Turkish Neurosurgery 2010, Vol: 20, No: 1, 73-76

Case Report

Giant Craniocervical Junction Schwannoma Involving the Hypoglossal Nerve: Case Report

Kraniyoservikal Bileşke Yerleşimli Dev Schwannoma Olgusu

Serdar KABATAS[1]
Tufan CANSEVER[2]
Cem YILMAZ[3]
Ebru DEMIRALAY[4]
Serdar CELEBI[5]
Hakan CANER[6]

1,2,3,6 Başkent University, Faculty of Medicine, Department of Neurosurgery, Ankara, Turkey
4 Başkent University, Faculty of Medicine, Department of Pathology, Istanbul, Turkey
5 Başkent University, Faculty of Medicine, Department of Anesthesiology, Istanbul, Turkey

ABSTRACT

OBJECTIVE: The authors present the case of a dumbbell-shaped schwannoma of the upper cervical spine involving the ventral rami of C-2 sensory root and rising through the foramen magnum up to the pontobulbar junction. The 27-year-old male patient complaining of hoarseness, imbalance and experiencing cervical pain and cervical muscle contractions for 2 months was admitted to the hospital. The cervical T1 and T2-weighted magnetic resonance (MR) images revealed the presence of a slightly hyperintense left C1–2 intra–extradural lesion which had eroded the clivus and odontoid process and enlarged the intervertebral foramen and was rising up to ponto-bulbar junction. A posterior approach was used to perform a suboccipital craniectomy and C1-2 laminectomy, including opening of the dura mater and gross-total removal of the lesion. The cerebrospinal MR image of the patient obtained at the early post-operative period revealed total removal of the lesion. The patient had hypoglossal nerve palsy and mild hemiparesis on the left side which had regressed almost totally at the 3-month follow-up. The far-lateral approach with the patient in the sitting position is very important and facilitates the total removal of the schwannoma. Simple suboccipital craniectomy provided enough exposure for total removal in this case.

KEYWORDS: Dumbbell-shaped, Schwannoma, Hypoglossal nerve, Spinal

ÖZ

AMAÇ: Schwannomlar spinal ve kranial sinirlerden kaynaklanan tüm intrakraniyal tümörlerin %8'ini oluşturur. Kraniyoservikal bileşkede oldukça nadir görülürler. Klinik bulguları bulundukları bölgeye göre oldukça değişkendir ve genellikle nörofibromatozisle birlikte görülür. Üst servikal bölgede yerleşen C2 duyusal kökünden kaynaklanarak foramen magnumdan geçip pontobulber bileşkeye kadar uzanım gösteren dambıl schwannom olgusu sunuldu.

OLGU: İki aydır süren horlama, dengesizlik ve üst servikal bölgede ağrı ve spazm şikâyetleriyle başvuran 27 yaşındaki erkek hastanın yapılan muayenesinde tüm kemik veter reflekslerinde hiperaktivite, sola trunkal ataksi ve lhermitte bulguları saptandı. T1 ve T2 ağırlıklı MR incelemelerinde heterojen yapıda milimetrik kalsifikasyon odakları olan sol C1–2 aralığından ekstradural bölgeye uzanım göstererek periferik bölgede hipoglossal sinir ile birleşen, klivus ve odontoid çıkıntı erozyonuna sebep olarak pontobulber bileşkeye uzanım gösteren kitle lezyonu saptandı. Suboksipital kraniyektomi ve C1–2 laminektomi ile yaklaşılan tümör tam olarak çıkartıldı. Lezyonun histopatolojik incelemesi sonucunda schwannom olduğuna karar verildi. Ameliyat sonrası erken dönemde hastanın sol hipoglossal sinir parezisi, hafif düzeyde sol hemiparezisi mevcuttu ve bu bulguları 3 ay içinde tama yakın geriledi.

SONUÇ: Bu tür vakalarda genelde suboksipital kraniyektomi ve farlateral transkondiler yaklaşım kullanılsa da suboksipital kraniyektomi ile yeterli görüş alanı sağlandı.

