C_1 and C_2 nerve sheath tumors: Analysis of 32 cases

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Abstract

Background: C_1 and C_2 nerve sheath tumors are to be considered as a separate clinical entity because of their unique clinical presentation, relation to the adjacent structures and surgical approaches when compared to their counterparts in other regions. **Aim:** To present the clinical characteristics, radiological findings, operative approaches, and outcomes. **Setting and Design:** Of the 32 patients, the study was retrospective in 22 patients and in 10 it was prospective. **Materials and Methods:** Patients' case records were analyzed. Diagnosis was established by magnetic resonance imaging of craniovertebral junction. Patients were evaluated by Yasargil grading and operative procedure was done accordingly. **Results:** In this series the tumors were multiple and had extradural extension and hourglass expansion. Surgical results were excellent in most patients when procedure was done early in the course of the disease. Best results in terms of complete removal and good neurological outcome were achieved with posterior or posterolaterlly located lesion. Of the 20 patients who had surgery by posterior approach, 19 (95%) patients had improvement postoperatively, while one (5%) patient remained unchanged. While of the 8 patients who had surgery by posterolateral approach, 6 (75%) had improvement postoperatively, one had no change and the other had neurological deterioration. **Conclusion:** C_1 and C_2 nerve sheath tumors are to be considered as a separate clinical entity. Operative results and recovery of patients are excellent when tumor is located posterior or posterolaterally and surgical procedure is done early in the disease course.

Key words: *Foramen magnum, hourglass tumor, nerve sheath tumors*

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Introduction

Nerve sheath tumors of C_1 and C_2 nerve roots are rare; they represent approximately 5% of all spinal nerve sheath tumors and 18% of cervical nerve sheath tumors.[1] Nerve sheath tumors in these locations are generally considered under spinal nerve sheath tumors[1-3] or foramen magnum tumors.[1,4-6] C_1 and C_2 nerve sheath tumors have certain specific characteristics such as multiplicity, hourglass expansion, and the relation to the vertebral artery and the specific surgical problems associated with it.[1,6-10] The unique osseous anatomy and biomechanical characters of this region needs special consideration.[11] This communication presents a review of the demographic profile, clinical and radiological findings, operative procedures and outcome in 32 patients with C_1 and C_2 nerve sheath tumors.

Materials and Methods

This study was conducted at the University Hospital from January 1988 to May 2005. During this period 242 cases of spinal nerve sheath tumors were operated. Cervical nerve sheath tumors accounted for 106 cases, of which 32 were in the C_1 and C_2 region. Of the 32 patients, in 22 patients the study was retrospective (January 1988 - December 2001) and in 10 patients it was prospective (January 2002 - May 2005). Follow-up period ranged between 6 months to 15 years.

The patient's clinical records were reviewed for demographic profile, clinical and radiological findings, operative procedures and outcome during the postoperative period and follow-up. In all the patients, diagnosis was established by magnetic resonance imaging (MRI) of craniovertebral junction. Computed

tomography (CT) of craniovertebral junction was done in selected patients in whom MRI showed large dumbbell tumor, to study osseous anatomy. Vertebral angiography was done in 15 patients with large dumbbell tumors to decide the side of extreme lateral approach in anterior and anterolateral tumors based on dominance of the vertebral artery. Patients were clinically evaluated by using Yasargil grading.[1,12] Grade 0 - no symptom, Grade 1- minor symptom, Grade 2- minor symptoms or signs, Grade 3 - marked symptoms and signs, and Grade 4- bedridden. Death in the postoperative period was considered as Grade 5. During the operation microscope and ultrasonic surgical aspirator were used. Follow-up records were analyzed to assess the functional recovery.

Results

C_1 and C_2 nerve sheath tumors accounted for 13.2% of all spinal nerve sheath tumors and 30.2% of all cervical nerve sheath tumor. Of the 32 patients, 20 were male, age ranged from 11 to 60 years with a mean age of 35.5 years. The tumor was common in middle aged males. The symptoms and signs at the presentation are given in Table 1. Preoperatively twenty (62%) patients were in Yasergil Grade 3, five (15.62%) patients in Grade 2, four (12.5%) patients in Grade 4 and three (9.37%) patients in Grade 1 [Table 2].

