

**CASE REPORT**

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***Secondary Scoliosis from a Retroperitoneal Ganglioneuroma in a 9-year-old Female: Case Report***

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**ABSTRACT**

Ganglioneuromas are rare benign tumors that tend to be asymptomatic until large enough to compress adjacent structures. These tumors are often detected during childhood evaluations for other pathologies. Retroperitoneal ganglioneuromas may be a secondary cause of scoliosis. Here, we present a case of a child who was found to have a retroperitoneal ganglioneuroma during an evaluation for scoliosis.

**KEYWORDS**

Ganglioneuroma, Scoliosis, Retroperitoneal mass

**INTRODUCTION**

Ganglioneuromas (GNs) are rare neuroblastic tumors of neural crest origin, and these tumors arise from either the sympathetic chain or the adrenal medulla.<sup>1</sup> GNs are considered benign tumors and have favorable outcomes when treated with resection alone.<sup>2</sup> Survival rates are comparable between partial resection and complete resection for cases when the latter is not possible.<sup>3-5</sup> GNs tend to be asymptomatic until they are large enough to compress adjacent structures. These tumors are often detected incidentally in childhood during evaluations for other pathologies.<sup>6,7</sup> Retroperitoneal GNs have been theorized to be associated with the development of scoliosis; however, reported cases that combine GN with scoliosis are scarce.<sup>8</sup> Here, we present a case of a young female with a retroperitoneal GN that was found incidentally during an evaluation for scoliosis.

**CASE REPORT**

The patient was a 9-year-old female with a history of scoliosis and upper respiratory infections who presented to the clinic with right hip pain. She under-

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went magnetic resonance imaging for evaluation of her scoliosis, and a large retroperitoneal mass was incidentally discovered. The mass was noted to be 9.9 x 6.4 x 5.7 cm and was found within the right psoas muscle. It appeared to emanate from the L2 foramen. The Cobb angle was noted to be 13 degrees as shown in Figure 1 and Figure 2.

The patient was taken to the operating room on June 3, 2019, for mass excision via a retroperitoneal approach with both pediatric and neurosurgery physicians. The case proceeded without complication. Pathology was sent to NIH/NCI, and the reports indicated that the mass was a ganglioneuroma, immature. The patient's hospital stay was uncomplicated, and she was discharged on postoperative day two. Due to the mild degree of scoliosis, a spinal correction was deemed unnecessary.

**DISCUSSION**

GNs are mature, differentiated peripheral neuroblastic tumors derived from neural crest cells. They can develop anywhere along the sympathetic nervous system and are commonly found in the posterior me-



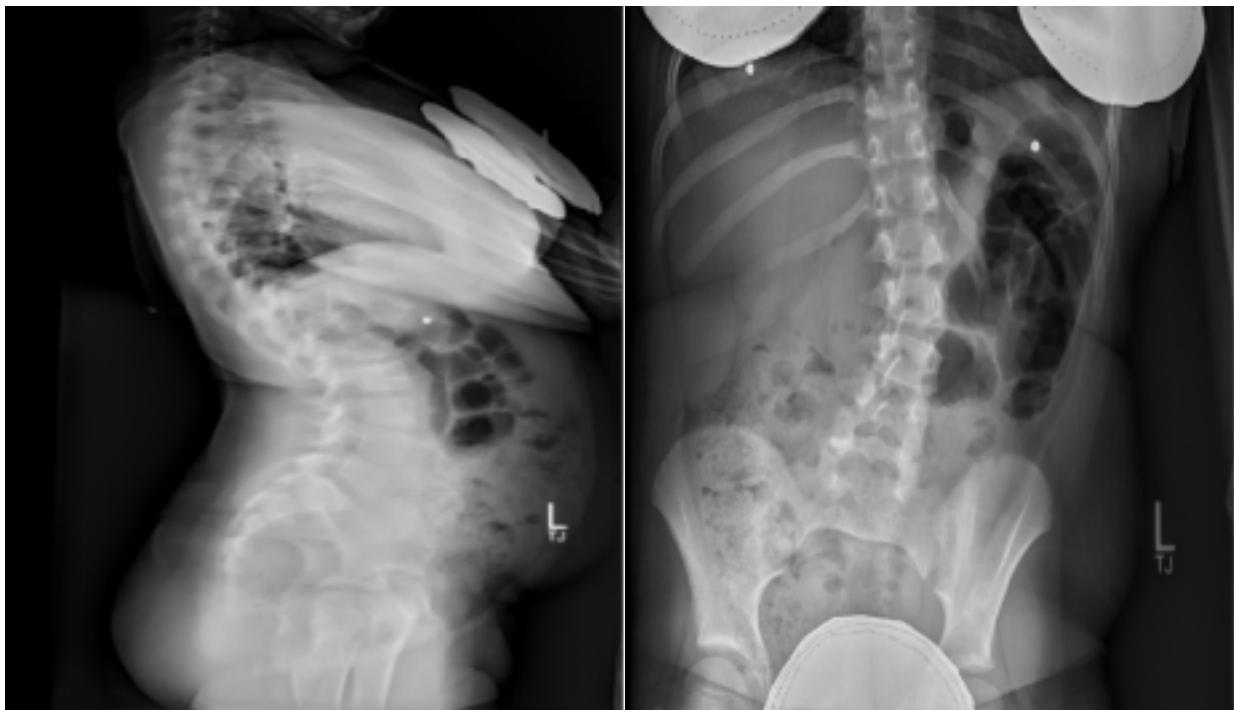


FIGURE 1. Abdominal radiographs.

