LATE RECURRENCE OF SPINAL NEURINOMA AFTER ITS SINGLE-STAGE TOTAL REMOVAL

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Dumb-bell schwannomas (neurinomas) account for 15 % of the total number of spinal neurinomas. Their surgical removal in the cervical spine is a technical challenge due to intricate anatomical relationships. The paper presents a clinical case of recurrence of the dumb-bell neurinoma at the cervicothoracic level 20 years after its total single-stage removal. The tumor recurrence was confirmed by the data of neuroimaging monitoring. Repeated removal was performed. The paper describes in details the course of the disease, clinical and radiological manifestations, and results of neurophysiological monitoring.

Key Words: dumb-bell tumor, schwannoma, recurrence of the neurinoma of the spinal root, surgical intervention, magnetic resonance imaging.

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Primary intra- and extramedullary spinal cord tumors account for up to 10 % of all primary tumors of the central nervous system [6], and their ratio to spinal cord and cerebral tumors is estimated as 1:4–1:6 [1].

Neoplasms possessing intracanal and paravertebral components with an isthmus in the intervertebral foramen have been referred to in the literature as "hourglass" or "dumbbell" tumors. This tumor growth is typical of hemangioblastomas, meningiomas, neuroblastomas, gangliomas, and neurinomas [5]. During their surgical treatment, not only intracanal but also paravertebral and intraforaminal tumor components are subject to removal; their total resection should be performed in one stage, if possible. Staged resection considerably increases the period of patient disability and the risk of surgical complications in subsequent interventions [3].

The incidence rate of spinal cord neurinomas is 0.3–0.4 cases per 100,000 people per year [15], but tumors with intracanal/paravertebral extension account for only 15 % of the total number [10, 12–14]. The cervical spine is the most common localization of dumbbell tumors [11–14]. However, even total resection of these neurinomas does not exclude the recurrence rate of 5 %, according to multicenter international databases [7].

Rare cases of extremely late recurrence of dumbbell neurinomas and almost complete lack of published data on the dynamics of sensory and motor defects estimated by electroneurophysiological techniques prompted the authors to report the case.

A male patient I. considers himself ill from the age of 31 years. In 1995, he marked weakness in his left upper limb, which gradually aggravated for the next two years. In 1997, MRI of the cervical spine revealed an extramedullary dumbbell tumor at the C5–C6 level on the left, which compressed the spinal cord (type IVb according to the classification by Sridhar et al. [16]). The intracanal component was 1.8 × 1.5 × 2.0 cm in size, and the extravertebral component was 3.0 × 3.5 × 4.0 cm in size (Fig. 1).

The patient underwent elective surgery that included single-stage total intra-extracanal resection of a C6 spinal root lesion using a neodymium laser through a hemilaminectomy approach on the left. According to a morphological study, the tumor was identified as Grade 1 neurinoma. After surgical treatment, an improvement in the form of pain regression was observed. In this case, there was persistent hypesthesia in the innervation area of the C6 spinal nerve root on the left, which gradually regressed in the subsequent years, so the patient could work in the specialty and lead normal life. In 2014 (17 years after surgery), a follow-up MRT examination revealed no signs of recurrent tumor (Fig. 2).

Twenty years after surgery, being in good health, the patient experienced pain in the lower cervical spine and left limbs as well as weakness in the left hand. In June 2017, at the age of 53 years, the patient underwent MRI that revealed a recurrent extramedullary neoplasm at the C5–T1 level, 5.5 × 2.2 × 1.6 cm in size, which extended into the postoperative defect of the left C6 lamina and into the left C6–C7 intervertebral foramen with compression of the spinal cord at the C5–T1 level (Fig. 3).

Neurological examination. The cranial nerves were normal. Strength in the upper and lower limbs: 5 points on the right and 4 points on the left; hand-grip strength decreased to 2 points. Tendon reflexes from the upper and lower limbs were moderately brisk, S > D. There was hypesthesia in the left upper limb below the elbow joint, in the innervation area of the C6–C8 nerve roots.

Before surgery, electromyography (EMG) of the upper limbs detected a decreased excitation function of the radi-

al and ulnar nerves on the left, especially in the distal parts. The conduction function of the left ulnar nerve was below the normal value and more proximal; the velocity at the forearm level was 49.0 m/s. On the left, M-responses from the *biceps* to stimulation of the musculocutaneous nerve were lower; upon stimulation of the median nerve, the *thenar* tension activity in the left hand was significantly lower, with M-responses being higher.

According to stimulation EMG of the lower limbs, the function of both sciatic nerve branches on the left was reduced in the distal parts: the H-reflex on the right was high and brisk, the delay was 29.9 ms; the reflex was much lower on the left, the delay was 34.1 ms, which corresponded to radiculopathy of S1 on the left.