T1-weighted MRI of craniovertebral junction showed iso to hypo signal intensity mass lesions while it was hyper intense on T2WI. Post-gadolinium diethylene triamine pentaacetic acid MRI showed homogenous brilliantly enhancing tumors. Based on the location of the mass lesion, the C_1 and C_2 nerve sheath tumors were classified into four types: anterior; anterolateral; posterolateral; and posterior. Six (18.75%) tumors were anterior [Figure 1, Figure 2], 15 (46.87%) were anterolateral, [Figure 3], 7 (21.87%) were posterolateral, and 4 (12.5%) were posterior [Table 3] in location. Of the 32 patients, 28 (87.5%) patients had single tumor (18 (64.28%) at C_2 and 10 (35.71%) at C_1) and four (12.5%) patients had multiple tumors. In 2 (6.25%) patients the tumor was purely intradural, in 10 (31.25%) patients it was extradural, and in 20 (62.5%) patients it was both intradural and extradural with hourglass expansion [Figure 4]. Vertebral angiography was performed in 15 patients with large dumbbell tumor. Feeding branches were arising from the vertebral artery at C_3 in 5 patients, at C_2 in 8 patients, and at C_1 in 2 patients. Vertebral artery was in contact with the tumor in 7 patients and shifted laterally by the tumor in 8 patients, and in one patient there was stenosis of the vertebral artery [Figure 5].

Surgical techniques included posterior, extreme lateral, and posterolateral approaches. Posterior approach

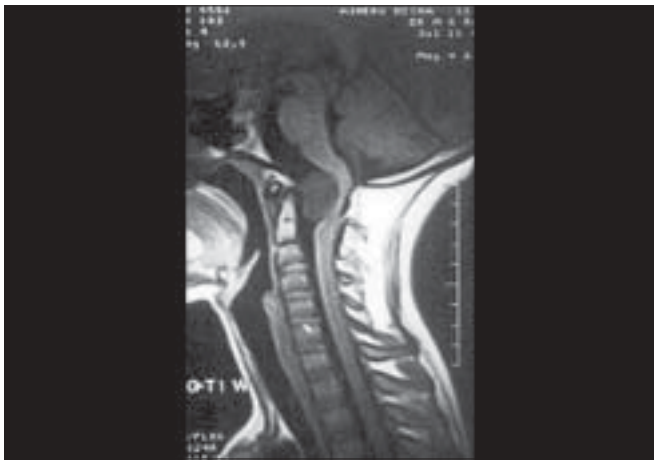

Figure 1: T1 weighted sagittal section of MRI of cervicomedullary junction showing iso to hypointense anteriorly placed tumor compressing the cord posteriorly

Table 1: Clinical features

	No. of patients (n = 32)	Percentage
Symptoms	30	93.75
Suboccipital headache		
Limb weakness	25	78.12
Numbness	20	62.50
Sphincter disturbances	9	28.12
Neck stiffness	4	12.50
Signs		
Spasticity	30	93.75
Suboccipital headache	20	62.50
Motor deficit	18	56.25
Spino-thalamic tract involvement	15	46.87
Posterior column involvement	12	37.50
Horner's syndrome	3	9.37
Gait disturbance	6	18.75
Facial hypoesthesia	6	18.75
Dyspnea at rest	5	15.62
IX-X nerve palsy	3	9.37
Lhermitte's sign	2	6.25

Table 2: Clinical evaluation of cases of C_1 and C_2 nerve sheath tumors based on Yasargil grading

Grading	Preoperative		Postoperative	
	n = 32	%	n = 32	%
0	0	0	1	3.12
1	3	9.37	4	12.5
2	5	15.62	23	71.8
3	20	62.5	2	6.25
4	4	12.5	1	3.12
5	0	0	1	3.12

