Retroperitoneal ganglioneuroma combined with scoliosis
A case report and literature review

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Abstract

Rationale: Ganglioneuroma (GN) is a rare tumour arising from the sympathetic nervous system. GN is constantly asymptomatic, easily ignored and likely damages other organs during tumour progression.

Patient concerns: The case report involved a 21-year-old girl who was admitted to a hospital because of a computed tomography result after her pregnancy examination showed retroperitoneal tumour and scoliosis. The scoliosis was considered as a tumour complication.

Diagnoses: The tumour was finally diagnosed as GN by pathological examination.

Interventions: We carried out surgical treatment and performed a pathological examination on postoperative tumour specimens.

Outcomes: The patient was followed up for 19 months and did not show tumour recurrence. However, the condition of the scoliosis did not improve.

Lessons: This paper reports a case of GN with scoliosis at the same time. GN is a benign tumour consisting of cells with a special origin. GN grows extensively and leads to different complications. Presently, pathological examination after an operation is the only approach to formulate an exact diagnosis. We should consider the possibility of retroperitoneal tumour, especially GN, if a patient suffers from scoliosis with an unknown cause. Thus, CT and MRI are needed to provide additional information that would help formulate a diagnosis.

Abbreviations: AIS = adolescent idiopathic scoliosis, CT = computed tomography, GN = ganglioneuroma, GNB = ganglioneuroblastoma, MRI = magnetic resonance imaging, T2WI = T2 weighted imaging.

Keywords: ganglioneuroma, retroperitoneal tumor, scoliosis

1. Introduction

Ganglioneuroma (GN) is a rare benign tumor occurring spontaneously. It can also occur during radiation therapy or chemotherapy,[11] accounting for 0.72% to 1.6% of primary retroperitoneal tumors.[12] This tumor is commonly found in the posterior mediastinum and retroperitoneum.[3] Patients suffering from GN do not usually present symptoms because of its nonfunctioning trait. However, several complications arise if the tumor is sufficiently large to press against the adjacent organs. In this report, we introduce a patient suffering from GN and its complications.

1.1. Case description

The patient was a 21-year-old Chinese woman diagnosed with a tumor behind the left peritoneum on computed tomography (CT) during her 33rd-week pregnancy examination in July 2016. The tumor remained clinically silent. All physical examination results were normal except for lumbar scoliosis. The patient did not have problems in daily activities nor experienced neurological symptoms. Her family members were in good condition.

The patient underwent abdominal enhanced CT after the cesarean operation in September 2016. The CT results were as follows. The scoliosis was located at the lumbar segment (Fig. 1). Several soft tissues suspected to be tumors were found at the apex of the scoliosis. Tissues in front of the left psoas major muscle were enhanced irregularly in the image and adhered to the aorta ventrals and wrapped around the left accessory renal artery. The maximum cross-sectional area of the tissues was 4.7 cm × 4.7 cm (Figs. 2A, B and 3).

We considered the tissues as a type of neoplastic disease that can lead to scoliosis, but the nature of the tumor remained unknown. Whether the tumor grows continually was also difficult to confirm. Several tumor markers, such as alpha-fetoprotein and carcinoembryonic antigen, were at normal levels.

Finally, the patient underwent resection of the retroperitoneal tumor after all physical examinations and routine laboratory
studies showed no contraindication. We selected left lateral rectus incision. During the surgical operation, 3 parts of the tumor (approximately 6 cm × 7 cm × 6 cm, 3 cm × 3 cm × 3 cm, and 3 cm × 3 cm × 4 cm, respectively) were connected by fibers. Parts of the tumor were hard and lobulated in shape but did not extend to the spinal canal. The tumor was exceedingly close to the main vessels and psoas, and it was difficult to separate them. Nevertheless, the nearby lymph nodes were in their normal form. The masses and fibers were excised entirely during the operation, whereas the normal tissues including the lymph nodes were preserved and all specimens were diagnosed as GN after pathological examination. The pathologists finally provided the following immunohistochemistry report: S100 (nerve fiber +), Syna (ganglion cell +), NeuN (−), and Ki67 (MIB-1) (−). No adverse and unanticipated events occurred during the treatments. The patient was followed up for 19 months and did not show tumor recurrence. However, the condition of the scoliosis did not improve.

