Thoracic Vertebral Ganglioneuroma: Treatment with 3D Printing Technology and Neurological Outcome of Spinal Cord Compression

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Abstract

Background: Ganglioneuroma (GN) is a very rare neoplasm of the central nervous system, accounting for 0.1% - 5% of all brain tumors and less than 10% of the lesions occur in the spinal cord. Because of the low incidence, there are few reports at home and abroad. It can occur in the cervical, thoracic and lumbar spines of the spine.

Method: In this case, we describe a rare GN that occurs in the thoracic 6-8 spinal canal and extends into the thoracic cavity in a 5-year-old female. Her symptoms were progressive sensory abnormalities in both lower limbs and weakness of both lower limbs for one year. The patient underwent detailed investigations including CT, MRI and 3D printing technology. Before operation, 3D model of tumors and corresponding tissue structures was printed at the ratio of 1:1 in size. As far as we know, this is the first case of spinal GN treated with 3D printing technology in the English literature.

Results: The patient had an uneventful postoperative recovery and no evidence of tumor recurrence could be demonstrated on clinical examination. Histopathological diagnosis was GN for all resected tumors.

Conclusion: In conclusion, resection of intraspinal GN occurring simultaneously in the spinal canal and thoracic cavity is effective for improving the symptoms of patients. Preoperative 3D printing based on imaging data is helpful to visually identify the location of tumors in the spinal canal and the degree of compression of spinal nerve roots before operation, and to judge the key part of compression of spinal cord by tumors during operation.

Keywords: Ganglioneuroma; 3D printing; Thoracic vertebra; Dumbbell tumor; Excision

Introduction

Ganglioneuroma (GN) (also known as ganglioneurofibroma) is a rare benign neurogenic tumor originating from the sympathetic nervous system. It can occur in the mediastinum, retro peritoneum [1-3] and spinal canal [4-7]. Intraspinal GN is rare, mostly solitary. There are also reports of multiple tumors distributed along the cervical sympathetic nerve. Individual case can develop into giant tumors [1]. Because of the low incidence, there are few reports in domestic and abroad. Accurate diagnosis often requires excision owing to the histopathological heterogeneity of these tumors and the risk of false negative biopsy results. We describe an unusual case of progressive sensory abnormality and weakness of both lower extremities caused by giant thoracic GN in a 5-year-old female. We have also briefly reviewed the pertinent literature.

Materials and Methods

Pathological analysis

In April 2017, a 5-year-old girl was hospitalized for progressive weakness of both lower limbs with severe paresthesia for 1 year duration. She also complained of intermittent numbness of both feet with occasional loss of balance on walking resulting in falls. And in the past month, incontinence of urine and stool occurred. There was no significant past medical history. On examination, she had a gibbus deformity at T7 tenderness on palpation. Detailed neurological examination revealed the upper extremities muscle force were V grade and the lower extremities muscle force were zero grade with obviously objective sensory loss to touch and pain. Moreover, proprioception and vibration were also reduced in both lower limbs. Bilateral knee and ankle jerks were hyper-reflexic.

Imaging examination revealed space-occupying lesions inside and outside the spinal canal, considering neurogenic tumors. X-ray showed elliptical space-occupying image on the left side, partial rib shadows disappeared, and the pedicle spacing of the left thoracic 6-8 was widened (Figure 1a). Three-dimensional CT reconstruction showed a homogeneous low-density mass at 6-8 level in the left thoracic spine, with a clear border. The ribs of thoracic 7 were eroded and wrapped by the mass (Figure 1b). Magnetic resonance imaging showed left side dumbbell shaped extramedullary lesion at the T6-T8 level extending outside through neural foramen, found low-signal intensity on T1-W MRI and mixed signal intensity on T2-W MRI. Both of the signals were uniformity and distinct borders. The corresponding segment of nerve root foramen was enlarged and grew dumbbell-shaped into the thoracic cavity. At the level of thoracic 7, the spinal cord was compressed by tumors and displaced to the right lower part of the spinal canal on the cross section of magnetic resonance (Figure 1c).

However, it is difficult to visualize the shape of the mass in the spinal canal and the compression boundary of the spinal cord by imaging examination. This prompted us to do detailed workup to using 3-D printing technology to print the corresponding segment solid tumors and thoracic appendages in 1:1 size of patient's body, which can objectively indicate the true condition of spinal cord compression and...
the location of tumors in the spinal canal, so that we can find the key parts of the compressed spinal cord by tumors. At the same time, we can directly observe the positional relationship between the corresponding spinal nerve roots and the tumors, reducing spinal cord and nerve root injury during resection of the tumors (Figure 2).

The decision was made for mass resection and surgical decompression after discussion with the patient and her family. Posterior decompression of T7 with hemilaminectomy followed by posterior instrumentation of pedicle screw fixation system from T5 to T9 was performed (Figure 3). In addition to thorough decompression of the thoracic cord and excision of the expanded posterior elements, transpedicular curettage of the vertebral body was done to ensure maximal removal of tumour.

Histopathology of the tumors excised during operation confirmed the diagnosis of spinal GN. The gross observation showed that the tumors were grey-white, about 0.2-0.3 cm long and 0.1 cm in diameter. Microscopically, the tumors showed spindle cell proliferation. Immunohistochemistry showed a large number of neurospecific enolase (NSE) produced in pathological section (Figure 4).

Considering the large size of intrathoracic tumors and excessive bleeding during operation, combined with the patient's wishes, extraspinal resection of intrathoracic tumors should not be performed for the time being. Cardiothoracic surgery should be performed to treat residual intrathoracic tumors half a year later. The patient was followed up for 1, 6, 12 and 22 months after operation, and no recurrence symptoms were found.