Table 3: Surgical approaches depending on location of tumors in relation to dura

Surgical approaches	Tumor location			Total cases
	Intradural	Extradural	Dumbbell	
Posterior	1	6	13	20
Posterolateral	1	2	5	8
Extreme lateral	0	2	2	4

was a standard midline posterior incision with bilateral laminectomy of C_1 and C_2 for the C_2 tumors and resection of the posterior arch of the atlas with or without suboccipital craniectomy for C^1 tumors. Posterior approach was used for posterior, posterolateral and anterolateral lesions with contralateral shift of the cord. It was used in 20 (62.5%) patients. The majority of lesions were excised completely [Tables 3 and 4].Extreme lateral approach was used for anterior or anterolaterally located lesions. Control of vertebral artery extra-periosteally was done to avoid troublesome bleeding from the venous plexus that lies inside the periosteal sheath surrounding the artery. Ipsilateral occipital condyle was excised. This approach provided a lateral exposure of the lesion, permitting safe dissection without retraction of the cord. This approach was done in 4 (12.50 %) patients [Table 4]. Posterolateral approach was used for posterolaterally located lesions which was a standard posterior approach enlarged laterally up to the transverse foramen of C_1 to include exposure of

Table 4: Surgical approaches depending on tumor location in relation to cord

Surgical approaches	Tumor location								Total	
	Posterior		Posterolateral		Anterior		Anterolateral		No.	%
	No.	%	No.	%	No.	%	No.	%		
Posterior	4	12.5	6	18.75	-	-	10	31.25	20	62.5
Posterolateral	-	-	1	3.12	3	9.37	4	12.5	8	25.0
Extreme Lateral	-	-	-	-	3	9.37	1	3.12	4	12.50
Total	4	12.5	7	21.87	6	18.75	15	46.87	32	100.0

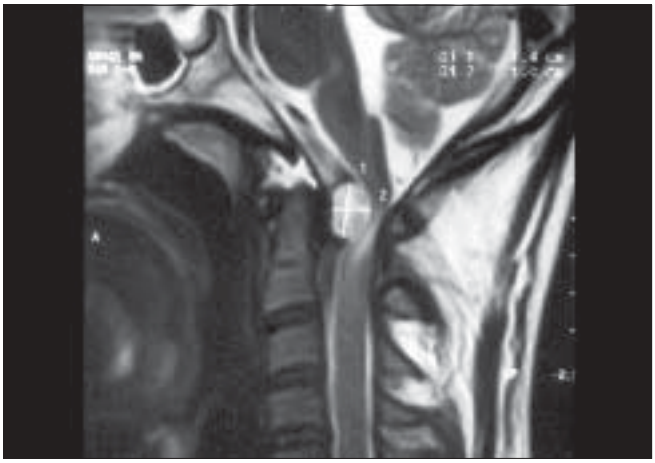

Figure 2: T2 weighted sagittal section of MRI showing anteriorly placed hyperintense tumor compressing the cord posteriorly with signal changes in the cord of same patient as of Figure 1

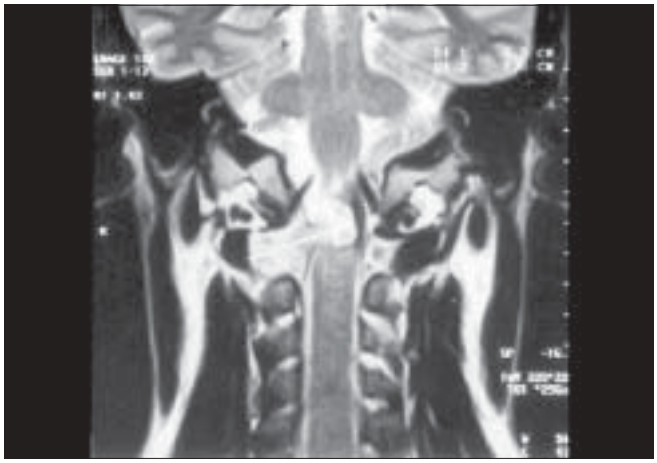

Figure 3: T2 weighted coronal section of MRI craniovertebral junction showing posterolateral dumbbell-shaped hyperintense tumor compressing the cord to opposite side

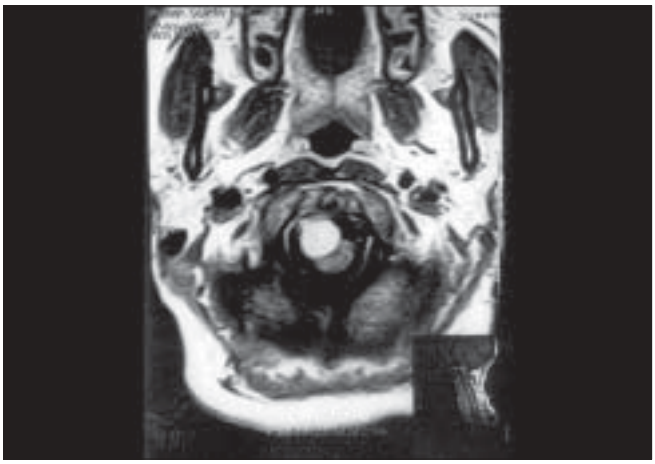

Figure 4: Post-gadolinium DTPA axial section of MRI cervicomedullary junction showing homogenous brilliantly enhancing anterolateral tumor compressing the cord posterolaterally

