

GIANT INVASIVE SCHWANNOMA OF CAUDA EQUINA WITH MINIMAL NEUROLOGIC DEFICIT: A CASE REPORT AND LITERATURE REVIEW

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A 53-year-old man presented with a history of slight weakness in the right lower limb. Giant invasive cauda equina schwannoma was diagnosed according to the criteria of Sridhar et al. Schwannomas are usually benign and common tumors arising from nerve sheath cells, particularly from sensory nerves. Giant invasive schwannomas, however, are rare, and most of patients with them present with severe neurologic deficits independent of daily activity, although in the case presented here, in spite of the large size of the tumor causing pedicle erosion, expansive destruction of the vertebral body and widening of the neural foramina, there were only minimal neurologic deficits. We have therefore decided to report this case, with a review of the relevant English literature emphasizing clinical presentations, plain film images and magnetic resonance image findings of giant invasive cauda equina schwannoma for early diagnosis and differential diagnosis.

Key Words: cauda equina, giant invasive schwannoma
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Schwannomas, also known as neurilemmomas, are usually common tumors arising from nerve sheath cells [1–3]. The peak incidence for spinal nerve sheath tumors is in the fourth decade. There is no sex predilection [3,4]. These tumors are most often seen in the cervical or lumbar region, and are usually found on the dorsal sensory roots [4–6]. Spinal schwannomas also arise from spinal root nerve roots, and are occasionally seen in the cauda equina [7]. Tumors of the cauda equina are relatively rare, and account for only 6% of spinal tumors [7]. The majority of histologic diagnoses have indicated schwannoma [6,7]. Most patients with giant invasive schwannomas of cauda equina present

with severe neurologic deficits independent of activity in daily life [7,8–11]. Herein, we present a case of giant invasive cauda equina schwannoma (diagnosed according to the criteria of Sridhar et al [12]) with pedicle erosion, expansive destruction of vertebral bodies and widening of the neural foramina with, but only slight neurologic deficits dependent on activities in daily life.

CASE PRESENTATION

A 53-year-old man presented with a history of intermittent weakness in the right lower limb for 2 years. There was no urinary or fecal incontinence. Neurologic examination revealed mild right lower limb weakness with decreased deep tendon reflex, but no sensory impairment. Muscle testing showed mild decreased muscle power of the right lower limb. Deep tendon reflex tests demonstrated mild decrease of the reflex of the right lower muscle and a rating of 1 for the reflex



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of right ankle jerks (a rating of 2 indicates normality). Notably, the intermittent mild weakness of right lower limbs did not affect daily activity.

A lumbar radiography showed erosion of the first, second and third pedicles of the lumbar spine; in particular, total disappearance of the second pedicle of the lumbar spine, widening of the first and third interpedicle distance in the anteroposterior view (Figure 1A) and scalloping of the second vertebral body of lumbar spine in the lateral view (Figure 1B) were observed. Under the impression of a potential space occupying lesion, magnetic resonance imaging (MRI) was performed. MRI demonstrated a huge spinal tumor, about $7.0 \times 3.5 \times 4.0$ cm in size, at the level of the first, second and third lumbar spinal canals; it also revealed erosion of the first, second and third pedicles and scalloping of the second vertebral body of the lumbar spine, as well as invasion of an extraspinous component through the interspinous foramen (Figure 2). MRI demonstrated an intradural spinal tumor (Figures 2A–D). After administration of contrast, homogeneous enhancement confirmed an intradural spinal tumor at the level of the first, second and third lumbar spinal canals (Figures 2E and 2F). Typically, nerve sheath tumors are isointense on T1-weighted images and have a typically marked high signal on T2-weighted images. Enhancement was intense and homogeneous [4,6]. According to image appearance, the preoperative

impression was an invasive giant cauda equine nerve sheath tumor [8].

