

## Lumbar Cystic Intradural Schwannoma: A Case Report

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### Abstract

**Background Data:** Intradural lumbar cystic schwannomas are very rare entity and only about 10 case reports have been reported in the literature. The diagnosis and management remains a challenge for clinical physicians.

**Purpose:** Our objective in presenting this particular case is to highlight the atypical radiological features of intradural spinal schwannoma and how to deal with it surgically and follow up.

**Study Design:** case report.

**Material and Methods:** A case of cystic intradural lesion in a previously healthy 29 years old male is reported. Low back pain and lately right sciatica were the only symptoms. After pre-operative evaluation and preparation, this patient was operated on prone position under general anesthesia.

**Results:** Although the lesion originates from nerve tissue, only 50% of cases have a direct relation with the nerve. Hence complete excision without sacrificing nerve roots is usually feasible and is curative in most cases as in our case. The patient recovered totally intact without any motor, sensory or sphincteric disturbance. The postoperative histological picture of cystic schwannoma, which was not in the preoperative list of differential diagnoses, is discussed.

**Conclusion:** A successful surgical outcome depends on early diagnosis [before neurological damage has occurred], meticulous investigation [on any intractable nonspecific low back pain], and complete excision. (2014ESJ066)

**Keywords:** Intradural, Schwannoma, Cystic, Lumbar

## Introduction

Schwannoma (neurilemmoma) is a benign slow-growing encapsulated tumour arising from the myelinated nerve sheaths. Schwannomas, together with meningiomas, are the most common intradural tumours.<sup>1</sup> Degenerative changes like haemorrhage, calcification, and fibrosis are commonly seen in schwannomas, but cystic changes are rare. Such tumours have been reported in the orbital region,<sup>13</sup> olfactory groove,<sup>12</sup> tentorial hiatus, posterior cavernous sinus,<sup>4</sup> pre-sacral region,<sup>7</sup> pancreas,<sup>14</sup> maxillary sinus,<sup>11</sup> spinal cord,<sup>8,13</sup> lesser sac,<sup>15</sup> and within the ventricular system.<sup>14</sup> Intradural spinal schwannomas are well-described benign lesions, accounting for approximate 30% of primary spinal tumors.<sup>1</sup> However, intradural lumbar cystic schwannomas are very rare entity and only 10 cases have been described in the literature.<sup>2</sup> We report another such case in a 29-year-old man.

## Materials and Methods

A 29 years old Palestinian married male, presented with 12 months history of episodic low back pain, following mild trauma while playing soccer. One month before admission the patient developed severe, excruciating right sciatica without numbness or sphincteric dysfunction. There is nothing relevant in his past medical history.

On admission, he was noted to be in a good general condition. The tone, power and reflexes were normal, symmetrical in all four limbs and the plantar response was flexor bilaterally. The only neurological finding was diminution of pinprick sensation in right L1 and L2 dermatomes. The patient had no cutaneous stigmata of neurofibromatosis. The patient's blood investigations, coagulation profile and chest x-ray were all unremarkable. Plain lumbosacral spine x-ray showed no abnormality. Magnetic Resonance Imaging of the lumbosacral spine showed evidence of intradural oblong cystic mass at the level of L2, measuring 1.6 x 1.7 x 3.7 cm. The lesion shows a hypointense signal on T1 weighted images and mixed signal on T2 weighted images, along with rim enhancement after contrast injection.

After pre-operative evaluation and preparation, this patient was operated on prone position under general anesthesia. After low back antiseptic sterilization and op-site draping, midline skin

