Intradural lumbar cystic schwannoma

Cheng-Ta Hsieh, MD, Wen-Chiuan Tsai, MD, Ming-Ying Liu, MD.

ABSTRACT

يعد الورم الشفاني الكيسي القطني الذي يظهر داخل الجافية من الأورام النادرة جداً، وقد تم الإفصاح عن 10 تقارير تناولت هذا المرض في الأدب الطبي. ويظل تشخيص وعلاج هذا المرض من التحديات التي يواجهها الأطباء. نستعرض في هذا المقال حالة نادرة لمريضة تبلغ من العمر 51 عاماً، وقد أتت إلى المستشفى بعد إصابتها بألم في أسفل الظهر، بالإضافة إلى ألم منتشر في المنطقة العلوية من الفخذ. لقد أظهر تصوير الرنين المغناطيسي للعمود القطني وجود كتلة كيسية داخل الجافية، وكان حجمها التقريبي35 x 17 x 18 مم، وقد ظهرت هذه الكتلة في الفقرتين L4 وL5، وكانت نسبة تركيز الإشارات المُرسلة منها مشابة لتلك المُرسلة من السائل الدماغي الشوكي. وفي أثناء العملية الجراحية تم العثور على كتلة كيسية محاطة بالكامل بمحفظة، وأكد التشخيص النسيجي للعينة المأخوذة من الكتلة تشخيص الحالة بالورم الشفاني. كما تمت مناقشة الأدب الطبي الذي ناقش هذا الورم النادر في المقال.

Intradural lumbar cystic schwannomas are a very rare entity and only 10 case reports have been reported in the literature. The diagnosis and management remains a challenge for clinical physicians. Herein, we report a 51-year-old female presenting with lower back pain and radiating pain at the left upper thigh. Magnetic resonance images of lumbar spine demonstrated an intradural cystic mass approximately 18 x 17 x 35 mm in size occupying L4 to L5, which appeared as the same signal intensity as CSF. At operation, a well encapsulated cystic mass was found. The pathological examination confirmed a diagnosis of schwannoma. The relevant literature was also reviewed.

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Intradural spinal schwannomas are well-described benign lesions, accounting for approximate 30% of primary spinal tumors.[1] However, intradural lumbar cystic schwannomas are a very rare entity and only 10 cases have been described in the literature.[1-5] Herein, we report a 51-year-old female with an intradural lumbar cystic schwannoma and review of the relevant literature. Our objective in presenting this particular case is to highlight the atypical radiological features of intradural spinal schwannoma.

Case Report. A 51-year-old female was admitted with a 4-month history of lower back pain, followed by radiating pain in the bilateral lower legs. She received open reduction and internal fusion at the L3-L4 level due to spondylolisthesis 9 years earlier, and a laminectomy and discectomy at the L5-S1 level due to herniated lumbar disc with root compression 7 years ago. At admission, physical examination revealed mild swelling in bilateral lower legs and foot. Neurological examinations showed dysesthesia in the L4-S1 dermatone. Weakness in the left lower legs (muscle power scored as grade 4) was noted. No abnormal deep tendon reflexes or pathological reflexes exist. The spinal radiographs revealed no endplate destruction, enlargement of spinal canal, or deformity of spinal structures. An MRI of the lumbar spine revealed an intradural cystic mass approximately 18 x 17 x 35 mm in size occupying the L4-L5 space, which appeared the same signal intensity as CSF (**Figure 1**). The adjacent cauda equina were displaced by this mass lesion. After intravenous gadolinium administration, the cystic mass showed a marginal enhancement (**Figure 2**). She underwent laminectomies from L4 to L5 for removal of the tumor. At operation, an encapsulated mass with abundant vessels in the capsule was found after opening the dura membrane. Only 2 small nerve roots were adhesive to the mass lesion. Other cauda equina was displaced and compressed by the mass. The tumor was totally removed. After opening the cystic mass, one hematoma with yellowish fluid was found. The pathological examinations revealed alternative Antoni A (hypercellular) and Antoni B (hypocellular) areas, which confirmed a diagnosis of schwannoma (**Figure 3**). Postoperatively, she had complete relief of symptoms

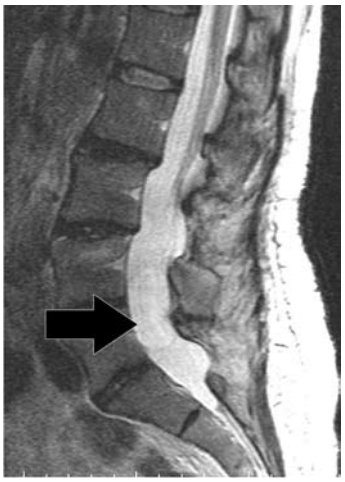

Figure 1 - Sagittal view of T2-weighted MRI revealing an intradural cystic tumor occupying L4-L5. The tumor appeared the same signal intensity as CSF.