This patient underwent surgery under this impression of a giant invasive cauda equina tumor at the level of first, second and third lumbar spinal canals. A complete L1–3 laminectomy was performed. The lamina was severely eroded to paper-thin margins and adherent to the dura mater. Paper-thin dura was also noted after the laminectomy. A huge well-defined, yellowish, elastic, encapsulated, and soft multilobular intradural tumor without dura invasion or destruction was seen; the tumor extended from the level of the first and second to the third lumbar spine filling the entire spinal canal through the enlarged bilateral intervertebral foramen of L1/2 and L2/3. The nerve root of the tumor's origin could not be defined during the operation. The posterior surfaces of vertebral bodies were deeply eroded, but the dura mater was intact and covered the surface of the eroded vertebral bodies. Bilateral first/second and second/third lumbar intervertebral foramen were enlarged by the huge tumor. Near total destruction of bilateral second lumbar pedicles with extraspinous invasion was noted. To prevent postoperative instability, the first and third lumbar vertebrae were fixed with transpedicle screws and rods.

The gross tumor was composed of yellowish elastic contents. The histology of the tumor specimen revealed Antoni type A characteristics with spindle cells in a

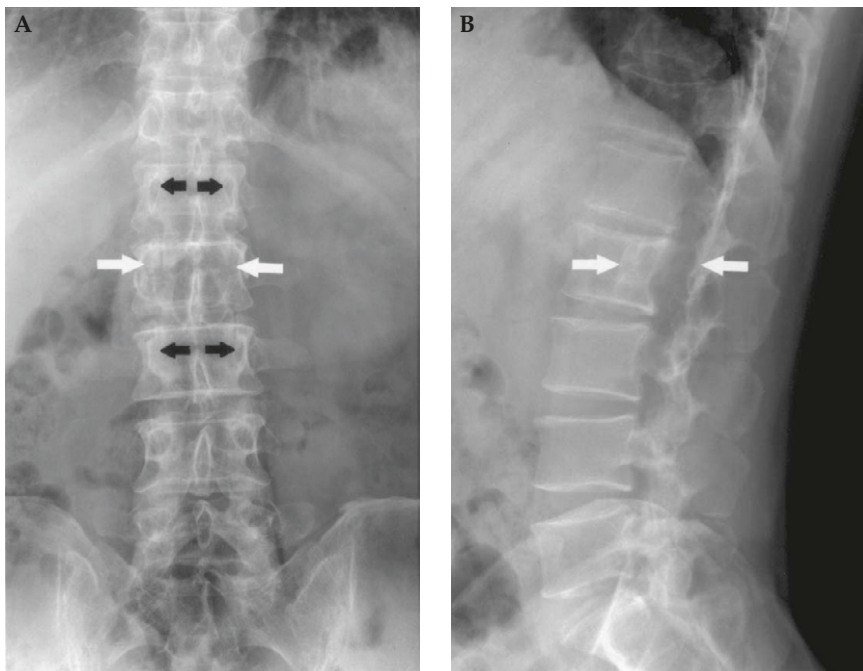


Figure 1. Lumbar radiography shows the erosion of the first, second and third pedicles of the lumbar spine, especially: (A) the total disappearance of the second pedicle of the lumbar spine (white arrows) and widening of the first and third interpedicle distances (black arrows) on anteroposterior view; and (B) scalloping of the second vertebral body of the lumbar spine on lateral view (white arrows).