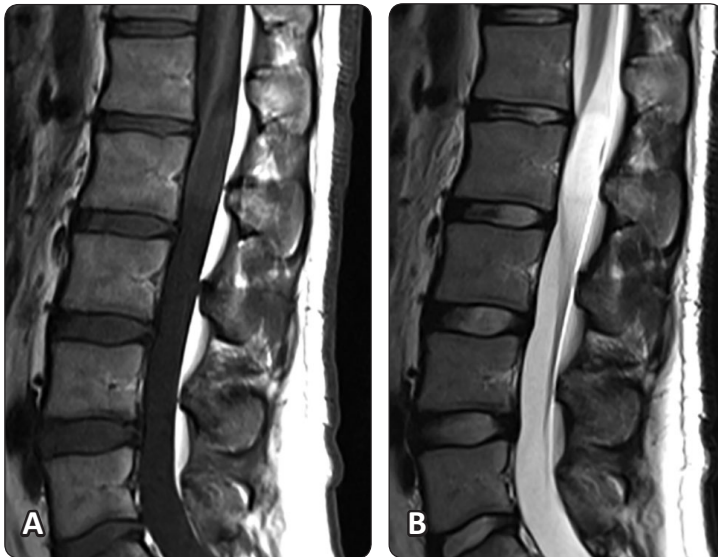
incision after localization with imaging intensifier was performed. Dissection in layers was done exposing L1 and L2 lamina with 25'' knife and bipolar coagulation. High speed drill laminectomy was done for L2 totally and the lower half of L1, exposing the thecal sac. Durotomy with 11'' knife was done then tenting the dura with 3/0 vicryl stitches. Dissection of the cystic lesion and bipolar coagulation of a small vascular territory going into the cyst were achieved, then removal of the cystic lesion in toto. Closure of the dura after adequate hemostasis was done, followed by closure in layers with Redyvac 12 in place for one day. Estimated blood loss was about 100cc only.

## Results

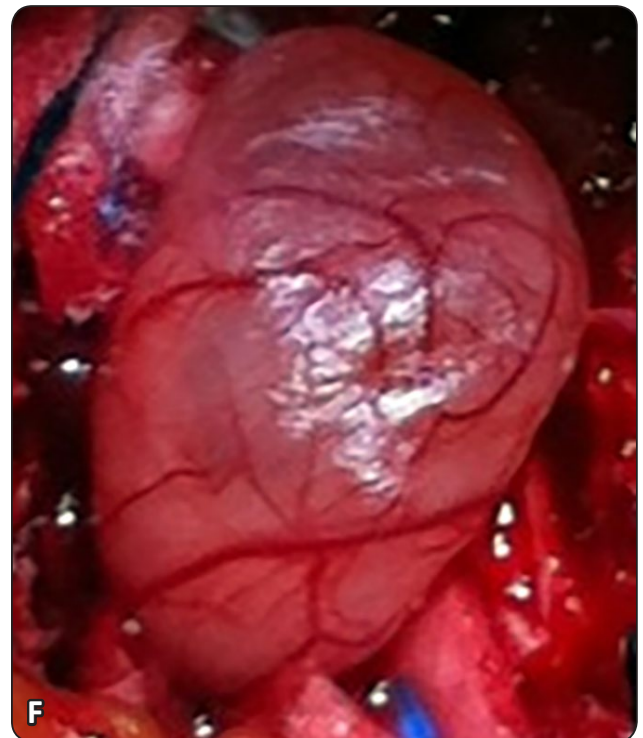
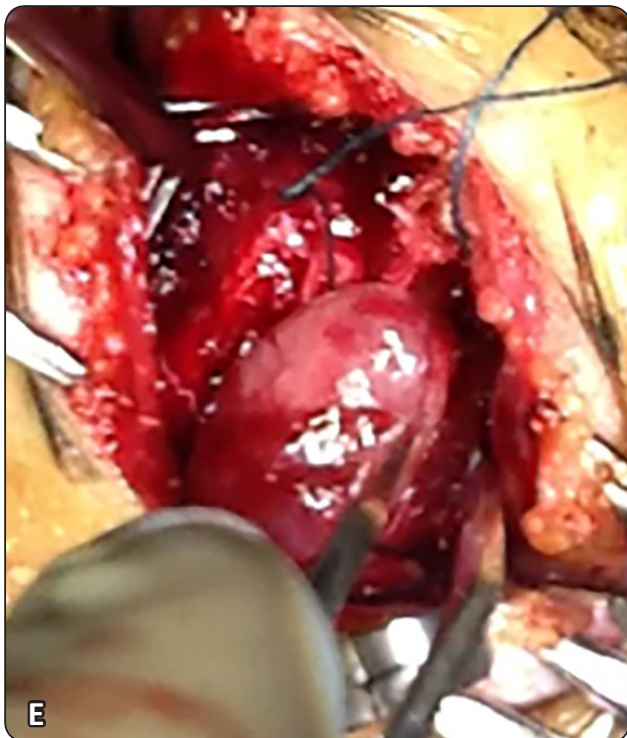
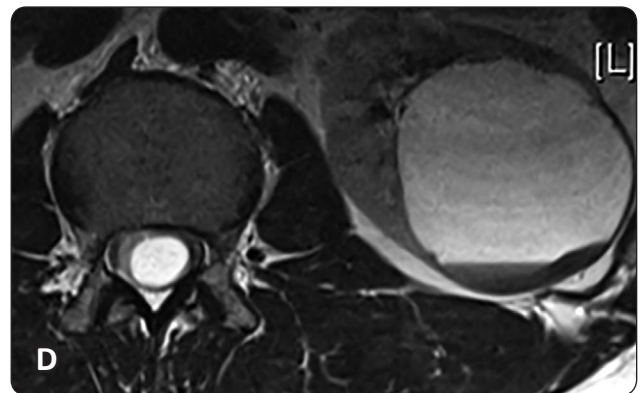
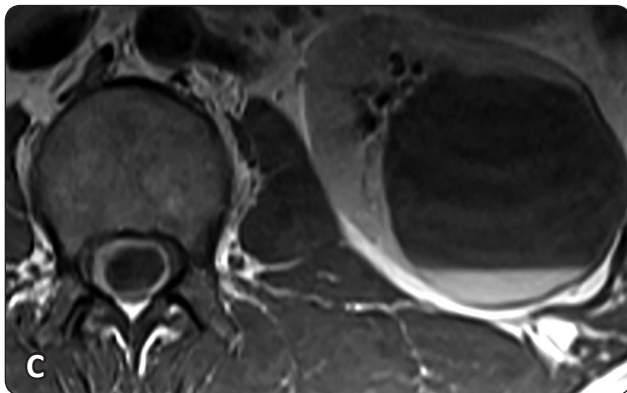
The patient recovered fully and managed to ambulate out of bed with a lumbar support in situ from the first postoperative day. The preoperative sensory deficit in the right L1 and L2 dermatomes settled quite nicely after 3 weeks from the date of surgery and the patient became neurologically free and follow up for one year clinically pain free and intact. Histological examination of the operative specimen (Figure 1) proved it to be a cellular schwannoma with no evidence of malignancy. No malignant cells were found in the cyst fluid either.