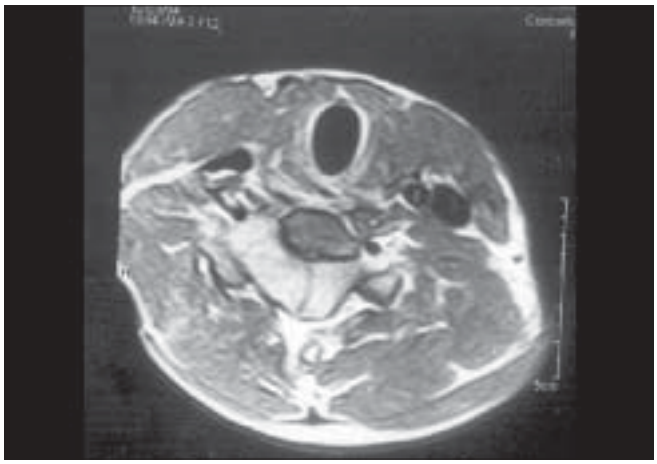

Figure 5: T1 weighted axial view of MRI at C_2 level showing right-side lateral dumbbell tumor displacing the cord to the left side and vertebral artery anterolaterally

the vertebral artery above C_1 and between C_1 and C_2. It was used in 8 (25%) patients [Tables 3 and 4]. The C_1 nerve root was difficult to identify in its extradural and extraspinal portion and with the naked eye because it was a very thin strand emerging between the vertebral artery and the groove of the atlas, but with the help of a microscope it was easily identified. The C_2 nerve root was easily exposed as it separates into anterior and posterior divisions while curving around the vertebral artery and it had a long course between the dural sac and the vertebral artery. Total resection was done in 26 (81.25%) patients, subtotal in four (12.5%) patients and partial in two (6.25%). Subtotal resection was defined as a removal leaving a remnant less than 5 mm in length, generally close to the vertebral artery. Partial resection was when a portion of tumor more than 5 mm in length was left. Partial resection was done when massive bleeding occurred in one of the anteriorly located tumors and in other dumbbell tumor. Bleeding was controlled by packing with gelfoam and surgical, and surgery was abandoned.

Postoperative neurological improvement was seen in 26 (81.25%) patients, neurological status was unchanged in 3 (9%) patients, deteriorated in 2 (6.25%) patients, and there was 1 (3.1%) death [Table 5]. Of the 20 (62.5%) patients in Yasargil Grade 3, only 2 (10%) deteriorated by one grade following surgery. However both improved to preoperative status at the time of discharge. The remaining 18 (90%) patients showed improvement in the neurological status by one or two grades at the time of discharges or during early follow-up period. Nine (28%) patients with bladder symptoms had marked improvement in their bladder symptoms within four weeks postoperatively, this includes a patient with bladder symptoms for more than four weeks [Table 2]. During follow-up 2 (6.2%) patients with residual tumor presented with recurrence of symptoms. Both were re-operated, only subtotal resection could be done. Massive adhesions and distorted anatomy limited total resection.

Of the 20 patients who had surgery by posterior approach, 19 (95%) patients had improvement postoperatively, while one (5%) patient remained unchanged. While only 6 (75%) of the 8 patients who had surgery by posterolateral approach had improvement. Of the remaining two patients, one had no change in the neurological deficit and the other had neurological deterioration. The neurological deterioration was attributed to the cord edema, which improved with steroid therapy. The death in one patient was related to postoperative pneumonia. None of our patient had spinal instability problems.