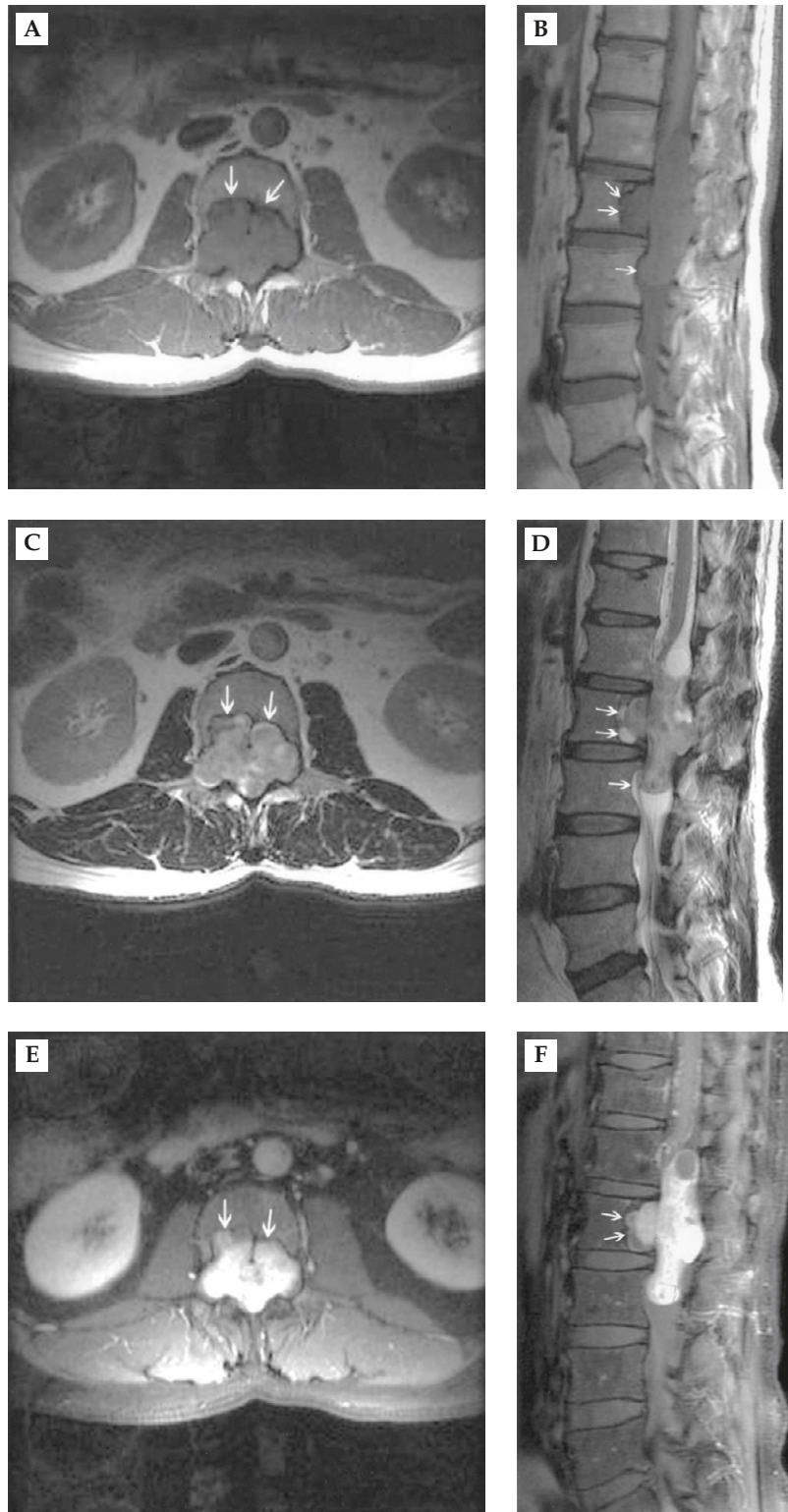


Figure 2. Magnetic resonance imaging (MRI) shows a 7.0×3.5×4.0cm intradural spinal tumor at the level of the first, second and third lumbar spinal canals with erosion of the first, second and third pedicles and scalloping of the second vertebral body of the lumbar spine. (A, B) T1-weighted MRI demonstrates isointensity in the intradural spinal tumor. (C, D) T2-weighted MRI shows hyperintensity in the intradural spinal tumor. (E, F) After administration of contrast, homogeneous enhancement indicated an intradural spinal tumor at the level of the first, second and third lumbar spinal canals.