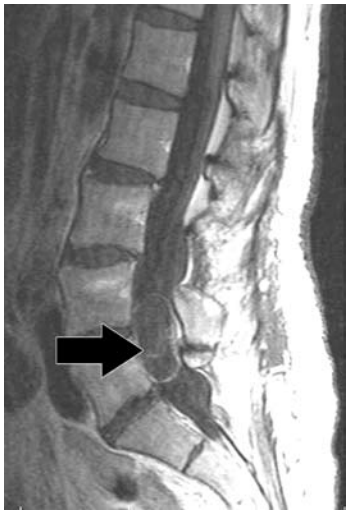

Figure 2 - After intravenous gadolinium administration, sagittal view of T1-weighted MRI revealing the cystic mass with a marginal enhancement.

and returned to work after discharge. No recurrent tumor was found during the follow-up period.

Discussion. Intradural spinal schwannomas are well-described slow growing benign tumors of the peripheral nervous system, arising from Schwann cells. The most common location of spinal schwannomas are the lumbar spine (48%).[6] It has been described that a schwannoma may display variable degenerative changes such as fibrosis, cytological atypia, calcification, hemorrhage, or cystic formations.[2] Cystic degenerations occurring in schwannomas are found in schwannomas of the orbital region, olfactory groove, tentorial hiatus, posterior cavernous sinus, presacral region, maxillary sinus, intramedullary spinal region, or intraventicular

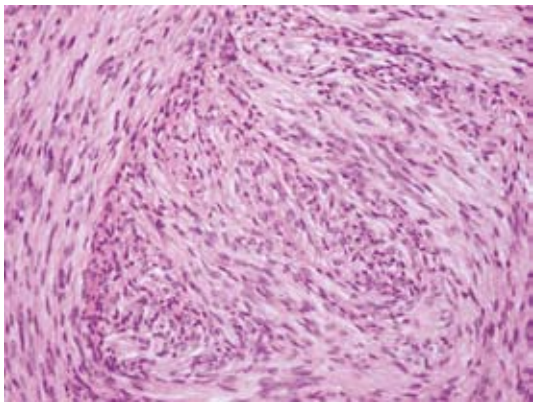

Figure 3 - Microscopically, the tumor was composed of alternative Antoni A (hypercellular) and Antoni B (hypocellular) areas (Hematoxylin & Eosin x 400).

region.[2] However, cystic schwannomas are rarely reported in the lumbar spine region and only 10 cases have been described in the literature.[5]

Schwannomas are usually encapsulated solid benign tumors. Various theories have been hypothesized to explain the cystic change occurring in schwannomas. Degeneration of the Antoni B portion of a neuroma can result in cyst formation and may then progress to form a larger cyst.[7,8] Central ischemic necrosis, thrombosis of the vessels with resultant necrosis plus hemorrhage or tumor neovascularity also can be caused by tumor growth resulting in cyst formation within the tumor.[1]

An MRI has been considered as the best tool to investigate the spinal cystic tumor. Generally, schwannomas appears as hypointensity on T1-weighted MR images and heterogenous intensity on T2-weighted MR images, based on the different components within the schwannoma.[9] The hypointensity on T2-weighted MR images often correspond to hemorrhage, dense cellularity or collagen deposition, whereas hyperintensity may represent cystic changes.[3] However, the radiological differential diagnosis of such a large spinal cystic tumor includes a cystic neurinoma, ependymoma, neurenteric cyst, epidermoid, bronchogenic cyst, cystic teratoma, Tarlov cyst, and arachnoid cyst.[5,8] A contrast study is preferred to differentiate schwannoma from other neoplasms. Rim enhancement of an intradural-extramedullary tumor on MRI should be considered as the diagnosis of schwannoma.[1,10] However, the exact diagnosis should be proven by pathological studies. The surgical intervention has been advocated if the neurological deficits are progressive. Total surgical excision is the best option to achieve the least chance of recurrence and greater chance of cure.[1,7-9]

In conclusion, our case showed a rare entity of intradural lumbar cystic schwannoma. The surgical outcome depended on the early diagnosis and complete excision.

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