## Discussion

Schwannomas are benign tumors arising from the embryonic neural crest cells of the nerve sheath of peripheral and cranial nerves. The first case was reported in 1910.<sup>16</sup> Schwannomas have predilection for sensory nerves and tend to arise from the dorsal roots.<sup>3</sup> Nerve sheath tumors such as schwannomas and neurofibromas account for 30% of all intraspinal masses.<sup>10</sup> Schwannomas are most frequently seen in the lumbar region. The purely intradural nerve sheath tumors increases from 8% in the upper cervical to 80% in the thoracolumbar region.<sup>6</sup> This may be explained by the anatomic features of the spinal nerve roots, which have a longer intradural component at the caudal portion of the spinal axis. Coexistence with other tumors, like meningioma, can be seen in patients with neurofibromatosis. The presenting symptoms of lumbar schwannomas can be rather confusing which commonly leads to delayed presentations. This is attributed to the fact that they are slowly growing lesions and the surrounding anatomic environment is permissive,



**Figure 1.** (A): MRI sagittal T1WI showing intra dural hypointense Cystic lesion pushing the intra dural structures anteriorly opposite the body of L2 vertebrae, (B): MRI Sagittal T2WI showing hyper intense intra dural well circumscribed cystic lesion with mass effect on the intradural nervous structures opposite the body of L2, (C): Axial MRI T1WI showing the same intradural well circumscribed cystic lesion, (D): Axial MRI T2WI showing the same intradural well circumscribed leion, (E,F): Intra operative image showing the intra dural cystic lesion after complete dissection from the adjacent nervous structure and before getting it out of the surgical field.



the presenting symptoms are nonspecific and the affected patients are young and otherwise healthy. Other symptoms like intracranial subarachnoid hemorrhage, hydrocephalus, acute low back pain and nonspecific abdominal pain can be encountered with such lesions.