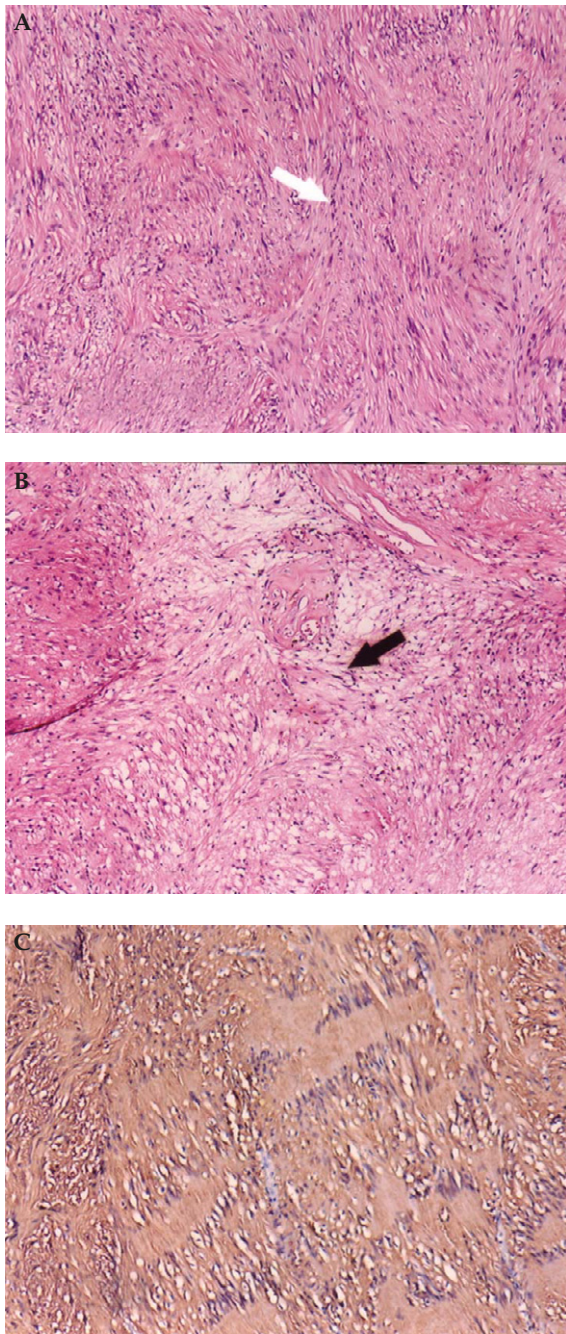


Figure 3. Histology (hematoxylin & eosin, 100×) of the tumor specimen revealed: (A) Antoni type A characteristics, with spindle cells in a palisade formation surrounded by an interstitial substance (white arrow); (B) Antoni type B characteristics, with irregular cells and a myxoid component (black arrow). (C) Immunohistochemical staining for S-100 protein is positively diffuse.

palisade formation surrounded by an interstitial substance (Figure 3A), and also Antoni type B characteristics with irregular cells and a myxoid component (Figure 3B). An immunohistochemical stain for S-100 protein was performed and demonstrated positive

staining (Figure 3C). Histology and immunohistochemical staining demonstrated a schwannoma.

The postoperative course was uneventful. The patient was immobilized for 5 days, and then mobilization was well tolerated. Local wound pain resolved within 1 week. The patient recovered full leg muscle power within 1 month of surgery without incidence of postoperative complications and had recovered without recurrence as confirmed by MRI of the lumbar spine at the 6-month follow-up.

DISCUSSION

Giant schwannomas of the cauda equina of the spinal canal, with pedicle erosion, widening of the neural foramen and severe neurologic deficits, have frequently been described. However, a giant invasive schwannoma in the cauda equina accompanied by minimal neurologic deficits, to the best of our knowledge, has not been reported previously. Sridhar et al proposed a classification system for spinal schwannomas as a means to define these giant lesions. Lesions that erode the vertebral bodies and extend posteriorly and laterally into the myofascial planes are classified as giant “invasive” spinal schwannomas [12]. Therefore, we reported this case as a giant invasive cauda equina schwannoma with minimal neurologic deficit. However, it is difficult to regard the correlation of the size of a giant invasive schwannoma to symptoms and signs as being typical in the clinical presentation in this case. We discuss the relationship between the sizes of giant schwannoma and the clinical presentation. Shimada et al revealed that cauda equina tumor size is correlated with preoperative symptom duration [13], which indicates that an enlarged size of schwannoma would increase the severity of the disease.

