



CAUDA EQUINA SYNDROME DUE TO SPINAL SCHWANNOMA WITH INTRATUMORAL HEMORRHAGE

İNTRATÜMÖRAL KANAMA SONRASI KAUDA EKUİNA SENDROMUNA NEDEN OLAN SPİNAL SCHWANNOMA

Ali Erdem YILDIRIM¹,
Cagatay OZDOL²,
Denizhan DIVANLIOĞLU¹,
Nuri Eralp CETİNALP¹,
Ahmed Deniz BELEN³

¹Ankara Numune Research and Education Hospital, Department of Neurosurgery, Ankara.

²Antalya Research and Education Hospital, Department of Neurosurgery, Antalya.

³Ankara Numune Research and Education Hospital, Department of Neurosurgery, Ankara.

SUMMARY

Cauda equina syndrome is defined as a group of symptoms including lower back pain, sciatica, saddle anesthesia, decreased rectal tone, perianal areflexia or hyporeflexia, bowel and bladder dysfunction, and lower-extremity weakness. There are several causes of this syndrome, including trauma, central disc protrusion, hemorrhage, and rarely, neoplastic invasion. In this case, we report cauda equina syndrome caused by intratumoral hemorrhage of a spinal schwannoma at the L4 level. Surgical resection of the tumor significantly improved the neurological symptoms of the patient.

Key words: Cauda equina syndrome, intratumoral hemorrhage, spinal schwannoma

Level of evidence: Case report, Level IV

ÖZET

Kauda ekuina sendromu; bel ve bacak ağrısı, ayak tarzı hipoestezi, 38 anal hiporefleksi, bağırsak ve mesane fonksiyon bozuklukları ile birlikte alt ekstremitelerde kuvvetsizlik semptomlarından oluşan klinik bir tablodur. Bu sendrom sıklıkla travma, santral disk protrüzyonu ve kanamalara bağlı gelişirken daha nadir olarak da tümöral lezyonlar nedeniyle oluşabilmektedir. Bu vaka takdiminde L4 seviyesine yerleşmiş bir intradural schwannoma olgusunun, tümör içine kanaması ile oluşan kauda ekuina sendromu sunulmaktadır.

Anahtar kelimeler: Kauda ekuina sendromu, tümör içine kanama, spinal schwannoma

Kanıt Düzeyi: Olgu Sunumu, Düzey IV

Address: Ali Erdem YILDIRIM,
Ankara Numune Research and
Education Hospital, Department of
Neurosurgery, Ankara.

Tel: 0505 6875519,

Fax: 0312 3103460

E-mail: alierdemyildirim@gmail.com

Received: 1st July, 2013

Accepted: 16th September, 2013

INTRODUCTION:

Cauda equina syndrome (CES) is a neurosurgical emergency caused by compression of the nerves below the level of the conus medullaris. The signs and symptoms are complex, with pain (unilateral or bilateral sciatica and lower back pain) and findings of sensory (in the saddle and perianal region), motor (weakness in lower extremity) and reflex (bulbocavernous, patellar and achilles) dysfunction^{1,9}.

Schwannomas in CES are generally sporadic, but they can also be seen in neurofibromatosis type 2. They arise from the transition zone of the central and peripheral myelin sheaths, called the Obersteiner-Redlich zone¹. The peak age at presentation is 40 years, with progressive symptoms indicating a slow-growing tumor. Spinal schwannomas commonly affect the sensory nerve roots in the cervical and lumbar regions⁵. Schwannomas have been reported as being associated with cauda equina syndrome due to subarachnoid or subdural hematomas; however, association with an intratumoral hemorrhage is unique^{3,7}. In the following report, a patient with cauda equina syndrome caused by an intratumoral hemorrhage is reported.

CASE REPORT:

A 24-year-old female was admitted to the neurosurgery clinic with complaints of severe back pain, paraparesis and urinary and fecal incontinence. She had been suffering from lower back pain for two years, which had improved two days previously. Neurological examination showed severe weakness of her bilateral lower extremities and bilateral sensory loss below the L1 level, with absence of rectal tonus.

Magnetic resonance imaging (MRI) demonstrated an intradural lesion, hyperintense on T1-weighted images and heterogeneously hypointense on T2-weighted images, located at the L4 level (Figure-1).

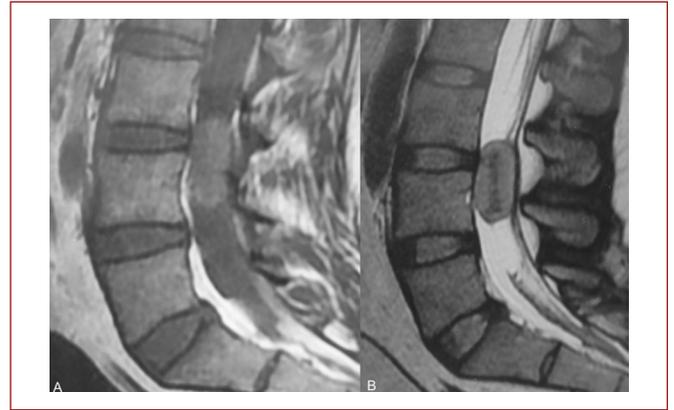


Figure-1. (A) MRI demonstrating an intradural lesion located at the L4 level that is slightly hyperintense on the T1-weighted image and (B) heterogeneously hypointense on the T2-weighted image.

She received urgent surgery and a L4 laminectomy was performed. The hemorrhaged tumor was seen in the cauda fibers (Figure-2), and total resection of the tumor was achieved using microsurgical techniques at the expense of an involved nerve root. Histopathological evaluation indicated a schwannoma with positive S100 immunostaining. In the early postoperative period, although her neurological deficits did not improve, there was no pain. She was taken into a rehabilitation program after discharge from the ward at the third day postoperatively. During the follow-up period, a total recovery in her neurological status was achieved six months after surgery.

