CASE REPORT

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Adolescent thoracic scoliosis due to giant ganglioneuroma: a two-case report and literature review

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ABSTRACT

Ganglioneuromas are rare benign tumors that arise from the sympathetic nervous system. The presentation of tumors is variable and associated with adolescent thoracic scoliosis. Herein, we present two case reports and a review of literature. The two cases involved 10 and 13-year-old patients who were asymptomatic for muscle pain or weakness, and ganglioneuromas were incidentally detected through imaging screening. Both patients underwent a two-stage surgery. The first stage involved detachment of the tumor from the spinal cord and simultaneously performing deformity correction surgery from the posterior aspect. The second stage was resection of the ganglioneuroma through the anterior approach without neurological problems. A two-stage surgery was necessary to excise the tumor and correct the deformity, thereby avoiding neurological problems and concurrently establish a pathological diagnosis. Commencing with the posterior approach proved to be safe and was more effective.

Keywords: ganglioneuroma, adolescent thoracic scoliosis, two-stage surgery, corrective fusion, tumor resection

Abbreviations:
GN: ganglioneuroma
MRI: magnetic resonance imaging

INTRODUCTION

Ganglioneuroma (GN) is a rare benign neurogenic neoplasm composed of ganglion and mature Schwann cells and nerve fibers. GNs are typically found in the posterior mediastinum or retroperitoneum of asymptomatic patients.[1] Although GN is a benign tumor, it constitutes 0.72–1.6% of primary retroperitoneal tumors, which can grow to an enormous size and pose

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serious consequences. In young individuals, it may cause spinal scoliosis.[2] In this study, we discussed two cases of thoracic scoliosis caused by giant GN, as well as the clinical characteristics of this tumor.

CASE REPORT

Case 1

A 10-year-old girl was referred to our hospital for thoracic scoliosis. Radiography revealed a neoplastic lesion in the thorax. She had no symptoms or neurological findings, except for rapid progression of scoliosis. Whole spine radiography showed a 71-degree scoliosis deformity at T7–T11 (Fig. 1A). Chest radiograph showed a huge shadow in the right thoracic cavity (Fig. 1B). Magnetic resonance imaging (MRI) revealed a soft tissue mass located on the convex apex side of the vertebral body and the tumor measured 9.0 x 7.5 x 3.0 cm, showing T1W hypodense, T2W heterogeneous hyperdense, and mild heterogeneous enhancement effect (Fig. 2A–C). Moreover, the tumor developed into spinal canal via intervertebral foramen of T8/9 (Fig. 2D). Computed tomography (CT) scan revealed bone proliferation at the corner of vertebral body of T9 (apex) (Fig. 2E). The patient subsequently underwent a needle biopsy to diagnose the pathology, revealing GN on thoracic scoliosis. A two-stage surgery was then planned. The first surgery was performed using a posterior approach to ensure a relationship between the tumor and the spinal cord. Laminectomy was performed to release the tumor from the spinal cord, followed by T2–L1 posterior correction and fusion combined with T9 pedicle subtraction osteotomy. Four months later, a second surgery was performed to resect the tumor using a lateral direct thoracic approach.

Pathological findings revealed CD56, S-100, neurofilament, and synaptophysin-positive spindle cells, indicating GN. No progression of scoliosis was observed at 8 years postsurgery (Fig. 1C), and no tumor recurrence was observed on imaging.