ANAHTAR SÖZCÜKLER: Dambıl, Schwannom, Hipoglossal sinir, Spinal

Received : 11.06.2009
Accepted : 27.07.2009

Correspondence address:
Tufan CANSEVER
E-mail: drtufan@gmail.com

INTRODUCTION

Schwannomas are benign tumors of the spinal and cranial nerves and represent about 8% of all intracranial tumors. Schwannomas located at the craniocervical region are rare. The clinical presentation is variable, depending on location, and can be associated with neurofibromatosis.7) We report a case with an unusual location of a spinal schwannoma presenting with hypoglossal nerve palsy associated with cerebellar signs and treated surgically with excellent result.

CASE REPORT

A 27-year-old right-handed male patient complaining of hoarseness, imbalance and cervical pain together with cervical muscle contractions for two months was admitted to the hospital. He had a ten-year history of occasional difficulty in cervical movements, a two-year history of gradually progressive gait disturbance and a two month history of hoarseness. A preoperative neurological examination revealed hyperreflexia in all extremities, partial hypoglossal nerve palsy on the left side, truncal ataxia to the left side and positive lhermitte sign. Postoperatively, the patient experienced mild left-sided hemiparesis and swallowing and breathing difficulties, which resolved in the early postoperative follow-up period.

Radiological findings

Magnetic resonance imaging (MRI) revealed an intradural-extradural dumbbell shaped, lobule contoured, and heterogenic mass with an intradural component located ventrally at the level of C1–C2 that was rising through the foramen magnum to the pontobulbar junction. The lesion was compressing the bulbus, both inferior cerebellar pedicles and the fourth ventricle (Figure 1 A,B). Cranial axial and coronal MRI examinations demonstrating the mass showed diffuse enhancement after intravenous Gadolinium injection (Figure 1 C,D). Post-operative cranial coronal and sagittal MRI showed total removal of the tumor. No contrast enhancement was seen at the operational area when IV contrast (gadolinium) was administered (Figure 2 A,B).

Surgical Findings

The tumor was totally removed via a posterior approach with a median incision and a C1–C2 laminectomy. The dura was then opened longitudinally and an additional T-shaped durotomy was made transversely toward the dural margin at the entrance of the nerve root (dural ring) to open the

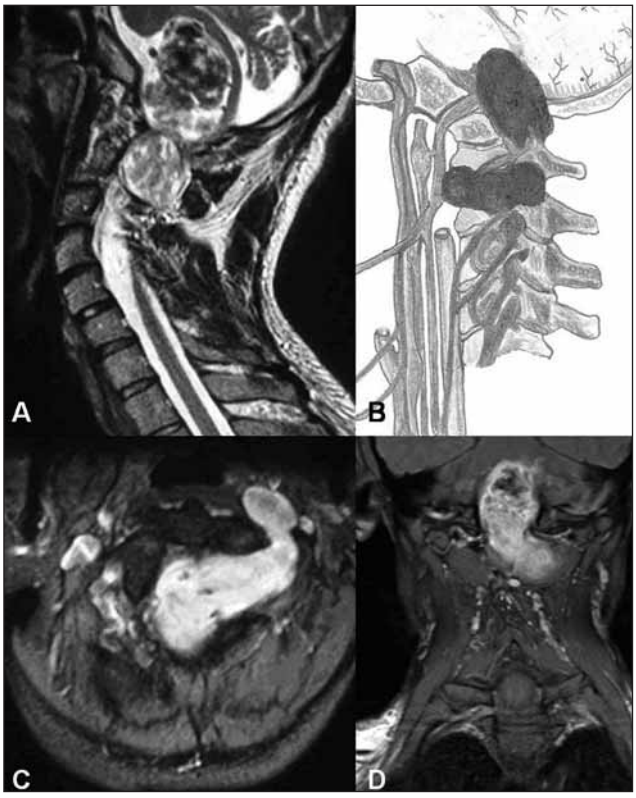

Figure 1: *Cranial sagittal T2-weighted MRI examination (**A**) and sagittal section illustration (**B**) showing an intradural-extradural dumbbell shaped, lobule contoured, and heterogenic mass with the intradural component, located ventrally at the level of C1–C2 which was rising through the foramen magnum to the pontobulber junction. The lesion was compressing the bulbus, both inferior cerebellar pedicles and the fourth ventricle. Cranial axial (**C**) and coronal MRI examinations (**D**) demonstrating the mass enhanced diffusely after intravenous Gadolinium injection.*