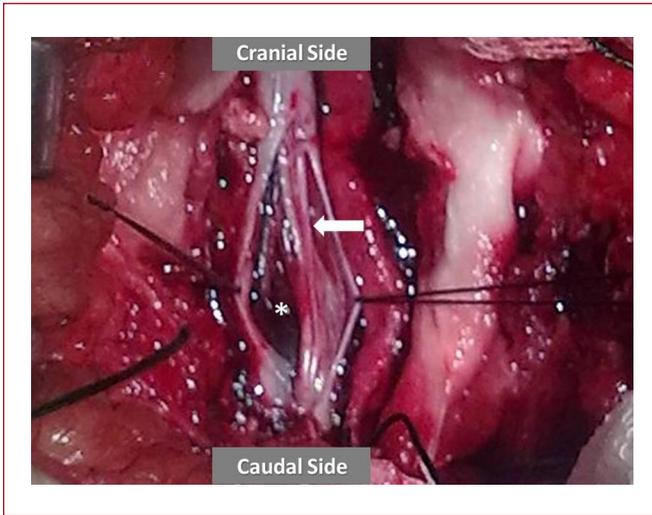


Figure-2. Intraoperative photograph of the lesion with intratumoral hemorrhage (asterisk) compressing the cauda equina fibers (arrow).

DISCUSSION:

Cauda equina syndrome is a very specific constellation of symptoms, including sciatica, lower back pain, saddle and perianal hypoesthesia or analgesia, decreased rectal tonus, lack of the bulbocavernosus, patellar, and achilles reflexes, bowel and bladder dysfunction, and lower extremity weakness¹. CES was first reported in 1934 by Mixter and Barr¹⁰. The most common cause of CES is central lumbar disc herniation⁹. However, intratumoral hemorrhage caused by ectatic and hyalinized blood vessels in a neoplasm has also been reported in the etiology⁶. Among all spinal tumors, the incidence of intratumoral hemorrhage is highest among ependymomas, followed by schwannomas⁶. Tumors around the cauda equina region may apply a traction force in the rostral to caudal direction along the spinal axis, facilitating intratumoral or subarachnoid bleeding⁶.

Tumors of that region can reach considerable sizes with no symptoms, as the area is large

and the nerve roots are mobile. The mean duration for diagnosis was reported to be four years by Cervoni et al.². Spinal schwannomas are generally slow-growing tumors causing progressive symptoms^{5,8}.

Cauda equina schwannomas presenting with acute paraparesis and urinary incontinence are usually associated with intratumoral hemorrhages. In the literature, trauma or anticoagulant drugs have been blamed for causing subarachnoid or subdural hemorrhages in cases with spinal schwannoma, which in turn resulted in CES^{4,6}. This case is unique for presenting CES caused by an intratumoral hemorrhage of a spinal schwannoma without any precipitating factors. A review of the literature also revealed that this case shows an unusual bleeding site for a spinal schwannoma, at the L4 level as opposed to at the thoracolumbar junction.

Intratumoral hemorrhage causing CES is quite rare, but still an urgent condition that should not be missed. Therefore, in cases of CES, MRI evaluation should be performed as soon as possible, in order to detect unusual pathologies, and early surgical decompression of the nerve roots should be carried out, as well as tumor resection in this case, in order to improve the neurological outcome.

REFERENCES

1. Bagley CA, Gokaslan ZL. Cauda equina syndrome caused by primary and metastatic neoplasms. *Neurosurg Focus* 2004; 16: 3.
2. Cervoni L, Celli P, Scarpinati M, Cantore G. Neurinomas of the cauda equina clinical analysis of 40 surgical cases. *Acta Neurochir(Wien)* 1994; 127: 199-202.

-
3. Chalif DJ, Black K, Rosenstein D. Intradural spinal cord tumor presenting as a subarachnoid hemorrhage: magnetic resonance imaging diagnosis. *Neurosurgery* 1990; 27: 631-634.
 4. Cohen ZR, Knoller N, Hadani M, Davidson B, Nass D, Ram Z. Traumatic intratumoral hemorrhage as the presenting symptom of a spinal neurinoma. Case report. *Neurosurgery* 2000; 93: 327-329.
 5. Conti P, Pansini G, Mauchaty H, Capuano C, Conti R. Spinal neurinomas: retrospective analysis and long term outcome of 179 consecutively operated cases and review of the literature. *Surg Neurol* 2004; 61: 34-44.
 6. Ichinose T, Takami T, Yamamoto N, Tsuyuguchi N, Ohata K. Intratumoral hemorrhage of spinal schwannoma of the cauda equina manifesting as acute paraparesis-case report. *Neurol Med Chir* 2009; 49: 255-257.
 7. Inoue T, Miyamoto K, Kushima Y, Kodama H, Nishibori H, Hosoe H, Shimizu K. Spinal subarachnoid hematoma compressing the conus medullaris and associated with neurofibromatosis type 2. *Spinal Cord* 2003; 41: 649-652.
 8. Jinnai T, Koyama T. Clinical characteristics of spinal nerve sheath 168 tumors: analysis of 149 cases. *Neurosurgery* 2005; 56: 510-615.
 9. Kostuik JP, Harrington I, Alexander D, Rand W, Evans D. Cauda equina syndrome and lumbar disc herniation. *J Bone Joint Surg* 1986; 68-A: 386-391.
 10. Mixter WJ, Barr JS. Rupture of the intervertebral disc with involvement of the spinal canal. *N Engl J Med* 1934; 211: 210-214.