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CASE REPORT Open Access

Single stage complete resection of giant dumbbell lumbar ganglioneuroma: a case report and review of the literature

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Abstract

Background: Ganglioneuroma is a rare benign tumor, which is usually located in the posterior mediastinum and retroperitoneum. Occasionally, it involves the paraspinal region with intraspinal extension and becomes dumbbell shaped. Dumbbell ganglioneuromas rarely affect the lumbar spine, and only nine cases of dumbbell lumbar ganglioneuromas have been reported previously.

Case presentation: We are reporting an extremely rare case of a giant dumbbell tumor in the lumbar spine. We performed a one stage total resection of the tumor with posterior approach and long-segment fixation and fusion. This approach is different from previous methods. Histopathological examination of the surgical specimen confirmed the diagnosis of ganglioneuroma. Thirty months follow up showed a satisfactory outcome.

Conclusions: Single-stage posterior tumor resection with fixation and fusion is effective and an appropriate choice for the treatments of lumbar dumbbell ganglioneuromas.

Keywords: Ganglioneuroma, Dumbbell, Lumbar spine, Surgery

Abbreviations: CT, Computed tomography; MRI, Magnetic resonance imaging; MEPs, Motor evoked potentials

Background

Ganglioneuroma is a rare, slow-growing, benign tumor that arises from sympathetic ganglion along the spinal column and adrenal glands [1–5]. Most of ganglioneuromas are found in the posterior mediastinum and retroperitoneum, and less than 10 % in the spine [6, 7]. Ganglioneuromas may be present in any part of the spine with more prevalence in the thoracic region [6]. Occasionally, the ganglioneuromas located in the paraspinal region extended intraspinally and became dumbbell shaped [3]. Dumbbell ganglioneuromas affecting the lumbar spine are extremely rare [1, 4, 8, 9]. Until now, only nine cases of dumbbell lumbar ganglioneuromas have been reported [1, 8]. To our knowledge, this is the first report regarding the complete resection of a giant dumbbell lumbar ganglioneuroma by a single posterior approach with fixation and fusion.

Case presentation

A 29-year-old female patient was accepted in February 2013 with a 25-years history of mild weakness and numbness in both legs. She experienced worse weakness on the left side with both pain in her back and left leg. X-ray examination in 2010 revealed destruction of T12, L1 and L2 vertebral bodies. Further magnetic resonance imaging (MRI) scan revealed a large intraspinal mass extending to the left paravertebral space from T11 to L3 levels. Operation was then suggested, but the patient refused. She suffered from progressive weakness and pain in both legs. After deliberating for 3 years, the patient finally agreed to surgical excision and was transferred to our hospital. The patient underwent an operation for bilateral strephenopodia when she was 10-years old.

Upon admission, neurological examinations revealed impaired muscular strength in both legs, which was grade 5- on right side and four on the left. Pinprick sensation was decreased below the level of the groin on both sides; more severe on the left. The tendon

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reflexes were intact. She had no pyramid signs. In addition, the patient was found to have mild bladder and bowel dysfunction.

A second lumbar MRI showed a large intraspinal soft tissue mass from T11 to L3 levels. This mass extended into the paravertebral space through the neural foramina of the left L1–3 levels, and elevated the left psoas major muscle. The tumor had well-defined margins and the adjacent part of thecal sac was extremely compressed. The mass was isointense on T1 and heterogeneous hyperintense on T2 weighted images with heterogeneous enhancement on gadolinium-enhanced T1-weighted images (Fig. 1). X-ray showed a mild lumbar scoliosis with a Cobb angle of 8° from the superior endplate of L1 to the inferior endplate of L4. Computed tomography (CT) scan showed that the tumor eroded the posterior vertebral bodies and bilateral pedicles on T12, L1 and L2 levels (Fig. 2).

