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Epidural spinal sarcoma

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In 1980–1994, we have treated 237 patients with spinal cord tumors. The purpose of this work is to evaluate the clinical manifestation, treatment and outcome of patients with epidural spinal sarcoma. There were 22 patients with primary epidural spinal sarcoma (16 men and 6 women), mean age 46.2 years, mean duration of illness 2.1 years. The diagnosis was specified by plain X-ray, myelography with In^{111}, majodyl, omnipaque and CT. No MRJ was available at that time.

Localization of tumor: cervical – 2, thoracic – 15, lumbar – 5. Preoperative motor deficit: 12 patients were paraplegic, 3 with paraparesis, 1 with tetraparesis, 1 with tetraplegia and 5 with radiculopathy. 21 patients were operated on and one patient received only radiotherapy. Total removal of tumor was applied in 7 cases and subtotal in 14 cases. Six patients received postoperative radiotherapy.

The results of treatment depend on the character of malignancy, duration of illness and preoperative condition of patients. General results of sarcoma surgery are poor – only 27.27% of patients improved.

Key words: sarcoma, spinal cord, surgery, outcome

INTRODUCTION

Primary spinal sarcomas originate in nerve roots, spinal substance, blood vessels and other spinal structures. They are mostly found in the epidural space of the spinal cord. Spinal cord compression, secondary to epidural tumor, may be also accompanied by vertebral body and arch destruction.

In this article, we retrospectively analyzed our 15-year experience in treating primary epidural spinal sarcomas to better characterize their behavior and prognosis.

MATERIALS AND METHODS

In 1980–1994, of the treated 237 patients with spinal cord tumors 22 patients had spinal sarcoma.

The clinical information was obtained from case histories of patients. We evaluated their demographics, clinical symptoms, neurological status, diagnosis, surgery and outcome.

The diagnosis was specified by plain X-ray, myelography with In^{111}, majodyl, omnipaque and CT. No MRI was available at that time. Data analysis was performed using Epiinfo and Statistica programs.

RESULTS

Among the 22 patients with spinal sarcoma there were 16 men and 6 women. The median age of the patients was 46.2 years (range, 17–78). Mean duration of illness till hospitalization: in 17 patients from 1 month to 1 year, in 5 patients from 2 to 12 years.

The duration and severity of neurological symptoms and the length of affected spinal segment determine the chance of neurological recovery.

Table 1 presents the histological structure of the malignancies.

Table 1. **Histological structure of the malignancies**

Histological structure	Numbe of patients
Chondrosarcoma	1
Fibrosarcoma	3
Angiosarcoma	1
Lymphosarcoma	3
Neurofibrosarcoma	2
Sarcoma of unknown origin	12

The patients presented with a variety of clinical symptoms. The majority of patients (80%) presented with pain, local or radicular, half of them reported back pain. A significant number of patients (77%) arrived to the clinic with motor deficits ranging from lower extremity weakness to paraplegia. More than 50% had paraplegia, 13.6% with paraparesis, 9% tetraparesis and tetraplegia, and only 22.7% with radiculopathy. Among the patients prevailed men with paraplegia ($p = 0.005$). Numbness was reported in 12 patients; 17 patients had bowels and bladder disorders.

Location of tumor: cervical – 2, thoracic – 15, lumbar – 5. Thoracic location prevailed (p = 0.001). According to spinal cord axis, all spinal sarcomas were epidural.

Spinal cord compression was made by tumor in all cases, among them in 7 cases the additional cause of compression was vertebral body and arch destruction due to sarcoma invasion.

If spinal lesions compress the spinal cord and cause neurological symptoms such as paraparesis, decompression is required to save the patient from permanent neurological deficit. Decompression and stabilization may improve neurological symptoms and pain relief.

Twenty-one patients were operated on and one patient received only radiotherapy. Total resection of tumor was performed in 6 cases and subtotal removal in 15 cases. Radiotherapy was given to 6 of 21 surgically treated patients. Tumor irradiation dose was 45 Gy for 21–30 days, single fraction 2–5 Gy.

Table 2 presents the character of motor deficit, surgery and postoperative results.

Table 2. **The character of motor deficit, surgery and postoperative results**

Character of motor deficit	Number of patients	Surgery		Postoperative results			
		total removal	subtotal removal	improved	worsened	unimproved	died
Radiculopathy	5	2	2	2	2	1	–
Paraparesis	3	–	3	2	–	–	1
Paraplegia	12	3	9	2	–	6	4
Tetraparesis	1	–	1	–	–	–	1
Tetraplegia	1	1	–	–	–	1	–

There were 3 recurrencies after subtotal removal of sarcoma. Postoperative outcome was worse in men (p = 0.04): in two cases neurological status worsened and six patients died.

Postoperative results are related with the duration of illness: among the 17 patients with the duration of disease from 1 month to 1 year there were 5 deaths (31.25%). Among 6 patients with the duration of illness more than 1 year there was 1 death. These numbers confirm the great malignancy of sarcoma and especially rapid progression to death.

Postoperative results are related with the neurological deficit of patients. From 3 patients with paraparesis, 2 improved and 1 died. From 12 patients with paraplegia, 2 improved, 6 unimproved and 1 died. Five patients had radiculopathy – 2 improved, 1 unimproved, 2 worsened. One patient with tetraplegia after the total removal of tumor unimproved and one patient with tetraparesis after subtotal removal of tumor died.