1.2. Literature review

We considered the scoliosis as a complication of GN in this case. However, cases that combine GN and scoliosis are scarce. Thus, the relationship between the 2 diseases is difficult to determine exactly. We attempted to find evidence for this relationship by using keywords “GN” and “scoliosis” to search on the Web of Science. We obtained 13 articles with full texts, in which 14 cases offered detailed characteristics, including sex, age, curve pattern, size, major symptoms, and other factors under consideration (Table 1).[^4-15] The amount of clinical data was insufficient for a meta-analysis. Thus, we only observed several similarities of GN from the above cases to obtain experience in judging, diagnosing, and treating.

2. Discussion

GN, as well as neuroblastoma and ganglioneuroblastoma (GNB), is a peripheral neuroblastic tumor. Among these tumors, GN is the most mature and consists of ganglion cells, nerve fibers, and Schwann cells. GN is recognized as a benign tumor usually diagnosed in older children and in adults.[17] Low recurrence rate and good prognosis are observed after the complete removal of this tumor. However, GNB is the most immature and malignant of the tumors above and occurs in younger patients mostly and may develop into GN.[17] During the development process, some neuroblastic cells, including neuroblasts, differentiating neuroblasts, and/or ganglion cells in GNB, enter various stages of maturation.[18]

Previous articles indicated similar GN morbidity between males and females.[19] However, the number of females suffering concurrently from GN and scoliosis is higher than that of males from the cases we studied. In our work, children and adolescents more easily acquired scoliosis than did the other age groups. Therefore, we supposed that etiology may be related to factors, such as sex and age. However, our evidence is insufficient to indicate the details.
GN stems from the sympathetic chain and can be located in the cervical region to the pelvic cavity. However, most of GN tumors are located in the posterior mediastinum and retroperitoneum. Most retroperitoneal GNs are nonfunctional and do not often exhibit special symptoms. Patients may be admitted initially to the hospital because of disease complications.

Our patient has never been in a hospital before the retroperitoneal tumor and scoliosis were found. As revealed by the cases we studied, most patients with GN were admitted to the hospital because of painless and progressive scoliosis (Table 1). Moreover, all of the tumors were located at the convex side of the curve. In our study of previous cases, left and right curves exhibit similar occurrence rates (Fig. 4). Several patients presented scoliosis with 2 curves. This result may be caused by irregular tumor location, but the exact mechanism remains unknown. In our operation, we found fibers between each part of the tumor. Thus, fibers might exist between the tumor and spine, which might pull the spine to the tumor. Moreover, a compulsive position caused by stimulation to tissues close to the tumor may be one cause. Several professors propose that tumors stimulate the epiphyseal plate and hence cause osteoepiphysis hyperplasia.

Table 1

| Authors     | Time | Year | SEX | Age (y) | Site | Size   | Curve/ cobb | Extend to the spinal canal | Major symptoms                                                                 | Surgery | Recurrence |
|-------------|------|------|-----|---------|------|--------|-------------|-----------------------------|---------------------------------|---------|------------|
| Bauer BL   | 1989 | Female | 16  | L1–L4   | 23×15 cm² | Right/20° | T11, L5     | Scoliosis, incipient paraparesis, pain in the left leg | Yes     | No         |
| Sampson MA  | 1991 | Female | 12  | T4–T7   | —     | Right/- | —           | —                           | Painless thoracic scoliosis     | No      |            |
| Xihua L    | 2004 | Female | 11  | T8–L1   | 5×5×4.1 cm³ | Right/80° | —           | —                           | Scoliosis                       | Yes     | No         |
| Velyvis JH  | 2005 | Female | 15  | T2–T7   | 8×8×2 cm³  | Left/36°  | T5          | Mild but persistent back pain | Yes     | No         |
| Lai PL     | 2005 | Female | 12  | T5–T11 (right), T12–L4 (left) | 10×9×7 cm³ | Right/95° | T9–T10      | Painless thoracic scoliosis | Yes     | No         |
| Qiu Y      | 2007 | Male   | 9   | T9–L1   | 10×9×7 cm³ | Left/-    | —           | —                           | Rapid progression of scoliosis | Yes     | No         |
| Qiu Y      | 2007 | Female | 9   | T3–T12  | 13.5×9×4 cm³, 11×7×4 cm³ | Right/105° | T11        | Mild but persistent back pain | Yes     | No         |
| Qiuzu L    | 2009 | Female | 41  | Postmediastinum | 25×10 cm³ | Left/-    | —           | Thoracic scoliosis, chest distress and pain | Yes     | No         |
| Kara T     | 2013 | Male   | 28  | T1–T7 (left), T8–L2 (right) | —     | —       | —           | Dyspnea and vomiting             | —       | —          |
| D’Eufemia R | 2014 | Female | 11  | T8–T9 (left) | 4.5×9.5 cm³ | Left/18°  | —           | Abdominal pain, nausea, vomiting, constipation | Yes     | No         |
| Yilmaz S   | 2015 | Female | 10  | L1–L3 (right), T11–L5 | —     | Right/10° | Left/ < 40° | Lumbar spine | Lower-back pain, intermittent tingling sensation in the left leg | Yes     | —          |
| Damir MK   | 2015 | Male   | 33  | T6–T11  | —     | Right/- | —           | —                           | Hypoesthesia of the T12, L1, and L2 vertebral levels | No      | —          |
| Ulusoy OL  | 2016 | Female | 7   | T12–L2  | —     | Right/- | —           | —                           | Right abdominal mass with mild pain | No      | —          |
| Yang YH    | 2016 | Female | 12  | T12–L2  | 13×8×6 cm³ | Right/33.7° | —           | —                           | —       | —          |