Discussion

C_1 and C_2 nerve sheath tumors are relatively rare tumors and exhibit certain unique features: multiplicity, hourglass expansion and their relationship to the vertebral artery.[1,6-9] Nerve sheath tumors of C_1 and C_2 nerve roots represent approximately 5-12% of all spinal nerve sheath tumors and 18-30% of all cervical nerve sheath tumors.[1,10] Nerve sheath tumors arising from C_2 nerve root is much more common than C_1 nerve root Bazooka *et al.*,[6] reported 18 tumors arising from C_2 nerve root and only one tumor from C_1 nerve root. Geodetic and Pallone [5] reported six tumors arising from C_2 nerve root and three from C_1 nerve root. George *et al.*,[1] described 15 cases from C_2 nerve root and six from C_1 nerve root. In our series of 32 tumors, 20 (62.5%) were from C_2 nerve root and 12 (37.5%) were from C_1 nerve root. Dumbbell-shaped tumor is the most common type of tumor arising from C_1 and C_2 nerve roots and accounts for more than 80% of the tumors.[1,10] Tumors arising form C_1 and C_2 are common in the middle age, they can also occur in the younger age.[1] In our series three patients were below 20 years of age at the time of diagnosis. C_1 and C_2 nerve root tumors present with features of high cervical cord compression. Most common symptoms reported in the literature are spasticity, followed by sub-occipital headache and motor deficit. Bladder involvement is a feature in 19-28.6% of cases and Lhermitte's sign is observed in 5% of cases.[1,10] The tumors are usually large in size at the time of diagnosis. It is due to the spacious spinal canal at this level.[10] Mean sagittal diameter at the level of the atlas and axis is 23 and 20 mm respectively while at sub-axial spine, the average diameter is only 15 mm. The cervical bulge of the spinal cord also begins below the axis.[10,11] At the occipitocervical and atlantoaxial levels, the facial pillars lie anterior to the nerve roots exiting through the intervertebral foramina; this also permits an exuberant growth of lesion outward since there is no posterior bony obstruction to its growth

Table 5: Outcome of surgical procedure

Surgical procedure	Post-operatively improved		Remained unchanged		Deteriorated		Dead		Total	
	No.	%	No.	%	No.	%	No.	%	No.	%
Posterior	19	95	1	5	-	-	-	-	20	62.5
Posterolateral	6	75	1	12.5	1	12.5	-	-	8	25.0
Extreme lateral	1	25	1	25	1	25	1	25	4	12.5

at C_1 and C_2 level.[10,11] There is often a delay in the development of clinical symptoms until these tumors attain a fairly large size.[8,10,11]

Various operative procedures include posterior approach (standard bilateral laminectomy with or without suboccipital craniectomy),[1,10,13,14] posterolateral approach,[1,7,10] (laminectomy and medial ipsilateral partial fasectomy), and extreme lateral approach[10,15-18] (laminectomy and ipsilateral occipital condyle excision). Posterior approach is simple and the standard procedure for posteriorly situated tumors, and involves lesser risk of instability. But posterior approach has inadequate access to anterior/anterolateral tumors and this approach has no access to extraforaminal tumors.[1,10,13,14] Posterolateral approaches are used in most cases in which an extradural extension and relation with the vertebral artery had been demonstrated on preoperative imaging.[1] It enlarges the usual opening laterally, and provides the best results in terms of either rate of complete resection or clinical improvement.[1] It permits access to any tumor extension, especially the extradural component close to the vertebral artery.[1,7,10] Extreme lateral approach described by Sen *et al.*,[17] is suitable for C_1 and C_2 tumors situated ventral or ventrolateral to the cord. Lateral approaches give a lateral access to the intradural portion of tumors, avoiding any manipulation on the spinal cord and medulla and any traction on the tumor when it extends anteriorly.[1,10,15-18]

The C_1 nerve root section does not produce any significant sensory deficit. Division of the proximal part of the C_2 nerve root generally produces only a very mild sensory deficit because of important distal anastomosis.[1] But distal branch division leads to hypoesthesia or anesthesia in corresponding territory.[1] Sectioning of the nerve root at the time of the removal of the tumor leads to a neurological deficit in a minority of cases. Postoperatively, neurological recovery occurs in significant proportion of patients. In the series reported by Sepal *et al.*,[19] 69% had neurological improvement after surgery while 12% remained unchanged and 16% deteriorated. Sensory deficit occurred in four of the 15 (26.66%) patients who were alive at follow-up. Krishnan *et al.*,[10] reported improvement by one or more grades in 13 of the 21 (61.91%) patients while 7 (33.37%) patients maintained their preoperative status. George *et al.*,[1] reported marked improvement in 90.5% patients and neurological deterioration in 5% of patients. There was no immediate postoperative death in this series. In our study following operation 26 (81.25%) patients improved neurologically, three (9%) patients remained unchanged, two (6.25%) patients deteriorated and there was one (3.1%) death.