Most patients with cauda equina schwannomas present with neurologic deficits independent of their activity in daily life [7,8–11,13] and yet, our patient had a giant invasive schwannoma that occupied the whole spinal canal with severe bony destruction, but the clinical presentations and neurologic examinations revealed minimal deficit and impingement upon activity in daily life. Why might this be so? Firstly, schwannoma has the characteristic of gradually increasing in size [1–5]. Due to this characteristic, the mean duration of symptoms from onset to tumor diagnosis has been recognized as 22 months [13]. We did not pay

greater attention to this disease owing to the slowly progressive symptoms. Secondly, the intradural space of the lumbar spine is larger than the intradural space of the thoracic and cervical spine. The main reason for the lower deficit is likely to be more available intradural space for tumor growth. Finally, early diagnosis was confirmed before presentation of symptoms and signs had evolved. These reasons may clarify why our patient had a giant spinal tumor occupying the whole spinal canal with extensive bony destruction and extraspinal invasion, but with minimal neurologic deficits. How might we better achieve early diagnosis? Shimada et al described symmetrical lower back pain and/or pain that radiates to both lower extremities and increases in the supine position as characteristic of cauda equina tumors. If the pain has the above features, MRI examination should be performed as early as possible to check for a cauda equina tumor [13].

Most schwannomas on spinal radiography show space-occupying lesions of the spinal canal without bony destruction. Giant schwannoma of the cauda equina often results in pedicle erosion, and widening of the neural foramen without destruction of vertebral bodies [1,2,5,6]. Giant invasive schwannoma of the cauda equina often results in considerable vertebral erosion [8,12]. What explains the destruction of vertebral body, pedicle erosion and widening of the neural foramen? Schwannomas involve bone structures by three mechanisms: (1) an extrasosseous tumor causing external compression of the bone; (2) intrasosseous neurilemoma, with direct destruction of bone; and (3) a tumor arising within the nutrient canal and growing in a dumbbell-shaped configuration, resulting in enlargement of the canal [1,2,5]. In our patient, the schwannoma arising from intradural nerve sheath cells and growing in the spinal canal resulted in external compression of the vertebral body and pedicle erosion.

For management of giant schwannoma of the cauda equina, complete excision is recommended [11,12]. Nonetheless, giant invasive schwannoma in the lumbar spine region is usually excised incompletely, because complete removal has the risk of sacrificing many nerve roots [7,9,12]. Therefore, although total resection is strongly recommended, in surgical intervention for giant schwannoma of the cauda equine, spinal roots should be preserved whenever possible. Giant invasive schwannomas often create pedicle erosion and destruction of the vertebral body that may cause instability of the spine. After tumor removal,

spinal fusion with instrumentation has been performed to prevent vertebral fracture [7,8].

In summary, we present a case of giant invasive cauda equina schwannoma with extensive bony destruction but minimal neurologic deficits. It should be noted that while the presentation of particular symptoms may be the foremost indication of schwannoma, further consideration of the context and investigation with MRI might enable the identification of schwannoma prior to the appearance of severe neurologic deficits. We have reviewed previous literature and emphasized the clinical presentations, plain film images, and MR images of giant cauda equina schwannoma for early diagnosis and differential diagnosis.

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巨大侵犯性馬尾部位的神經鞘細胞腫瘤表現輕微症狀 — 病例報告以及文獻回顧

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一位 53 歲男性因右下肢輕微無力，經診斷馬尾巨大侵犯性神經鞘細胞瘤，神經鞘細胞腫瘤 (schwannoma) 通常是較常見良性腫瘤，一般是來自於感覺神經的神經鞘細胞，但巨大侵犯性神經鞘細胞瘤相當罕見，且於大部分情形會造成神經學方面缺損，進而影響日常生活，而本例之馬尾巨大侵犯性神經鞘細胞瘤雖造成週邊骨頭壓迫及破壞，但是病人只有輕微神經學的缺陷，因此我們決定報告此不尋常的病例。並回顧文獻作者也回顧以往文獻針對巨大侵犯性神經鞘細胞腫瘤的臨床表現、影像學上的發現加以討論，藉以早期診斷及鑑別診斷。

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