Figure 1: (a) X-ray showed elliptical space-occupying image on the left side. (b) CT showed a homogeneous low-density mass at 6-8 level in the left thoracic spine, with a clear border. The corresponding segment of nerve root foramen was enlarged and grew dumbbell-shaped into the thoracic cavity. (c) MR showed the spinal cord was compressed by tumors and displaced to the right lower part of the spinal canal.

Figure 2: (a) The black arrow showed the area after laminectomy according to the range of the blue square in b. (b) The blue square showed the key parts of the compressed spinal cord by tumors.
Discussion

GN is predominant in children and adolescents [8]. There is no obvious difference in incidence between men and women [9]. The tumors originated from sympathetic ganglion cells, mostly located in the sympathetic nerve chains on both sides of the spine and adrenal medulla [10], but rare in central nervous system, especially in the spinal cord [8,9,11,12]. The reported incidence of GN is 1 per million populations [13]. Since the lumbosacral ganglion can be located in the spinal canal, lumbosacral GN is relatively common [14].

The onset of the disease is insidious. The main clinical manifestations are pain, fixed location and progressive aggravation. Intraspinal GN is a small early lesion that causes sensory abnormalities in the innervation area without motor disturbance until the spinal cord is compressed and stimulated. With the further development of tumors, the growth of tumors in the spinal canal and compression of the adjacent spinal cord may lead to compression of the spinal cord conduction tract, resulting in impairment of the upper and lower spinal cord conduction tract function, resulting in motor and sensory dysfunction below the level of the tumors, and spinal cord incision syndrome may occur in severe cases. When the growth of tumors exceeds the compression of the surrounding tissues by the spinal canal, the clinical symptoms and corresponding signs of extra medullary space occupancy are presented. The final diagnosis depends on the pathological examination after operation.

The capsule of GN is intact, and its imaging appearance is irregular, oval, with more clear boundary. A few GN can be accompanied by calcification [15]. X-ray films showed enlargement of corresponding
nerve root foramen, compression depression of vertebral body, marginal sclerosis, widening of pedicle spacing. Vertebral canal angiography showed extramedullary space occupancy, mostly incomplete obstruction, with brush-like obstruction. CT plain scan showed homogeneous low density mass with clear margin, enlarged nerve root foramen, dumbbell-shaped outward growth, contralateral displacement of spinal cord under compression, enlarged distal subarachnoid space under compression. Generally, it cannot be enhanced or only can be slightly linear enhancement after enhanced CT scan [16]. On MRI, gangliocytoma usually behaves like all other central nervous system tumours, with low-signal intensity on T1-W, high-signal intensity on T2-W and moderately heterogeneous enhancement [12,17,18]. On T2WI, curved or linear low-signal foci were seen in high-signal tumors. These low-signal tissues represent crisscrossed Schwann cells and collagen fibers. Lonergan and co-authors considered this low signal focus to be a characteristic feature of GN [19].

Intraspinal GN grew slowly, lobulated or nodular, hard and well-defined, with a capsule and a gray-red or gray-white section. It consists mainly of nerve fibers, Schwann cells, mature ganglion cells and mucus matrix [20-23]. Light microscopy of spinal GN showed that ganglion cells were dense and uneven, often clustered and disordered [24]. Glial cells are composed of various components, mostly fibrous or hair cell astrocytes, sometimes accompanied by cell astrocytes, occasionally oligodendrocytes. Immunohistochemistry showed that positive markers were polyclonal and monoclonal antibodies to neurofibroma, neurospecific enolase (NSE), nerve growth factor receptor (TrkA), myelin basic protein (MBP), and S-100 [5,25,26]. Immunohistochemical reactions revealed strong marking by synaptophysin in ganglion cells and intense labeling by glial fibrillary acidic protein in axonal processes, Schwann cells, and satellite cells. Therefore, some scholars believe that the immunophenotypic characteristics reveal a diagnosis of GN [22,27]. GN with lipomatous component reported as a distinct entity in the literature recently shows mature adipose tissue within a GN [28,29].

In this case, 3D printing technique combined with imaging examination was used to find the key parts of the vertebral body compressed by tumors before treatment, and then hemilaminectomy was performed. Tumor resection need not be carried out urgently, because the abundant blood vessels of the tumors themselves are easy to bleed and difficult to stop bleeding, so posterior internal fixator devices should be performed before tumor resection. Only when the tumors are removed and the anesthesiologist is in close contact with the patient's vital signs, and timely blood transfusion can ensure the patient's life safety. There are also some problems encountered during the operation. The biggest problem is the separation of the tumors from the nerve roots and spinal cord when the adhesion is obvious. It is very difficult to completely remove the tumors with severe adhesion, then; the only way was a little peeling and biting off. In this treatment, we found that the decompression of the lamina and removal of the key compressive spinal cord tumors were the most important. Because the tumors in the spinal canal are provoked by resection of tumors, the tumors in the extra vertebral body can increase or even compress the important organs around them. In addition, in order to reduce the recurrence and malignancy of tumors, cardiothoracic surgeons should be invited to resect the tumors in the thoracic cavity as soon as possible after the patient's condition is stable.

Conclusion

In conclusion, surgery is an effective method for the treatment of intraspinal GN. Preoperative application of 3D printing combined with imaging examination to evaluate the location of tumors is helpful to guide the operation. Preoperative blood preparation and internal fixation of vertebral body before resection of tumors are extremely important. Of course, treatment of the huge dumbbell-shaped tumors inside and outside the spinal canal depends on the cooperation of many disciplines to achieve satisfactory results.

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References