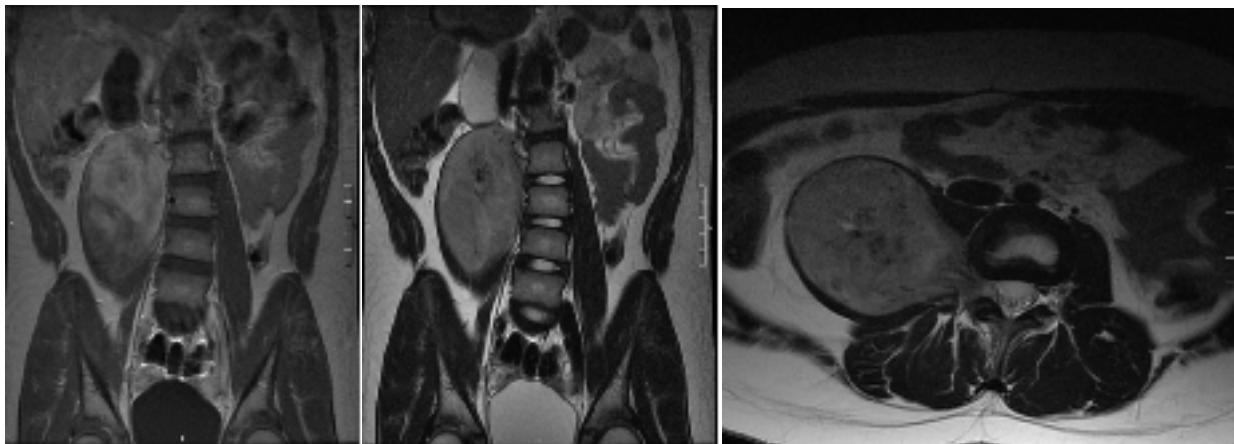


FIGURE 2. MRI of the spine.

diastinum or retroperitoneum.<sup>4</sup> These tumors exhibit slow growth, yet they tend to reach large dimensions since they typically remain clinically silent until they produce symptoms from the compression of locoregional structures.<sup>1</sup> Additionally, most will remain asymptomatic and will be diagnosed incidentally during imaging for other evaluations.

Cases that combine GN with scoliosis are rare. Previous studies reported that females are more likely than males to have concomitant scoliosis and GN. Children and adolescents are more likely to develop scoliosis

in association with GN than those of other age-groups.<sup>9</sup> Therefore, there is a potential susceptibility for younger females to develop scoliosis with GN, but the data are too scarce to be conclusive.

The theoretical mechanism of a tumor leading to scoliosis has been attributed to the stimulation of the epiphyseal plate, causing osteoepiphysis hyperplasia.<sup>8</sup> Yang et al. proposed three mechanisms by which a paravertebral ganglioneuroma causes scoliosis: (1) expansive tumor growth leading to damages in the lateral and anterior aspects of the vertebrae;



(2) scoliosis mechanically stimulated by the tumor; and (3) simultaneous occurrence of paravertebral GN.<sup>10</sup> Wang et al. published a recent case report and literature review indicating that all GNs associated with scoliosis were found on the convex side of the spine.<sup>9</sup> Notably, in our case, the mass was on the concave side of the spine, possibly suggesting more of a pure mass effect than the above-stated mechanisms.

Imaging studies are a vital adjunct to the diagnosis of GN due to its clinical silence. In the pediatric population, ultrasonography offers a useful initial test if there is any suspicion of a lesion.<sup>11</sup> However, MRI offers better characterization and is typically included in the work-up for retroperitoneal masses.<sup>12,13</sup> Ultimately, there is a need for tissue biopsy to definitively diagnose GN. Tissue diagnosis is typically obtained at the time of surgical resection.

Whenever possible, a complete resection of a GN should be accomplished. If there is a concern for the possibility of serious complications due to complete resection, then partial resection is an acceptable alternative.<sup>14</sup> Depending on the degree of scoliosis, a two-stage procedure may be undertaken. The first procedure will correct the deformity of the spine, and the second procedure will resect the tumor.<sup>9</sup> In our case, a single procedure to completely resect the mass was deemed appropriate, due to the mild degree of scoliosis.

## CONCLUSION

GNs are benign tumors that grow slowly but extensively, leading to various complications from mass effects. While imaging is a useful adjunct for diagnosis and operative planning, a definitive diagnosis requires tissue sampling. Resection of a GN is considered curative with a favorable prognosis. While primary (idiopathic) scoliosis is more common in younger age groups, secondary causes, including retroperitoneal masses such as GNs, should be ruled out.

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