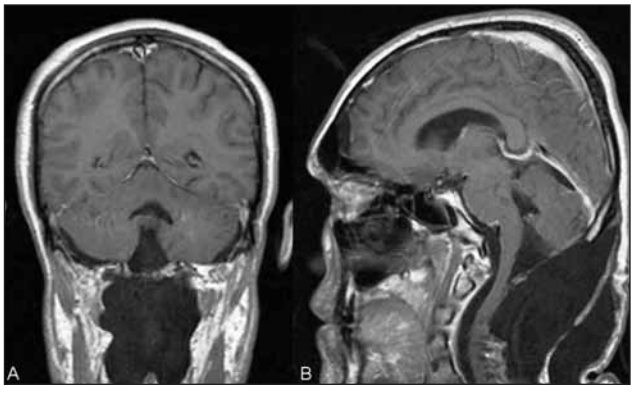

Figure 2: *Post-operative cranial coronal (**A**) and sagittal (**B**) MRI showed total removal of the tumor. No contrast enhancement was seen at the operational area when IV contrast (gadolinium) was administered.*

dural ring. The pontobulbar junction, cerebellum and medulla spinalis were exposed to the C3 level. The intradural component, which was relatively hard and did not adhere to the spinal cord, was extracted after a microsurgical ultrasonic aspirator was used to debulk the mass. The intradural mass was removed first and the extradural part was then removed by resecting the cervical C2 nerve branches distal to the lateral margin of the tumor and then dividing it in the part close to the dura. All affected C2 sensory and motor rootlets, the normal parts of which were elongated and tortuous, were sectioned, and the dura was closed primarily.

Histolopatological findings

The result of the intraoperative pathological examination of the cut end of the cervical nerve branches was negative. Histopathological examination showed typical features of a schwannoma; Antoni A areas (densely cellular, arranged in short bundles or interlacing fascicles) and Antoni B areas (fewer cells, organized with more myxoid component) (Figure 3 A,B). The tumor cells were bland, uniform spindle cells with wavy nuclei. The tumor showed extensive hyalinization and calcification and Antoni A areas constituted approximately 10% of the tumor volume (Figure 3 C,D).

DISCUSSION

Spinal schwannomas, which account for approximately 30% of primary spinal tumors11), originate predominantly from the sensory root of the spinal nerve2) and occasionally from its motor root.13) Although the cervical4), thoracic10), or lumbar predilection of spinal neuromas has been