The radiographic findings are sometimes non-pathognomonic which can only point out to the presence of a cystic lesion, as it is the case in our patient. MRI is the preferred imaging modality. Schwannomas generally have low to intermediate signal intensity on T1 weighted images. On T2 weighted images, they may be heterogenous with focal areas of hyperintensity and hypointensity, corresponding to cyst formation, hemorrhage, dense cellularity and collagen deposition. With gadolinium contrast, the cystic lesion is dense and heterogenous at times, though smaller lesions may be homogenous. A rim enhancement of an intradural extramedullary tumor suggests the diagnosis of schwannoma.

Definitive verification of the diagnosis is certainly based on histological analysis. The hallmark histological feature of schwannoma is the Verocay bodies. The degree of cellularity and shape of the cells with its arranged pattern can differentiate between Antoni A and Antoni B types. Immunohistochemical staining can further aid in the diagnosis as benign schwannomas show diffuse immunoreactivity for S-100 protein.

Dense Antoni A pattern comprises more than 90% of the tumor area with a more uniform pattern, a lack of Verocay bodies and frequent lymphocytic infiltration. The cystic changes of schwannoma are likely attributed to mucinous degeneration, ischemic necrosis, hemorrhage and the formation of microcysts.

Although the lesion originates from nerve tissue, only 50% of cases have a direct relation with the nerve. Hence complete excision without sacrificing nerve roots is usually feasible and is curative in most cases. Prognosis is usually excellent with the exception of the melanotic variant, malignant forms and cases of neurofibromatosis.

As schwannomas are benign tumors with a slow growth rate, the diagnosis of extracranial schwannomas may pose a challenge to the care giver when few symptoms are observed. In the case we report, there is a striking contrast with the paucity of

symptoms and the size of the tumor. This highlights the importance of suspecting of a cystic lumbar nerve sheath tumor when symptoms associated to lumbar spinal cord, or nerve roots compression are encountered. Moreover, schwannomas should be included in the differential diagnosis of a cystic mass in the spinal region.<sup>5</sup>

The differential diagnosis of such cystic lesion includes cystic schwannoma, cystic neurofibroma, epidermoid, epidermoid cyst, arachnoid cyst, cystic lymphangiomas, cystic teratoma and cystic meningioma. Until 2011, only 11 predominantly cystic intradural schwannomas have been reported in the literature.<sup>9</sup>

## Conclusion

A successful surgical outcome depends on early diagnosis [before neurological damage has occurred], meticulous investigation [on any intractable nonspecific low back pain], and complete excision.

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## الملخص العربي

**الورم الكيسي القطني والذي يظهر داخل غشاء الأم الجافية**

المقدمة: يعد الورم الكيسي القطني والذي يظهر داخل غشاء الأم الجافية من الأورام النادرة. هذا وقد تداولت فقط إحدى عشر تقرير هذا النوع من الأورام ونحن هنا نعرض بالتفصيل حالة من هذه الحالات حيث يعد تشخيص وعلاج مثل هذه الحالات يحتاج إلى مهارة عالية.

تصميم الدراسة: تقرير حالة

الهدف: الهدف من عرض هذه الحالات هو إبراز الطرق الجديدة في تشخيص وعلاج مثل هذه الحالات النادرة

عينة المرضى: رجل ٢٩ سنة يعاني من ألم أسفل الظهر ممتد إلى الساق والقدم اليمنى.

النتائج: تم إزالة الورم بالكامل مع الحفاظ التام على كافة الأعصاب والأغشية المحيطة بكامل حالتها.

الخلاصة: التشخيص الدقيق والمبكر لمثل هذه الحالات واستخدام التقنيات الحديثة أثناء التدخل الجراحي يؤدي إلى نتائج جيدة.