Table 3 shows the causes of deaths.

The early postoperative mortality rate is high: 6 men (27.7%) died. Within 6–12 postoperative days 4 patients died from pulmonary and cardiovascular complications, and 1 month after operation 2 patients died from exhaustion. The general results of our 22 patients are poor – only 6 (27.27%) improved. The only way to improve the results is early diagnosis and surgery.

DISCUSSION

We report characteristics and treatment of 22 patients with spinal sarcoma treated in Neurosurgical Clinic of Kaunas Medical University Clinics during 15 years.

Primary central nervous system sarcomas most often affect young and middle-aged patients (1, 2). The median age (46.2 years) of our patients was within this category.

Gender distribution in our study showed a higher frequency in males. Spinal sarcoma occurs at a higher frequency in thoracic spine because of a greater number of thoracic segments relative to the cervical and lumbar region (3). Also in our group sarcoma of thoracic localization prevailed. It corresponds to the literature data, as most of the spinal lesions present as extradural masses (4). In our patients all tumors were extradural.

The patients presented with a variety of clinical manifestations. As mentioned above, the majority of our patients (80%) complained of pain (generalized, localized or radicular) with half of these reporting back pain (5).

As reported, epidural spinal cord compression results in motor deficit in 66% of patients and in 30% in paraplegia. In our group, 77% of patients had motor deficit and 54.55% paraplegia (6).

Bowels and bladder disturbances were similar.

It is emphasized that careful preoperative planning is necessary before surgical extirpation of tumor.

Surgical excision is the primary form of treatment.

Laminectomy resulted in a transient improvement of neurological symptoms in nearly all patients. The best chance to cure chondrasarcoma of the spine is *en*

Table 3. **Causes of deaths**

Causes of deaths	Number of patients
Embolia of pulmonary artery	2
Pneumonia	1
Cardiovascular insufficiency	1
Exhaustion	2

block surgical excision without spilling neoplastic cells into the wound (1, 2, 4, 5).

Irradiation as adjuvant treatment may be applied to radiosensitive tumors (2).

Irradiation is advocated to prevent local reccurrence after chondrosarcoma biopsy or inadequate surgical excision (4).

The extent of surgical resection has on impact on tumor recurrence (2). In our study, spinal sarcomas were mostly subtotally removed and we had 3 early recurriencies

In rare cases may occur metastasis of angiosarcoma to a lung, and pleural effusion may develop after total removal of tumors followed by radiation therapy (6).

There was reported a metastatic neurofibrosarcoma of spine. Metastases to the thoracic spinal cord caused paraparesis to a patient from neurofibrosarcoma of the femoral nerve. Malignant spinal metastases remain a rare complication of neurofibromatosis with a very poor prognosis (7).

Improved prognosis is achieved with local radiation treatment, especially when tumor mass resection is incomplete (8). Data in the literature showed that sclerosing epithelioid fibrosarcoma in deep skeletal muscles of adults has a tendency to local recurrence and late metastases (9). Partially resected tumors do appear associated with a higher mortality (11). In our group of patients, 27.27% died in an early postoperative period. Prognosis in high grade spinal sarcomas seems to be better than in glioblastoma multiforme (10).

The small number of patients in our series does not provide statistical data enough to evaluate the prognosis.

CONCLUSIONS

1. Epidural spinal sarcoma prevailed among men ($p = 0.08$) with thoracic localization ($p = 0.01$) and with paraplegia ($p = 0.05$).

2. The results of treatment depend on the character of malignancy, duration of illness and preoperative condition of patients.

3. General results of epidural spinal sarcoma surgery were poor – only 27.27% of patients improved.

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EPIDURINĖ STUBURO SARKOMA

Santrauka

Epidurinė sarkoma yra retas navikas. Aptarėme 22 ligonius (16 vyrų ir 6 moteris; vidutinis amžius 46,2 metai), gydytus dėl stuburo sarkomos. Darbo tikslas – įvertinti šių ligonių klinikinius požymius, gydymą ir baigtį. Pastebėta palyginti trumpa susirgimo trukmė iki hospitalizacijos: 17 ligonių nuo mėnesio iki metų, 5 – 2–12 metų. Daugiausia ligonių kreipėsi dėl paraplegijos, 13,6% – dėl paraparezės ir 9,1% – dėl tetraplegijos ir tetraparezės. Iš jų 21 operuotas, vienam skirtas tik spindulinis gydymas, 7 ligoniams atliktas totalinis ir 14 – subtotalinis naviko šalinimas. Šešiems ligoniams po subtotalinio naviko pašalinimo skirtas spindulinis gydymas. Stebėti trys naviko recidyvai, 6 ligoniai (27,27%) mirė ankstyvuoju pooperaciniu periodu.

Išvados: 1. Dažniausiai nustatyta krūtinės srities epidurinė sarkoma jau esant paraplegijai. 2. Gydymo rezultatai priklausė nuo naviko piktybiškmo, ligos trukmės iki hospitalizacijos ir ikioperacinės būklės. 3. Po operacijos pagerėjo tik 6 (27,27%) ligonių būklė.

Raktažodžiai: sarkoma, nugaros smegenys, gydymas, baigtis