CASE REPORT

A huge extradural ganglioneuroma of the lumbar spine

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1. Introduction

Ganglioneuromas are benign tumors that are well circum-scribed and consist of ganglion cells and Schwann cells. They arise from the ganglion cells of the sympathetic nervous system, and are most commonly localized in the posterior mediastinum, followed by the retroperitoneum, cervical region, and adrenal gland. We encountered a rare paraspinal neurogenic tumor with extension into the spinal extradural space that presented with a dumbbell shape and bony indentation. The clinical features, radiographic analysis, and surgical approach are briefly discussed.

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2. Case Report

The case concerns a 37-year-old woman with hepatitis C who had a history of alcohol consumption. She sought...
Figure 1  A left retroperitoneal tumor, measuring 6.8 cm × 4.6 cm × 14.4 cm.

Figure 2  A heterogeneous mass (white arrows) measuring 8.0 cm × 6.0 cm × 5.5 cm, with minimal enhancement in different postcontrast phases, located in the spinal canal at the level of L1–L2 extending through the left L1–L2 neuroforamen and left psoas muscle: (A) precontrast image; (B) postcontrast image.

Figure 3  Indentation of the L1 and L2 vertebral bodies (yellow arrows) and widening of neuroforamen (white arrows).
medical advice because of impaired liver function. The routine abdominal ultrasonography revealed a left retroperitoneal tumor measuring $6.8 \text{ cm} \times 4.6 \text{ cm}$ (Fig. 1). She did not have obvious neurologic deficits, except for low back pain and occasional left buttock soreness. She was admitted to our hospital, and neurologic examination confirmed the absence of limb weakness or numbness. Computed tomography (CT) and magnetic resonance imaging (MRI) were then performed.

Figure 4  (A, B) Images are T1-weighted with contrast. (C, D) Images are T2-weighted. A dumbbell-shaped extradural tumor showing hyperintensity on T2-weighted imaging and heterogeneous postcontrast enhancement shown on L-spine magnetic resonance imaging.

Figure 5  These three images are T1-weighted with contrast and reveal that the spinal cord is displaced to the right side.
The abdominal CT showed a heterogeneous mass (8.0 cm × 6.0 cm × 5.5 cm) with minimal enhancement in different postcontrast phases, located in the spinal canal at the level of L1–L2 extending through the left L1–L2 neuroforamen and the left psoas muscle. Indentation of the L1 and L2 vertebral bodies was noticed (Figs. 2 and 3). MRI confirmed it to be a dumbbell-shaped extradural tumor, hyperintense in T2-weighted image and heterogeneous postcontrast enhancement. The spinal cord was displaced to the right (Figs. 4 and 5). Neurogenic tumors, including Schwannoma and neurofibroma, were first considered. The patient was admitted for surgical intervention.

Under general anesthesia with endotracheal intubation, the patient was placed in the prone position. The skin incision was made as shown in Fig. 6. We performed laminectomy from T12 to L2, with lateral extension of L2 in particular. The tumor was totally extradural and attached tightly to the left L2 root. We removed the tumor entirely and preserved all nerve roots. Transpedicular screw fixation from T12 to L3 was done after tumor removal (Figs. 7 and 8). The intraoperative frozen section revealed suspected neurofibroma. The specimen was grossly grayish and elastic (Fig. 9). The histopathologic examination showed a tumor with low cellularity and a loose stroma. It consisted of a cluster of ganglion cells and Schwann cells (Fig. 10). An immunohistochemical study demonstrated S-100 (+) (Fig. 11). The patient had a regular postoperative hospital stay without complications and was discharged on the 13th postoperative day.

3. Discussion

Ganglioneuromas are tumors arising from the neural crest cells. They rarely involve the spinal cord, but when this happens, they are frequently dumbbell shaped. Spinal ganglioneuromas are usually benign in nature and remain asymptomatic until they are large enough to compress the spinal cord and nerves, resulting in localized pain, radiculopathy, and weakness of the extremities.

In our literature review, only two cases of pure intradural extramedullary ganglioneuroma have been reported. Our case was a totally extradural ganglioneuroma in the lumbar spine with a dumbbell-shaped growth pattern and a widened neuroforamen. The characteristics of

Figure 6  Median-and-paramedian skin incision as shown.

Figure 7  (A, B) The tumor (🗗) covered the spinal cord. (C) Laminectomy from T12 to L2, especially lateral extension at L2, was performed. The roots, L1 (∘), L2 (∘), and L3 (∗), were all preserved.
Transpedicular screw fixation from T12 to L3 was done. We skipped the right pedicle of L2 because it was too small owing to indentation with tumor.

The specimen is grossly grayish to yellowish and elastic.

Hematoxylin and eosin stain. The background shows low cellularity and loose stroma. It consists of a cluster of ganglion cells (*), and Schwann cells, which are shown in the background with spindle shaped and round nuclei.
ganglioneuroma are bony erosion and widening of the neuroforamen; however, our case was different, in that it showed indentation of the vertebral bodies.

Because of the benign nature of ganglioneuromas, there is usually no need for medical treatment or radiotherapy for such tumors. The complete removal of a ganglioneuroma is usually curative. Paravertebral neurogenic tumors with intraspinal extension can be diagnosed and evaluated with very high accuracy using CT in adjunction with MRI. In conclusion, surgical resection of such a tumor in our case proved to be safe and feasible. Spine fixation may be necessary after removal of the tumor.

References