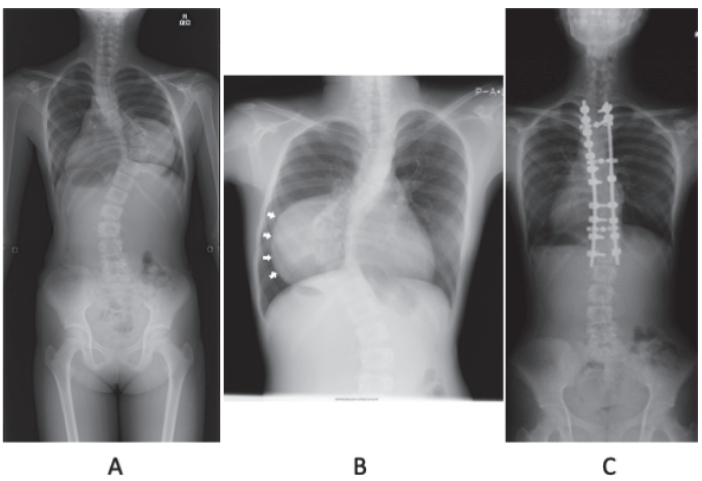

Fig. 1 Radiographic evaluation of spinal surgery outcome (Case 1)
Fig. 1A: Whole-spine posterior-anterior radiograph showed a 71-degree scoliosis deformity at T7-T11.
Fig. 1B: Chest radiograph anterior-posterior showed a huge shadow in the right thoracic cavity.
Fig. 1C: Whole-spine posterior-anterior radiograph after surgery 8 years.

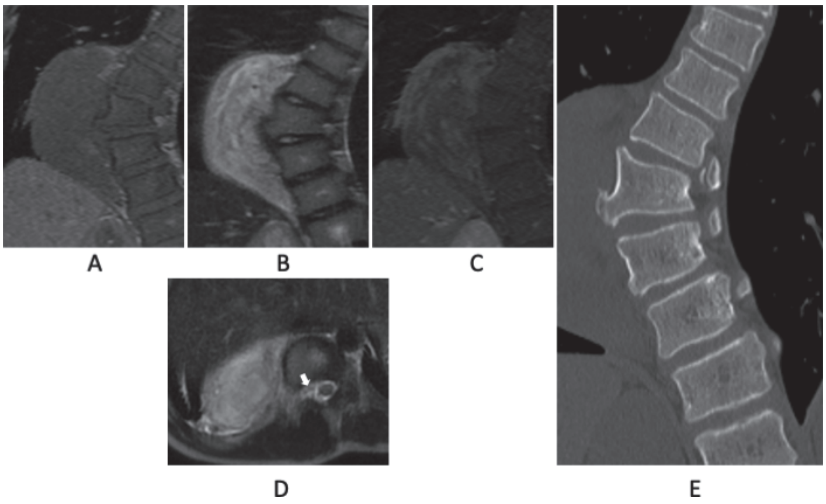

Fig. 2 Multimodal imaging evaluation of tumor characteristics, spinal involvement (Case 1)
Fig. 2A: MRI revealed a soft tissue mass located on the convex apex side of the vertebral body and the tumor measured 9.0x7.5x3.0 cm, showing hypodense on T1W.
Fig. 2B: MRI in T2W heterogeneous hyperdense on coronal view.
Fig. 2C: MRI in T1W with Gadolinium on coronal view with mild heterogeneous enhancement effect.
Fig. 2D: MRI in T2W revealed that tumor developed into spinal canal via intervertebral foramen of T8/9 on axial view (white arrow).
Fig. 2E: CT scan revealed bone proliferation at the corner of vertebral body of T9.
MRI: magnetic resonance imaging
CT: computed tomography

Case 2

A 13-year-old boy was diagnosed with a thoracic spine tumor at the age of 6 years after underwent open-chest surgery at age 6 due to pectus excavatum (there was no biopsy at that time). He had been under the supervision of a pediatric surgeon; however, at 13 years of age, he was referred to our department because of scoliosis progression. The patient did not report back pain or neurological abnormalities. Radiography indicated a 69 degrees of scoliosis at T7–T11 (Fig. 3A), and chest radiograph showed a large shadow overlapping the thoracic spine (Fig. 3B). MRI revealed a soft tissue mass measuring 9.0 x 6.6 x 2.4 cm located on the apex of scoliosis. The tumor exhibited T1W isodense, T2W hyperdense, and a homogeneous contrast effect on the right convex side of T6/7–T10/11, with T9/10 right foraminal extension (Fig. 4A), intracanal tumor development along the vertebral arch with no tumor behind the lamina (Fig. 4B). The patient was diagnosed with tumor-related scoliosis and underwent a two-stage surgery. In the first stage, part of the tumor was resected at the T9/10 right foramen using a posterior approach to avoid traction of the spinal cord. Subsequently, T2–L1 posterior corrective fusion was completed. The second-stage involved tumor resection through a lateral direct thoracic approach, which was performed 4 months later. Pathological findings revealed S-100, neurofilaments, and synaptophysin-positive spindle cells; however, tumor necrosis was not observed, which resulted in GN. Postoperatively, no progression of scoliosis or recurrence of the tumor was observed over a period of 4 years and 8 months (Fig. 3C) besides normal symptoms and MRI images after surgery.