A posterior approach was adopted to remove the mass. Motor evoked potentials (MEPs) monitoring was applied during surgery. The patient was placed into a prone position. A longitudinal midline incision from the T9 to L5 levels was made. The bilateral paraspinal muscles were dissected subperiosteally to gain a better exposure of the laminae and left the transverse processes exposed. Laminectomy was performed from T12 to L3 to open the spinal canal followed by transversectomy and facetectomy on the left side from L1 to L3. The tumor was localized in epidural space extending to the retropleural space through the intervertebral foramen. Under the microscope, the intraspinal canal portion of the tumor was initially internally decompressed, and the adhesions between the tumor and dura were dissected carefully. The tumor was bloodless and the intraspinal part was totally resected piecemeally. The tumor was found to involve the motor and sensory roots of L1and L2 and

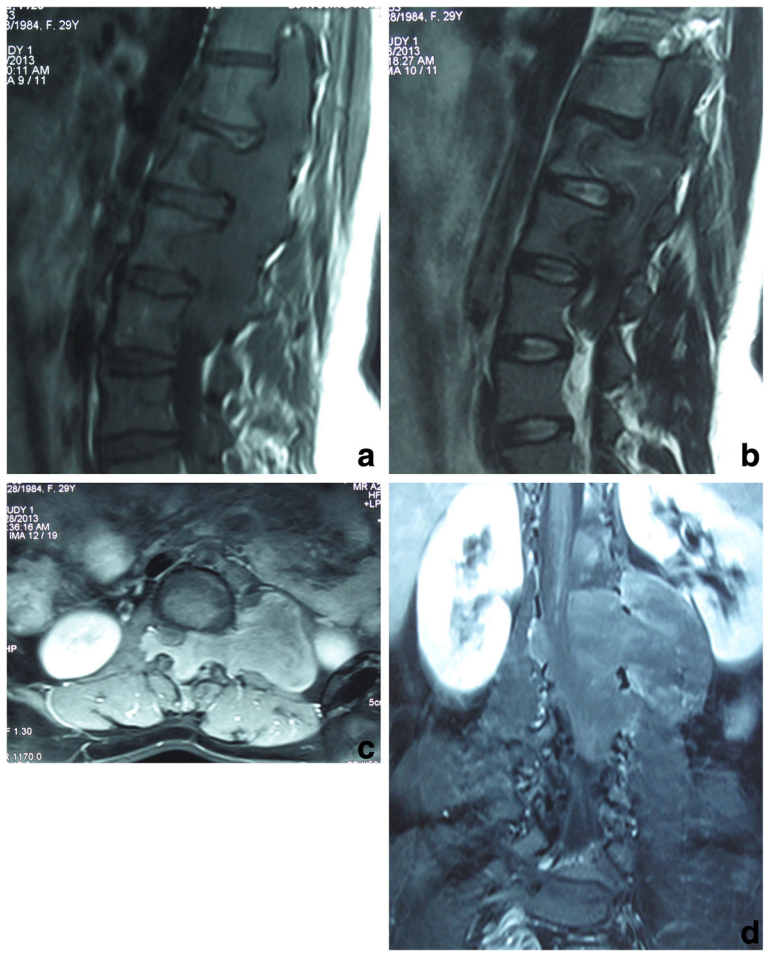

Fig. 1 Preoperative lumbar MRI scans. Sagittal MRI images show a large mass extending from T11 to L3 vertebral body levels, which is isointense on T1 (**a**) and heterogeneous hyperintense on T2 (**b**). Contrast T1 axial and coronal images (**c**, **d**) show a dumbbell-shaped component of the tumor which extends through the L1–3 foramen and lies beneath the left psoas muscle. The tumor mass displaces the thecal sac to the right and erodes the posterior vertebral body