Therefore, on the left, the muscle tension activity in the upper and lower limbs was reduced; there were signs of myeloradiculopathy, with lateral differences in the electrical tension activity being more pronounced than the differences in responses to stimulation (strength in points), which indicated the prevalence of myelopathy manifestations.

Local status. The postoperative skin scar without signs of inflammation was visualized on the posterior neck surface in the projection of the C3–T2 spinous processes. Paravertebral palpation at the C5–C6 level on the left revealed a sharp tenderness with irradiation to the left upper limb.

The clinical neurological examination and MRI findings indicated the presence of an extramedullary tumor at the C5–T1 level on the left, type III according to the classification by Sridhar et al. [16], which suggested a recurrent neurinoma. Elective surgery was decided upon.

Surgery protocol. With the patient in the sitting position and his head fixed in a Mayfield head clamp, the C5 and C7–T1 laminae on the left were skeletonized through a linear incision along the old scar, with a 2.0 cm caudal extension. A left C7–T1 hemilaminectomy with partial resection of the lower edge of the left C5 lamina was performed. The dura mater with areas of old defect bulged out in the trepanation window through

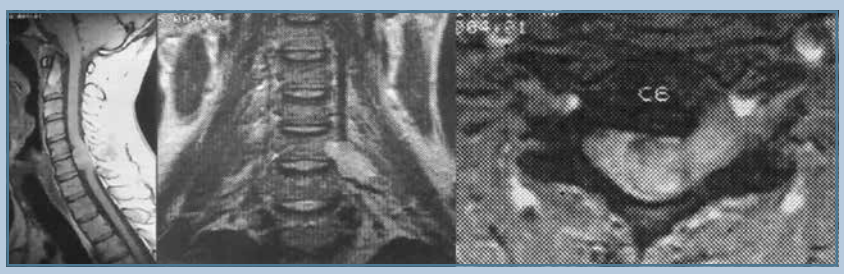

Fig. 1
MRI scans of the cervical spine and spinal cord of the patient I. before surgery in 1997

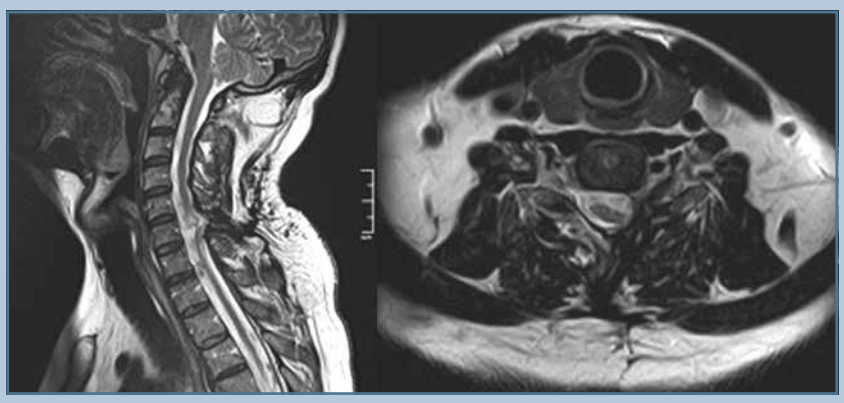

Fig. 2
MRI scans of the cervical spine and spinal cord of the patient I. 17 years after surgery

which the thickened bluish-cherry arachnoid membrane prolapsed. By using microsurgical instrument and microscopic magnification of ×12.0, the dura mater was opened in the bulging area, and the edges were pulled apart using holders. The arachnoid membrane was tightly adherent to the spinal cord and had large-spotted dark-brown patches (area of old hemorrhages). The spinal cord bulged out in the bone defect area. In the ventrolateral part of the dural sac on the left, there was a grayish-cherry mass of soft consistency, with a well-defined capsule, which displaced and deformed the spinal cord. The tumor capsule was coagulated and dissected using a microsurgical technique. Internal decompression of the capsule was performed using a vacuum aspirator, which was accompanied by mild bleeding. In the tumor cavity, cysts filled with brownish liquid were found and evacuated. The anterior C6 nerve root was of a grayish-cherry color and was invaded by the tumor. The C6 nerve root was excised in an intact portion. The remaining neoplasm located subdurally and blindly ended in the C6–C7 intervertebral foramen was totally resected. The CSF spaces were decompressed, which enabled free CSF circulation in the dural sac cavity. In the absence of continued bleeding signs, the dura mater was continuously sutured (Prolene), and the suture was covered with Tachocomb plates. There were no signs of liquorrhea. The wound was sutured in layers.