Both patients provided informed consent for the submission of their data and images for publication.

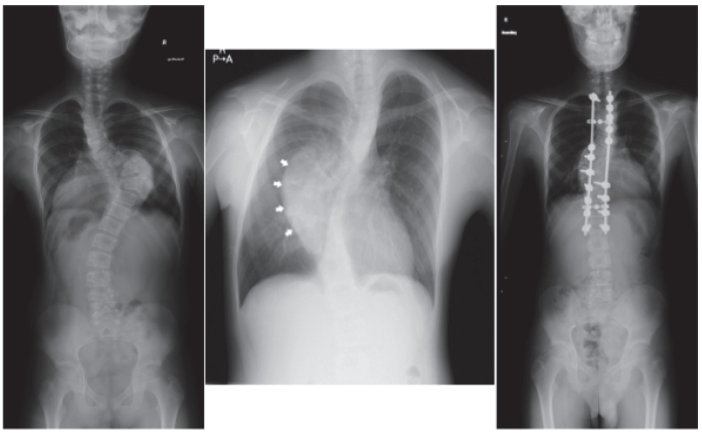

Fig. 3 Long-term radiographic follow-up of spinal surgery outcome (Case 2)
Fig. 3A: Whole-spine posterior-anterior radiograph showed 69-degree of scoliosis at T7-T11.
Fig. 3B: Chest radiograph anterior-posterior showed a large shadow overlapping the thoracic spine.
Fig. 3C: Whole-spine posterior-anterior radiograph after surgery 4 years 8 months.

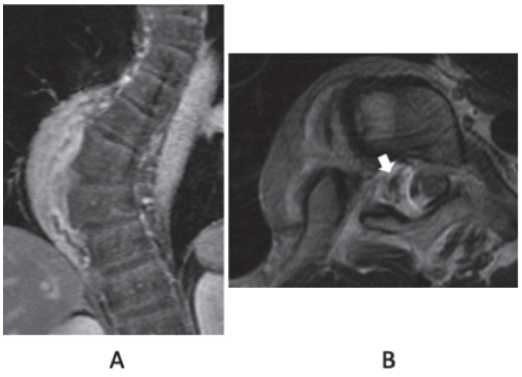

Fig. 4 MRI evaluation of tumor characteristics (Case 2)
Fig. 4A: MRI revealed a soft tissue mass located on the convex apex side of the vertebral body and the tumor measured 9.0x6.6x2.4 cm, showing hyperdense on T2W.
Fig. 4B: MRI in T2W revealed that tumor developed into spinal canal via right intervertebral foramen of T9/10 on axial view (white arrow).
MRI: magnetic resonance imaging

DISCUSSION

GN is a rare benign tumor composed of ganglion cells and Schwannian stroma.[3] The prevalence of GN is estimated to be 0.1–0.5% of all central nervous system tumors,[4] with the typical age at diagnosis being approximately 7 years.[5] A slight female predominance was noted,[3] ranging from 1.13:1 to 1.5:1. The posterior mediastinum (41.5%), retroperitoneum (37.5%), adrenal gland (21%), and neck (8%) are the most frequently affected areas. In our study, the tumor mass was located in the posterior mediastinum.

A literature review on GN with scoliosis is presented in Table 1.