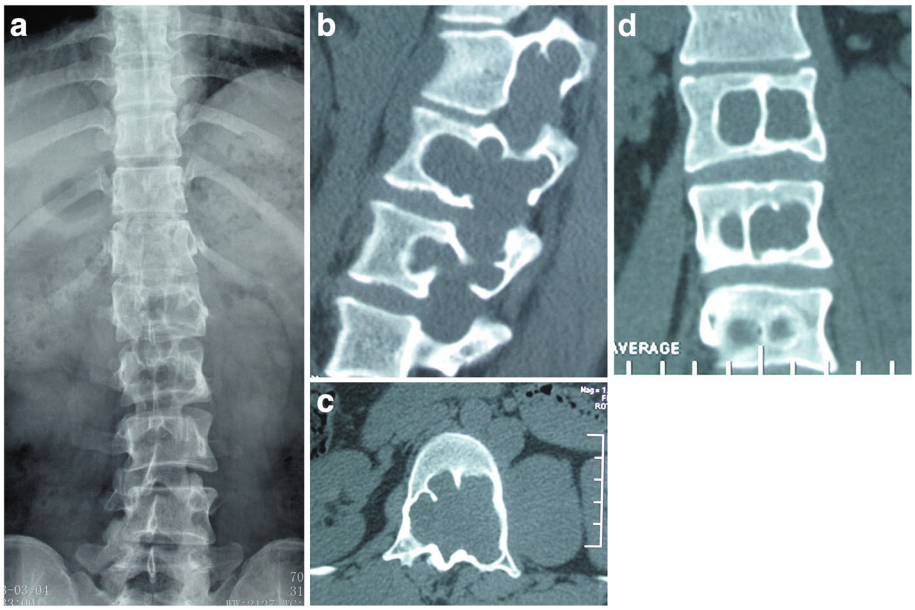

Fig. 2 Preoperative X-ray (**a**) shows a mild lumbar scoliosis with a Cobb angle of 8°. Sagittal (**b**), axial (**c**) and coronal (**d**) CT scans show that the tumor erodes extensively into the vertebral bodies, pedicles, and possible epidural space

the entire affected nerve roots were sacrificed. Then, the paravertebral tumor with its distal stump was exposed and carefully enucleated. The tumor was closely attached to the sympathetic ganglia while the adhesions were separated. Two nerves arose from the tumor, and were resected together with the tumor. Finally, the tumor was completely resected. The pedicle screws were inserted into the bilateral pedicles of T10–T12, L4 and L5. Two titanium rods were bent to curve along the contour of the spinal column and then connected longitudinally between screws. The bone graft harvested from spinous process and lamina was placed into the gutter of the resected facet joints and eroded vertebral body. A cross-link was connected and the incision was closed.

After the operation, the patient had complete resolution of pain without any aggravated sensations. She experienced slightly worsened weakness in left leg (from grade 4 to 4-). On discharge, she could walk independently.

Histopathological examination showed that the neoplasm was composed of spindle-shaped cells with elongated, thin and wavy nuclei, which were morphologically identified as Schwann cells. Moreover, the ganglion cells, which had pale to eosinophilic cytoplasm and a large vesicular round nucleus with a prominent nucleolus, were scattered or aggregated in the Schwann cell-proliferating area. No significant mitotic activity was found and MIB-1 labeling index was less than 2 %. The immunohistochemical analysis revealed that the ganglion cells were positive for S-100 and NF, while the Schwann cells stained positive for S-100, SOX-10. These results established a histological diagnosis of ganglioneuroma (Fig. 3).

During the 30 months of follow-up, motor functions in the legs recovered almost completely within 12 weeks. The patient had no clinical symptoms and maintained an extremely active lifestyle. X-ray, CT and MRI scans were performed at 1 week, 3 months, and 2 years after surgery (Fig. 4). There were no signs of tumor recurrence, no instrumentation-related complications, and spinal alignment was maintained.

Discussion

Ganglioneuroma is a rare disease with nonspecific clinical manifestations and is difficult to diagnose before operation. The differential diagnosis was mainly with other neurogenic tumors [10–13]. The patient developed symptoms when she was 4 years old. The early onset was more consistent with ganglioneuroma, as schwannoma and neurofibroma are more common in adults [10, 13]. The MRI scans of the patient showed low intensity on T1 images and heterogeneous high intensity on T2 images (Fig. 2). The features on T2-weighted images correlated with ganglioneuroma. The appearance is presumed to be caused by a combination of ganglion cells [1, 2, 14]. This patient's interverteberal foramens between L1-2 and L2-3 were widened which was consistent with ganglioneuroma, as schwannoma and neurofibroma rarely invaded more than one foramen [8, 13]. Schwannoma was a benign tumor arising from the sheath of the spinal nerve roots [15], and this tumor arose from sympathetic ganglia. The diagnosis of ganglioneuroma was confirmed by histology.

In multiple or huge tumors, complete resection at all sites is very difficult. Thus subtotal or partial resection