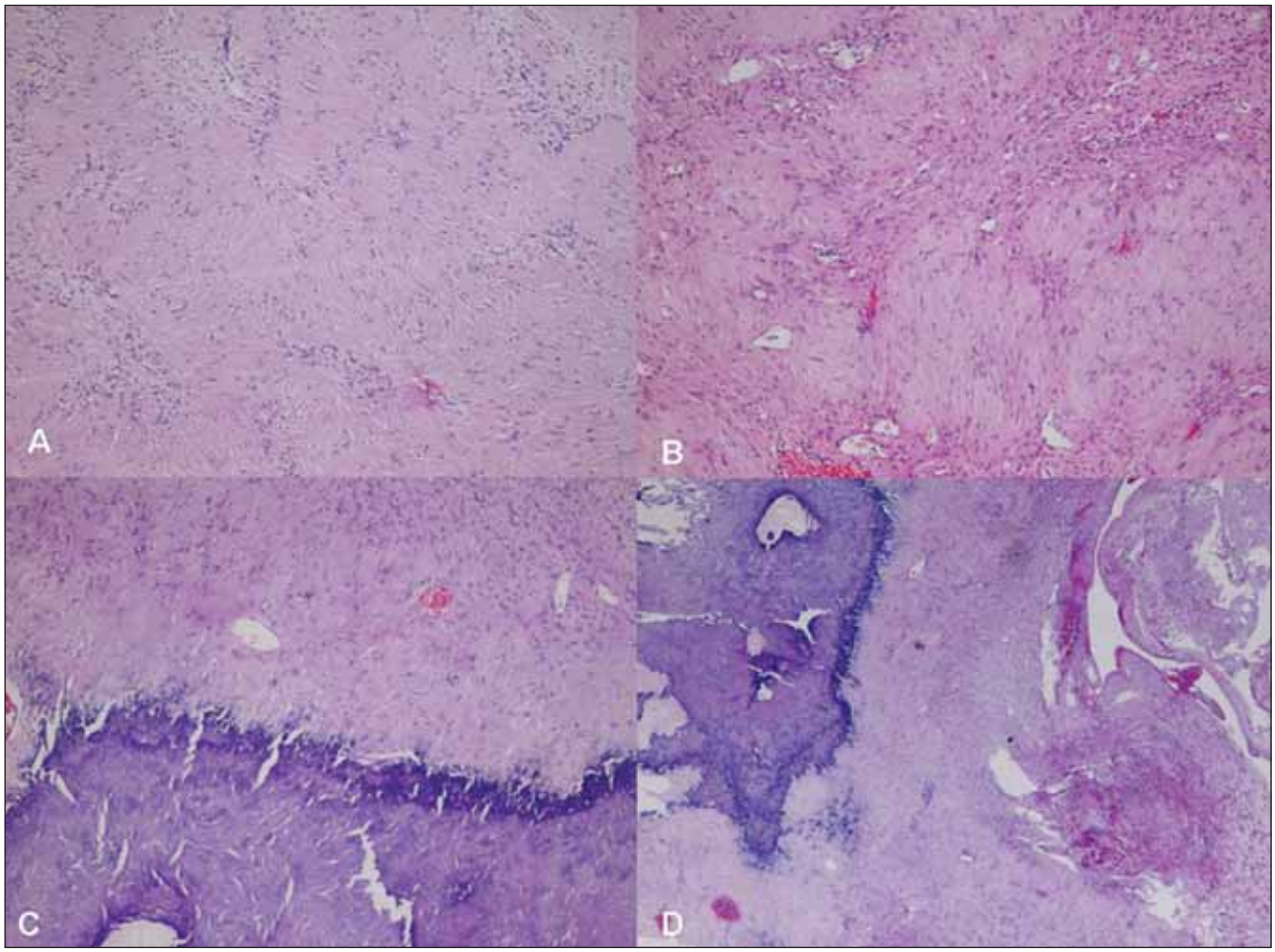

Figure 3: *Antoni A areas (densely cellular, arranged in short bundles or interlacing fascicles) and Antoni B areas (less cells, organized, more myxoid component) (**A, B**) (Hemotoxylin and eosin, original magnification X 100). The tumor cells were bland, uniform spindle cells with wavy nuclei. The tumor showed extensive hyalinization and calcification and Antoni A areas constituted approximately 10% of the tumor volume (**C,D**) (Hematoxylin and eosin, original magnification x 100).*

mentioned10), neuromas of the first two cervical nerve roots are uncommon, accounting for only 5.3% of all spinal neuromas and 18% of cervical neuromas.1)

The term "dumbbell" is used to reflect the tumor growth pattern. It is classified in relation to the intervertebral foramen.12) However, C2 neuromas are anatomically exceptional because there is no intervertebral foramen between C1 and C2. Therefore, dumbbell neuromas of the C2 nerve root will have an intraspinal-extraspinal canal and/or intradural-extradural pattern. Intradural tumors originating from within the nerve sheath or intervertebral foramen are more likely to extend ventrally toward the spinal cord.6) Extension of a C2 neuroma to the hypoglossal nerve causing hoarseness due to hypoglossal nerve involvement has never been described. This route of this extension is the anastomosis between the distal upper cervical plexus and hypoglossal nerve.