In the early postoperative period, there was moderate worsening of the neurological deficit in the form of aggravating weakness in the left limbs (up to

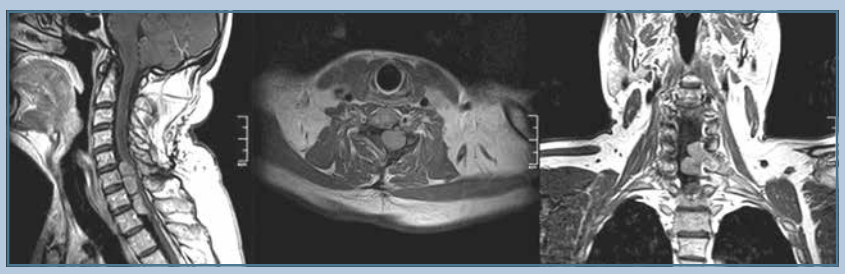

Fig. 3
Contrast-enhanced MRI scans of the cervicothoracic spine of the patient I. that revealed a recurrent neurinoma at the C5–T1 level

3 points), more pronounced in the left hand (up to 1 point).

On the 5th postoperative day, the patient underwent control contrast-enhanced MRI (Fig. 4) that confirmed total tumor resection.

In the early postoperative period, neurological symptoms in the patient who was on exercise and massage therapy partially regressed: strength in the left hand increased proximally to 4 points; peripheral hand paresis remained at the same level. There was no change in the lower limb motor function, but the radicular pain disappeared. The wound healed by primary intention. The patient was discharged in a satisfactory condition on the 15th postoperative day.

A histological examination of the material confirmed the diagnosis of neurinoma: the tumor tissue consisted of elongated cells predominantly with fusiform and rounded nuclei (Fig. 5a); the cells were tightly arranged and formed periodic patterns; there were tumor areas with a solid-reticular pattern. In the tumor stroma, there were a significant number of sinusoidal vessels (in places, with thickened walls), focal sclerosis, and moderate diffuse-focal lymphoid infiltration (Fig. 5b).

An immunohistochemical analysis showed positive staining of tumor cells with S100 antibodies, STAT6 antibodies (single cells), and CD34 antibodies (vessels). The proliferation marker Ki 67 staining index was 8–10 %. According to the morphological structure, the tumor was a cellular schwannoma (Grade I) with signs of growth activity.

Seven months after surgery, the patient's condition remained satisfactory. There were persistent complaints of moderate weakness in the left limbs, more pronounced in the left hand: strength in the upper and lower limbs was 5 points on the right and 4 points on the left, with hand-grip strength decreasing to 2 points. There was hypesthesia of the C6–C8 spinal nerve roots on the left in the innervation area.

A neurophysiological examination using electromyography and somatosensory evoked potentials revealed a severe decrease in function of the median and ulnar nerves on the left, in their proximal parts; distal radial nerve function was significantly reduced, with M-responses from the brachioradial muscle (more proximally) and the activity of its tension being at the preoperative level. Excitation functions of the musculocutaneous and axillary nerves differed little from those on the right; M-responses and activity of *m. biceps* and deltoid muscle tension were slightly higher compared to the preoperative levels.

On the right, M-responses from the hand muscles had a low amplitude, and their delay to stimulation of the distal median and ulnar nerve trunks was normal; nerve conduction velocities in the cubital fossa-wrist region were normal (median nerve, 56.8 m/s; ulnar nerve, 57.9 m/s) and almost did not differ from those detected before tumor resection.

The tension activity of all lower limb muscles was decreased, with myelinopathic conduction functions of the tibial and peroneal nerves being bilaterally decreased by 7 to 30 % compared to the preoperative levels. The activity on the left was lower, but these lateral differences (signs of myelopathy above the lumbosacral level) were also present before surgery.

Sensory conduction to the cortex was most reduced and delayed from the C7–T1 level on the left. Parameters of conduction from the right upper limb also demonstrated signs of moderate myelopathy at the cervical level (Fig. 6). Sensory conduction from the lower limbs was reduced and mostly delayed from the left lower limb; sensory conduction from the right lower limb was delayed and desynchronized.

Of particular importance was preservation of somatosensory evoked potentials to stimulation of the distal median and ulnar nerves, which contrasted with severe functional failure of the motor portions of these nerves in the distal parts.

Therefore, the neurophysiological data were consistent with the neurological symptoms and surgical findings: neoplastic spinal cord compression and surgical intervention mostly affected the anterior portions and anterior nerve roots at the C6–T1 level.

Late recurrences may be associated with various spinal cord tumors: Babiyan [2] reported a recurrent meningioma in the thoracic spine 19 years after tumor resection. Feiring and Barron [8] presented two cases of tumor recurrence 27 and 28 years after surgery. The authors believed that recurrence was associated with incomplete tumor resection.