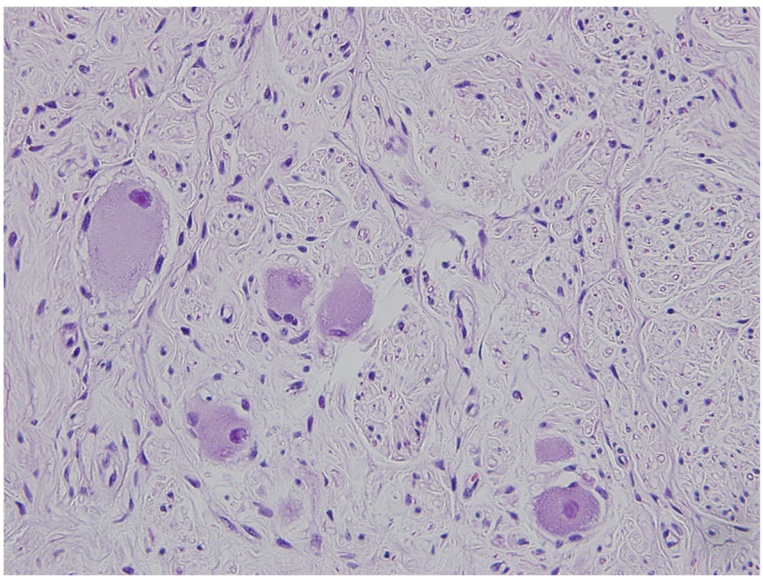

Fig. 3 Photomicrograph of the tumor. The tumor consists of large ganglion cells with interlacing bundles of spindle cells, which is consistent with ganglioneuroma

can be an acceptable choice [2, 3]. The purpose of the surgery is to decompress the spinal cord because the paraspinal part of the tumor is usually asymptomatic. Although subtotal resection has been proved to be beneficial in this situation, it is a general agreement that total resection is the best option for symptomatic solitary ganglioneuroma and the long-term prognosis is very good [1, 4, 8]. The role of radiotherapy and chemotherapy are limited to the treatment of ganglioneuroma due to their benign biological nature [4, 16].

There are various surgical approaches for managing spinal ganglioneuromas [1, 2, 4, 8, 17]. A combined posterior-posterolateral approach was used to remove the intradural and extradural parts of tumor extending through the foramina [8]. Pang et al. described a combined anterior and posterior approach in entirely removing a lumbosacral ganglioneuroma. The technique was used to treat a 36-year old female patient, and was performed by the general surgical, neurosurgical and orthopaedic team [4]. The combined approaches were technically challenging, and the potential morbidity and mortality were associated with these large resections [17]. In this case, despite the large tumor size, total resection was achieved by a single posterior approach. The posterior approach provided the surgeon with a familiar orientation of all intraspinal structures and left the patient with only one wound. The tumor was localized in epidural extending to the retropleural space. The vertebral body, bilateral pedicles and facet joints were eroded by the tumor, and the bone destruction provided an exposure of part of the tumor. Further transversectomy and facetectomy greatly improved the visibility of the tumor. With the procedure of the tumor resection piecemeally, the tumor was totally exposed and finally excised completely. In conclusion, the posterior approach may be a useful method for removing spinal ganglioneuroma.

Spinal ganglioneuromas often involve the nerve roots [1–4]; sacrifice of the entire spinal nerve is usually required for a total resection. In the study of Kim [15], 15 patients had large schwannomas with extradural components, which necessitated sacrifice of the entire motor and sensory radix. It was found that 11 (76 %) of these 15 did not develop any deficits referable to the involved myotome or dermatome. Some studies indicate that risks of causing disabling neurological deficits after sacrificing these roots are small [15, 18]. Regarding ganglioneuromas, there is no correlative literature. In this case, the tumor involved the entire left L1and L2 nerves diffusely, including motor and sensory roots. It is impossible to remove the tumor without sacrificing the affected nerve roots. During the operation, no motor responses were noted with stimulation of the tumor or of the lateral roots. The entire left L1and L2 nerve roots and two involved distal nerves were resected together with tumor. After the surgery, the patient had slight deterioration of the muscle strength in left leg from grade 4 to 4- withthe decreased sensations remained unchanged. The weakness was expected due to L1 and L2 nerve roots resection. The effect of weakness was small or non-existent. She was ambulating independently at discharge and the strength of both lower limbs completely recovered within 12 weeks. It was suggested that the incorporated nerves were no longer functional at the