Table 1 Literature review of ganglioneuroma presenting with scoliosis

No.	Case	Age (years)/Sex	Scoliosis location	Curve/ Cobb	Presentation	Treatment	Outcome
1	Bauer et al,[6] (1989)	16/F	L1-L4	Right/20°	Scoliosis, incipient paraparesis, pain in the left leg	TR	2 years, NR
2	Sampson et al,[7] (1991)	12/F	T4-T7	Right/-	Painless thoracic scoliosis	Open biopsy	No data
3	Xuhua et al,[8] (2004)	11/F	T8-L1	Right/80°	Scoliosis	TR	NR
4	Velyvis et al,[9] (2005)	15/F	T2-T7	Left/36°	Mild but persistent back pain	Ant. excision and post. decompression	RS, NR
5	Lai et al,[10] (2005)	12/F	T8-T11	Right/95° Left/60°	Painless thoracic scoliosis	Two-stage excision and fusion	Partial scoliosis correction, NR
6	Spiegel et al,[11] (2006)	14/F	T5-T7	Right/-	Asymptomatic	Two-stage excision and fusion	RS, NR
7	Qiu et al,[5] (2007)	9/M	T9-L1	Left/-	Asymptomatic	Two-stage excision and fusion	RS, NR
8	Qiu et al,[5] (2007)	14/F	T3-T12	Right/105°	Mild persistent back pain	Two-stage excision and fusion	RS, NR minor loss of correction
10	Kara et al,[12] (2013)	28/M	T1-T9	Left/109°, Right/97°	Dyspnea and vomiting	TR	Recurrence
11	D'Eufemia et al,[13] (2014)	11/F	T4-T11	Left/18° Right/10°	Abdominal pain, nausea, vomiting, constipation	Post. excision	RS, NR
12	Yilmaz et al,[14] (2015)	10/F	T11-L5	Left/<40°	Lower-back pain, intermittent sensation in the left leg	Post. excision	Symptom persisted
13	Demir et al,[15] (2015)	33/M	T6-T11	Right/-	Scoliosis	TR	No data
14	Yang et al,[4] (2016)	12/F	T10-L4	Right/33.7°	Claudication, right abdominal mass, progressive deformity	Two-stage excision and fusion	RS, NR
15	Ulusoy et al,[16] (2016)	7/F	T12-L2	Left/-	Left hypoesthesia at T12, L1, L2 dermatomes	Follow up	Scoliosis.
16	Barrena López et al,[17] (2018)	40/M	L5-S1	Right/-	Right lower limb pain, weakness, abnormal gait	Post. excision	11 × 6 mm^2 residual tumor, pain
17	Wang et al,[18] (2018)	21/F	L1-L2	Left/-	Asymptomatic	Excision	RS, NR stable scoliosis
18	Takebayashi et al,[19] (2019)	33/M	L1-L2	Left/-	Urinary retention left lower limb numbness	Two-stage extradural and intradural excision	RS
19	Elnady et al,[20] (2020)	17/F	T5-T9	Right/50°	Back pain, bilateral lower limb numbness, progressive deformity	Single-stage post excision and fusion	NR
20	Zhang et al,[21] (2020)	35/F	T4-T7	Right/-	Asymptomatic	TR	NR
21	Gaddipati et al,[22] (2021)	13/F	L3-L4	Left/15°	Mild lumbar back pain	One-stage excision of the lesion and fusion	NR

Ant.: anterior
F: female
L: lumbar
M: male
NR: no recurrence (months follow-up)
Post.: posterior
RS: resolution of symtoms
T: thoracic
TR: totally resected

Many patients have no symptoms; therefore, the tumor can slowly develop through the spinal foramina, adopt a dumbbell shape, and cause neurological symptoms due to nerve root and/or spinal cord compression.[9] Diagnosis may be delayed because of the absence of symptoms. In Case 1, the patient was diagnosed with idiopathic scoliosis, and the previous physician was unaware of the presence of the tumors.

GN is sufficiently soft to encircle nearby blood vessels or expand into a lacuna. Occasionally, CT or MRI can show foot processes and a dumbbell shape.[23] Among the MRI findings, the "whorled sign" is recognized as typical of GN. The tumor exhibits this indication as high intensity on T2-weighted imaging, with some low-signal and nodular features. Although, whorled sign is not seen in our report, this sign corresponds to Schwann cells and collagen fibers in the tumor[24,25] and should be noted when surveying imaging.