Various surgical approaches for dumbbell cervical tumors, such as a single posterolateral, anterolateral1,3,8) or lateral approach9,14), and combined posterior and anterior or anterolateral approaches in two stages5), have been described, together with their respective advantages and disadvantages, as well as limitations of the exposed area. Kyoshima et al6) considered that dumbbell C2 schwannomas can be satisfactorily managed with a posterior approach, which provides enough extraspinal and intraspinal access without the need to be concerned with postoperative spinal instability, because of the anatomic lack of an intervertebral foramen between C1 and C2, and the very wide C1–C2 interspace. Division of the proximal segment of the cervical C2 nerve root generally produces only a very mild sensory and motor deficit, if any, and then in a very limited area because of distal anastomosis of the cervical plexus.1) Resection of the cervical C2 nerve branches distally to the lateral margin of the tumor in our patient, caused motor impairment of the hypoglossal nerve due to anastomosis between the hypoglossal nerve and upper cervical plexus or may be due to hypoglossal nucleus impairment by the excision of the tumor from the brainstem. Hypoglossal nucleus impairment was thought to be less likely because the ventrally located intradural component was extracted after debulking, without damage to the spinal cord or brainstem as the intradural component of the tumor did not adhere to the brainstem.

Dumbbell-shaped schwannoma of the craniocervical junction based on the motor root of the cervical spine extending to the hypoglossal nerve is a rare clinical entity. Radiological techniques are not always helpful in identifying these lesions and a schwannoma should be included in the differential diagnosis of lesions in this area.

REFERENCES

1. George B, Lot G: Neurinomas of the first two cervical nerve roots: A series of 42 cases. J Neurosurg 82: 917–923, 1995
2. Hajjar MV, Smith DA, Schmidek HH: Operative Neurosurgical Techniques: Indications, Methods, and Results. Philadelphia: W. B. Saunders Co, 2000: 1843–1854
3. Kaynar MY, Hanci M, Sarioğlu AC: Intraspinal schwannoma of the accessory nerve. Br J Neurosurg. 13(4):429-31, 1999.
4. Hori T, Takakura K, Sano K: Spinal neurinomas: Clinical analysis of 45 surgical cases. Neurol Med Chir (Tokyo) 24: 471–477, 1984
5. Komisar A, Blaugrund SM, Camins M, Mangiardi J: Combined approach for excision of cervical nerve tumors with dural extension. Head Neck 15: 153–157, 1993
6. Kyoshima K, Uehara T, Koyama J, Idomari K, Yomo S: Dumbbell C2 schwannomas involving both sensory and motor rootlets: report of two cases. Neurosurg 53: 436-439, 2003
7. Leal Filho MB, Borges G, Ferreira A, França D, Mello P: Schwannoma of the craniocervical junction: Surgical approach of two cases. Arq Neuropsiquiatr 61: 639-641, 2003
8. Lot G, George B: Cervical neuromas with extradural components: Surgical management in a series of 57 patients. Neurosurg 41: 813–822, 1997
9. Ryu H, Nishizawa S, Yamamoto S: One-stage removal of a large dumbbell-shaped cervical neurinoma without laminectomy or interbody fusion in a child. Br J Neurosurg 13: 587–590, 1999
10. Salah S, Horcajada J, Perneczky A: Spinal neurinomas: A comprehensive clinical and statistical study on 47 cases. Neurochirurgia (Stuttg) 18: 77–84, 1975
11. Seppala MT, Haltia MJ, Sankila RJ, Jääskeläinen J, Heiskanen O: Long-term outcome after removal of spinal schwannoma: A clinicopathological study of 187 cases. J Neurosurg 83: 621–626, 1995
12. Shephard RH, Sutton D: Dumb-bell ganglioneuromata of the spine with a report of four cases. Br J Surg 45: 305–317, 1958
13. Urich H, Tien RD: Russel and Rubinstein's Pathology of Tumors of the Nervous System. 6 ed. London: Arnold, 1998: 141–193
14. Verbiest H: A lateral approach to the cervical spine: Technique and indications. J Neurosurg 28: 191–203, 1968