L = lumbar vertebra, T = thoracic vertebra, — = not mentioned.
Our literature review showed that some patients experienced back pain potentially derived from spinal deformity. Some patients complained of vomiting and abdominal pain that may be caused by tumors compressing the stomach or other digestive organs. Moreover, compression of the diaphragm muscle may explain why some patients exhibited dyspnoea. Commonly, clinical symptoms caused by paravertebral tumor mostly depend on the range of the spine close to the mass. Two patients suffered from pain and tingling sensations in the legs, which might be caused by spinal cord disease. By imaging examinations, we observed that the tumor extended into the canals of the spine and possibly affected the spinal nerves. Similarly, other neurologic diseases, such as changes in gait and weakness of the muscles and senses, can occur if the mass compresses the spinal cord. Hypertension and diarrhoea are observed if the tumor secretes catecholamine and vasoactive intestinal peptide.

GN is symptomless and lacks specific diagnostic methods. Thus, certain examinations, such as imageology, play an important role in diagnosing this disease. GN is sufficiently soft to encircle nearby blood vessels or expand into a lacuna. Thus, the shapes of GN can vary. Occasionally, foot processes and a dumbbell shape can be observed in CT or magnetic resonance imaging (MRI). However, appearances depend on the contents of the tumor. If the main part of GN is mucus substrate, then CT imaging will resemble that of a cyst. Meanwhile, T2-weighted imaging (T2WI) shows a high signal in MRI. If ganglion cells and fibers are the major structures, then CT imaging will show an increased density, and T2WI will reveal a slightly high signal. For enhanced CT examination, we can observe slightly intensified parts, which fit our report. A previous study suggested that a low-signal part called the swirl sign can be found in MRI when the mass is shown in a high signal; this sign is the characteristic appearance of GN. However, our patient in this case did not undergo MRI; thus, we failed to diagnose GN through the swirl sign. Moreover, GN should be distinguished from Schwannoma and cystic teratoma in imaging examinations.

In general, surgery is the only potential curative treatment to retroperitoneal tumors. Tumor resection should be performed as soon as the tumor is diagnosed. Given the large size and special location of the tumor, a definite diagnosis is difficult to formulate before clinical auxiliary testing with sufficient experiments for diagnosis. Thus, the nature of the mass can be confirmed after pathologic examination. Patients with severe scoliosis are suggested to undergo a second operation on the spine. Treatment should be individualised to fit each patient. Given that the patient in this report was asymptomatic in neuropahty, she opted out of spine treatment at that moment. Treatment outcome depends on the patient’s condition. A large Cobb angle extends the recovery time of scoliosis.

3. Conclusion

GN is a benign tumor consisting of cells with a special origin. GN grows extensively and leads to different complications. Presently, pathological examination after an operation is the only approach to formulate an exact diagnosis. We should consider the possibility of retroperitoneal tumor, especially GN, if a patient suffers from scoliosis with an unknown cause. Thus, CT and MRI are needed to provide additional information that would help formulate a diagnosis.

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