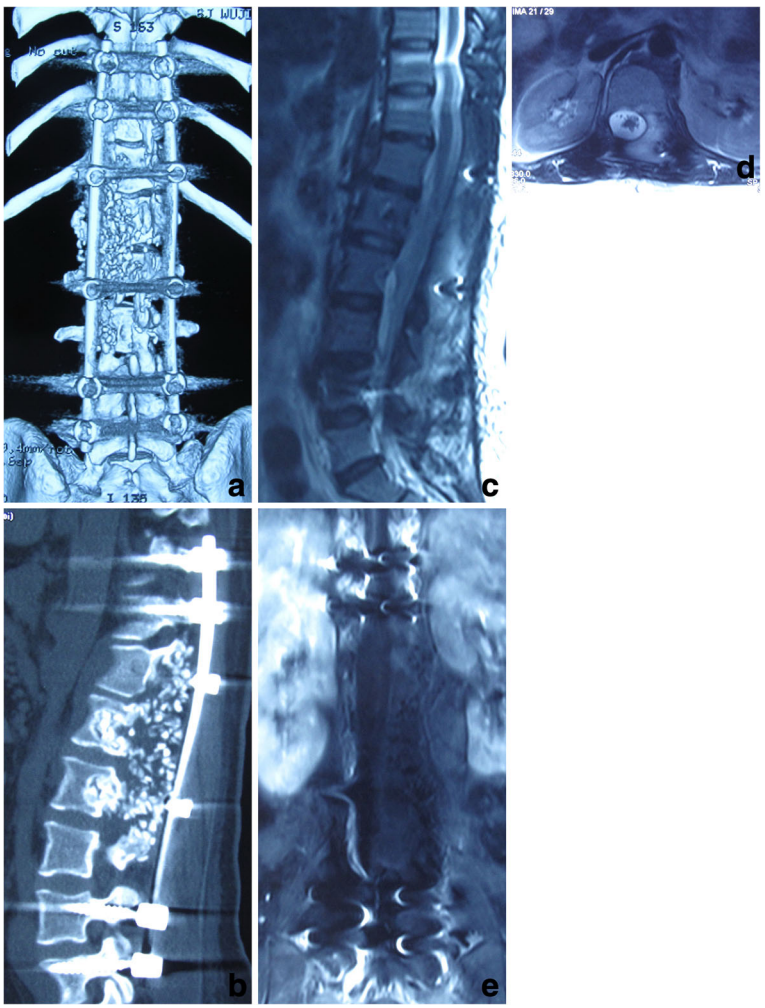

Fig. 4 Lumbar CT (**a**, **b**) and MRI scans (**c**, **d**, **e**) obtained at 12 months after surgery. There are no residual tumor, no decompression of the thecal sac, no evidences of instrumentation loosening and bone fusion. The spinal alignment is well maintained

time of surgery thus sacrificing the nerve roots would not cause disabled neurological deficit.

There are sporadic reports that ganglioneuromas may cause painless progressive spinal deformity [5, 10, 14]. The proposed mechanism for the scoliosis is the tumor expansion to the nerve root and resultant destruction of the lateral and anterior vertebral elements [5, 19]. Ganglioneuromas can grow to huge size while remaining asymptomatic and mimic the idiopathic scoliosis of adolescent. The treatment for this entity consists of complete surgical resection of the tumor and correction of the deformity; the outcome is usually highly positive [14]. In this case, the tumor involved the vertebral body, bilateral pedicles and facet joints, which leaded to spinal instability. This patient already had mild scoliosis as a result of bone destruction and pain. Although laminectomy, facetectomy and transversectomy provided a good exposure of the tumor, the spinal instability was aggravated. The fixation and fusion were necessary. Because some pedicles were eroded, the pedicle screws were applied at the levels of T10–T12, L4 and L5, and a crosslink was placed to increase the rigidity of the structure. The multisegmental posterior instrumentation and reconstruction addressed all elements of the 3-column spinal stability principle [20], and showed a favorable result. Multiple level laminectomies may cause spinal instability in the long term, however fusion or laminoplasty can enhance stability to the spine [8, 20]. In some cases of severe scoliosis or facetectomy, spinal instrumentation and fusion may be needed.

Conclusions

A rare case of giant dumbbell ganglioneuroma in the lumbar spine was presented with a tumor involved the paraspinal region with intraspinal extension. Complete resection was achieved by a posterior approach. Single-stage posterior tumor resection with fixation and fusion

may be effective and appropriate choices for the treatments of lumbar dumbbell ganglioneuromas.

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Authors' contributions
Wang ZW collected the patient data and performed the primary writing of the Manuscript. Jian FZ and Wang XW participated in the design and coordination of the study and assisted in the writing of the manuscript. Chen L collected the data of the patient and drafted the manuscript. All authors read and approved the final manuscript.

Competing interests
The authors declare that they have no competing interests, and manuscript is approved by all authors for publication.
I would like to declare on behalf of my co-authors that the work described was original research that has not been published previously, and not under consideration for publication elsewhere, in whole or in part.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Ethics approval and consent to participate
Ethical approval and consent to participate was obtained from the Ethics Committee Board of the participating hospital.

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