In our study, the entire mass was located on the convex side and adjacent to the apex of the curve, similar to other reports. One possible explanation is that the tumor stimulated the spinal epiphyseal plate on the affected side, causing it to overgrow.[26] Moreover, the paravertebral muscle of the convex side was affected by the tumor, which caused muscle atrophy,[10] the mechanism underlying this scoliosis is not yet fully understood. Compression or invasion of nearby nerve roots or chronic compression of nerves and muscle structures can lead to gradual weakness and atrophy of the affected muscles. In our two cases, no muscle atrophy was noted, however this is also a symptom to be aware of to avoid missing GN clinically.

Surgical approaches for the treatment of scoliosis with GN remain controversial, with various experts advocating for different methods. Wang et al performed tumor excision without addressing scoliosis, showing scoliosis without improvement.[18] In contrast, Lai et al advocated two strategies for a case of severe scoliosis with paravertebral GN: anterior resection of the tumor and release of the scoliosis, followed by posterior spinal corrective fusion.[10] In 9 of the 21 cases we reviewed, only tumor removal was performed, and in 7 cases, tumor removal was combined with scoliosis correction. Huang et al offered two distinct approaches advocating for tumor resection from both the posterior and anterior aspects.[27] Zhang's approach revolves around a one-stage surgery performed posteriorly to anteriorly.[21] Conversely, Qiu et al presented two stages: one involving anterior tumor resection and release and the other focusing on scoliosis correction.[5] Bauer's approach encompasses both posterior tumor resection and correction, as well as anterior tumor resection.[6] The decision to perform corrective scoliosis surgery is based on a number of factors, including the severity of the curvature, the progression of the curve, the presence of symptoms, and the likelihood of more severe complications. In cases of GN combined with severe scoliosis, surgery is required to correct the scoliosis and resect the tumor. In six of the eight patients who underwent both tumor removal and scoliosis corrective fusion, a planned two-stage surgery was performed. Notably, certain reports have raised concerns about neurological complications resulting from tumor development within the spinal canal.[14,19] In this report, our operation is divided into two stages combined with intraoperative neuromonitoring to avoid possible neurological complications. In the first-stage operation, a posterior approach and laminectomy were performed with resection of the intracanal tumor and correction of scoliosis. In the second-stage operation, gross tumor resection was performed. Our method has significant advantages in terms of neurological safety. Severe scoliosis can dramatically alter the anatomy of the spine, making traditional approaches more difficult due to disfiguring anatomy. A direct lateral approach may provide a more direct path to the tumor without having to move around the spinal deformity site. The anterior approach may require more manipulation with this device, which may increase the risk of complications. In our two cases, the tumor is located in a more lateral position in the chest cavity, a direct lateral approach can provide better access to the affected area while minimizing manipulation on the spine. This is particularly pertinent, as

previous reports have highlighted the risk of neurological deficits stemming from the traction of tumors within the spinal canal during anterior surgery.[6] Surgery for this disease requires different approaches in different positions. Therefore, we divided the surgery into two stages to reduce invasiveness. The time between the two stages of surgery ranges from 1 to 7 weeks[4]; however, the interval time should be considered based on the patient's recovery condition after the first operation. If they recover well and show no signs of surgical contraindications, the timing for subsequent procedures can be determined accordingly. In the two cases, the second stage was performed after 4 months, coinciding with the patient's summer vacation, which was convenient for the patient and the family.

CONCLUSION

Management of GN coinciding with scoliosis varies. We report successful results following a two-stage excision and corrective fusion of a giant thoracic GN accompanied by scoliosis. Scoliosis can be safely corrected by resecting a tumor in the spinal canal during the first stage of surgery. This is a useful method to reduce the invasiveness of surgery by dividing it into two stages.

AUTHOR CONTRIBUTIONS

YY: substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work. Drafting the work. Final approval of the version to be published. TH, GY, TB, HA, SO, KI, TY, KK, and YM: substantial contributions to the acquisition, analysis, or interpretation of data for the work. Revising it critically for important intellectual content. Final approval of the version to be published. DH: substantial contributions to the conception or design of the work. Revising it critically for important intellectual content. Final approval of the version to be published. YM: substantial contributions to the conception or design of the work. Revising it critically for important intellectual content. Final approval of the version to be published.

CONFLICTS OF INTEREST

The authors have no conflicts of interest to disclose.

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