperation because of CSF leak and purulent meningitis, the second, with ependymoblastoma metastases from the posterior fossa to the thoracic spinal cord, died 19 days after laminectomy because of intoxication and exhaustion.

DISCUSSION

P. Bailey and H. Cushing (12) formally classified ependymoma as a distinct neoplastic entity. In 1954 Greenwood J. Jr. (13), using bipolar cautery and magnification loup, reported six complete removals of intramedullary ependymomas. The mortality rate was 22%, but there was no tumor recurrence. In our patients the mortality rate was 9.1% and recurrence 4.5%. Radiation therapy may be beneficial for incompletely removed cauda equina ependymomas. Radiotherapy after total excision of tumor is unclear. The majority of all intramedullary ependymomas are benign and potentially resectable (1, 2, 4). Even extremely large or extensive tumors must be completely removed, because reoperation for recurrent tumor is extremely difficult, especially in patient with previous radiotherapy. Incomplete removal is a factor for recurrence. Recurrencies (15–19%) are usually local, but can be delayed for several

Table 3. **Outcome related to initial therapy**

Therapy	Outcome			
	Recovered	Improved	Unimproved	Died
Total excision (n = 13), radiation (n = 2)	5	6	2	–
Subtotal excision (n = 9), radiation (n = 5)	–	2	5	2

years (14). We had 1 (4.5%) recurrence after subtotal removal of tumor. Other authors reported that extirpation surgery is the main therapy, and the role of radiation is less defined (15, 16). The incidence of malign spinal ependymoma varies within 1–10%. In our group of patients there were 5 (22.7%) cases of malignancy.

There is a clearcut difference in the agresiveness and prognosis among ependymoma in different areas: intracranial ependymoma has a worse prognosis than those occurring on spinal levels. A relatively benign prognosis has been reported in filum terminale ependymoma as compared to intracranial ependymoma.

CONCLUSIONS

1. The outcome of spinal cord ependymoma depends on tumor localisation: results are better in cervical and cauda equina tumors in comparison to tumors of thoracic localisation ($p = 0.014$).

2. The risk of unimprovement in patients with paraplegia and paraparesis and thoracic tumor localisation is OR = 11.3 [1.28, 318.5] $p = 0.024$.

3. Preoperative neurological condition of the patient and tumor localisation are the main prognostic factors.

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NUGAROS SMEGENŲ INTRAMEDULINĖ EPENDIMOMA

S a n t r a u k a

1980–1994 metais operuoti 265 ligoniai, sergantys nugaros smegenų navikais, iš jų 22 (8,3%) – intrameduline nugaros smegenų ependimoma (10 vyrų ir 12 moterų).

Darbo tikslas: išnagrinėti ependimomų klinikos ypatumus ir chirurginio gydymo rezultatų priklausomybę nuo priešoperacinės neurologinės ligonio būklės, naviko lokalizacijos ir operacijos radikalumo. Diagnozė nustatyta remiantis neurologinio tyrimo, rentgenokontrastinių (su In^{111}, RISA J^{131}, majodilu, omnipaku) ir kompiuterinės tomografijos, taip pat mielografijos tyrimų duomenimis. Duomenų analizė atlikta panaudojus Epiinfo ir kitas statistines programas.

Nustatyti priešoperaciniai motorikos defektai: radikulopatija (5), paraparezė (8), paraplegija (5), tetraparezė (4). Radikaliai pašalinus navik[1] pasveiko 5, 6-ių ligonių būklė būklė pagerėjo dviejų ne. Po subtotalinio naviko pašalinimo pagerėjo 2, nepagerėjo 5 ligonių būklė, 2 žmonės mirė. Penkiems ligoniams navikas buvo piktybinis, 2 – metastazės iš galvos smegenų į nugaros smegenis ir vienas recidyvas. Spindulinis gydymas po operacijos skirtas 7 ligoniams.

Išvados: 1. Nugaros smegenų ependimomos baigtis priklauso nuo naviko lokalizacijos. Lyginant su krūtininės lokalizacijos navikais ($p = 0,014$), kaklo ir arklio uodegos srities navikų chirurginio gydymo rezultatai geresni.

2. Rizika nepagerėti ligoniams su paraplegija ir parapareze bei torakaline nugaros smegenų naviko lokalizacija OR = 11,3 [1, 28, 318, 5] ($p = 0,024$).

3. Pagrindiniai prognoziniai veiksniai yra priešoperacinė neurologinė ligonio būklė ir naviko lokalizacija.

Raktažodžiai: nugaros smegenų intramedulinė ependimoma, chiruginis gydymas