The largest number of recurrences is observed in neurofibromatosis. An analysis of the surgical outcomes in 87 patients with recurrent neurinomas in neurofibromatosis I (NF1) and II (NF2) [12] revealed their association with a genetic defect in chromosome 17 g in NF1 patients. The recurrence rate in these patients was 10.7 % after 5 years and 28.2 % after 10–15 years. For NF2 (central type of neurofibromatosis), the

recurrence rate at 5 years was 39.2% and close to 100.0 % at 9 years.

We could find a very small number of reports devoted to recurrent solitary spinal neurinomas; they most often recur in the first two years after surgery [7]; however, there were reports of their more delayed recurrences – 12 and 18 years after removal [4, 9].

Conclusion

The presented clinical case indicates that the microsurgical technique used for gross resection of intra-extravertebral dumbbell tumors can not provide relapse-free survival; benign neurinoma of the spinal cord roots may recur even 20 years after its total resection. We are inclined to think that insufficient radicalness of surgery is the cause of tumor recurrence.

The study had no sponsorship. The authors declare no conflict of interest.

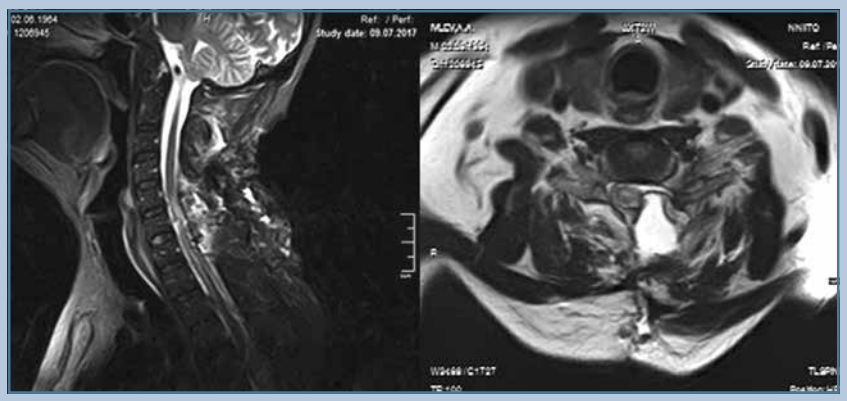

Fig. 4
Contrast-enhanced MRI scans of the patient I. on the 5th postoperative day: a small CSF cyst in the surgical area is seen; no tumor tissue is detected

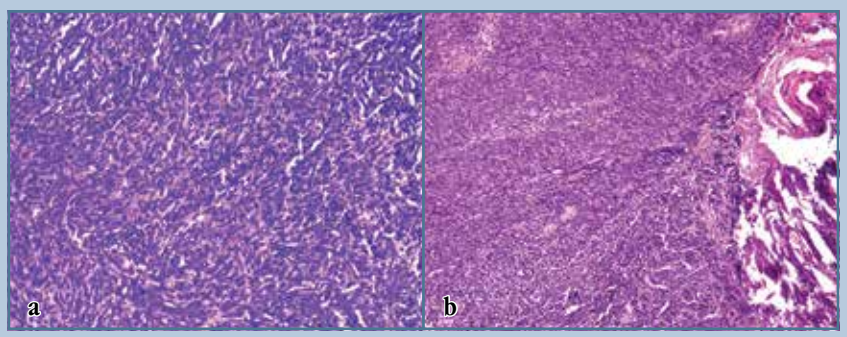

Fig. 5
Histology of the patient I. specimen, staining with hematoxylin and eosin, magnification 100:
a – elongated cells; fusiform and round nuclei; **b** – lymphoid infiltration

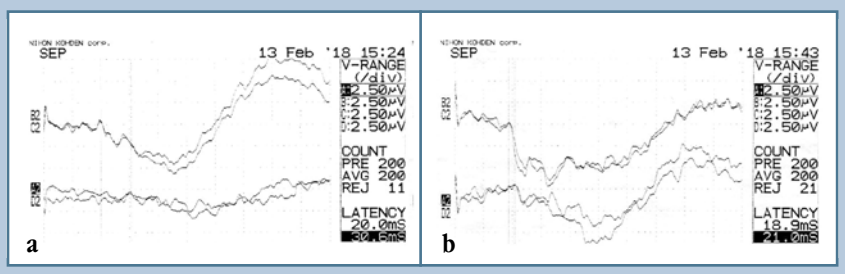

Fig. 6
Somatosensory evoked potentials to stimulation of the median (upper curves) and ulnar (lower curves) nerves on the left **(a)** and right **(b)** upper limbs

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