In conclusion C_1 and C_2 nerve sheath tumors have special features as compared to other spinal sites and the tumors at this region should be considered as a separate clinical entity. Surgical procedures are to be done early in disease course to achieve better outcome.

References

1. George B, Lot G. Neurinomas of the first two cervical nerve roots: A series of 42 cases. J Neurosurg 1995; 82:917-23.
2. Kernighan J. Tumours of the spinal cord. Arch Patrol 1941; 32:843-83.
3. Levy WJ, Latched J, Hahn JF, Sawhny B, Bay J, Dohn DF. Spinal neurofibromas: A report of 66 cases and a comparison with meningioma. Neurosurgery 1986; 18:331-4.
4. Cohen L, Macramé D. Tumors in the region of foramen magnum. J Neurosurgery 1962; 19:462-9.
5. Geodetic B. Pallone A. Benign intramedullary tumors of the foramen magnum. Adv Tech Stand Neurosurgery 1988; 16:83-120.
6. Yasuoka S, Okazaki H, Daube JR, MacCarty CS. Foramen magnum tumors. Analysis of 57 cases of benign extramedullary tumors. J Neurosurg 1978;49:828-38.
7. Bartholomew JC, Crocked A. Bilateral posterolateral approach to mirror-image C-2 neurofibromas: Report of four cases. J Neurosurg 2001; 94:292-8.
8. McCormick PC, Stein BM. Spinal cord tumors in adults. In: Youmans JR, editor. Neurological Surgery. Vol. 4. 4th ed. Philadelphia: WB Sounders Company; 1994. p. 302-23.
9. Klekamp J, Samii M. Surgery of spinal nerve sheath tumors with special reference to neurofibromatosis. Neurosurgery 1998; 42:279-90.
10. Krishnan P, Behari S, Banerji D, Mehrotra N, Chabra DK, Jain VK. Surgical approaches to C_1-C_2 nerve sheath tumors. Neurol India 2004; 52:319-24.
11. Welling B, Park YK, Al-Mefty O. Primary extramedullary tumors of the craniovertebral junction. In: Dickman CA, Spetzler RF, Sonntag VK, editors. Surgery of the craniovertebral junction. 1st ed. New York: Theime; 1988. p. 240-2.
12. Yasargil MG, Mortara RW, Curcic M. Meningiomas of basal posterior cranial fossa. Adv Tech Stand Neurosurg 1980; 7:3-15.
13. Smith DA, Schmeidek HH. Tumors of the nerve sheath involving the spine. In: Sundaresan N, Schmeidek HH, Schiller A, Rlosenthal DM, editors. Tumors of the spine: Diagnosis and clinical management. Philadelphia: WB Saunders; 1980. p. 226-7.
14. Zeidman SM, Ellenbogen RC, Ducker TB. Intradural tumors. In: Clarke CR, editor. The Cervial spine. 3rd ed. Philadelphia: Lippincott Raven; 1996. p. 487-603.
15. Banerji D, Behari S, Jain VK, Pandey T, Chhabra DK. Extreme lateral transcondylar approach to the skull base. Neurol India 1999; 47:22-30.
16. Pritz MB. Evaluation and treatment of intradural tumors located anterior to the cervicomedullary junction by a lateral suboccipital approach. Act Neurochir (Wein) 1991; 1213:74-81.
17. Sen CN, Sekhar LN. An extreme lateral approach to intradural lesions of the cervical spine and foramen magnum. Neurosurgery 1990; 27:197-204.
18. Nanda A, Vincent DA, Vannemreddy PS, Baskaya MK, Baskaya CA, Chanda A. For lateral approach to intradural lesions of the foramen magnum without resection of the occipital condyle. J Neurosurg 2002; 96:302-9.
19. Seppala MT, Haltia MJ, Sankila RJ Jaaskelainen JE, Heiskanen O. Long Term outcome after removal of spinal neurofibroma. J Neurosurg 1995; 82:572-7.

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