MARSHALL JOURNAL OF MEDICINE
Expanding Knowledge to Improve Rural Health.

Volume 6 | Issue 2 Manuscript 1254

2020

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

Andrew J. Weaver, Keitaro Nakamoto, and Daniel A. Beals

Author Affiliations

Andrew J. Weaver (*Marshall University Joan C. Edwards School of Medicine, Huntington, West Virginia*)

Keitaro Nakamoto (*Marshall University Joan C. Edwards School of Medicine, Huntington, West Virginia*)

Daniel A. Beals (*Marshall University Joan C. Edwards School of Medicine, Huntington, West Virginia*)

Corresponding Author

Andrew J. Weaver MD

Marshall University Joan C. Edwards School of Medicine

Huntington, West Virginia

Email: weaveran@marshall.edu

Recommended Citation

Weaver, Andrew J.; Nakamoto, Keitaro; and Beals, Daniel A. (2020) "Secondary Scoliosis from a Retroperitoneal Ganglioneuroma in a 9-year-old Female: Case Report," *Marshall Journal of Medicine*: Vol. 6: Iss. 2, Article 10.

DOI: 10.33470/2379-9536.1254

Available at: https://mds.marshall.edu/mjm/vol6/iss2/10

DOI: 10.33470/2379-9536.1254

Author Footnote: Patient Consent: The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their name and initials will not be published and due efforts will be made to conceal their identify, but anonymity cannot be guaranteed. Funding: No funding or grant Authorship: All authors attest that they meet the current ICMJE criteria for authorship. Conflict of Interest: None Author Contribution: 1. Andrew Weaver: literature search, drafting article 2. Keitario Nakamoto: drafting article 3. Daniel Beals: Manuscript concept, analysis, critical revision

Secondary scoliosis from a retroperitoneal ganglioneuroma in a 9-year-old female: a case report

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.[1] GNs are considered benign tumors and have favorable outcomes when treated with resection alone.[2] Survival rates are comparable between partial resection and complete resection for cases when the latter is not possible.[3-5] 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.[6,7] Retroperitoneal GNs have been theorized to be associated with the development of scoliosis; however, reported cases that combine GN with scoliosis are scarce.[8] 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 underwent 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.

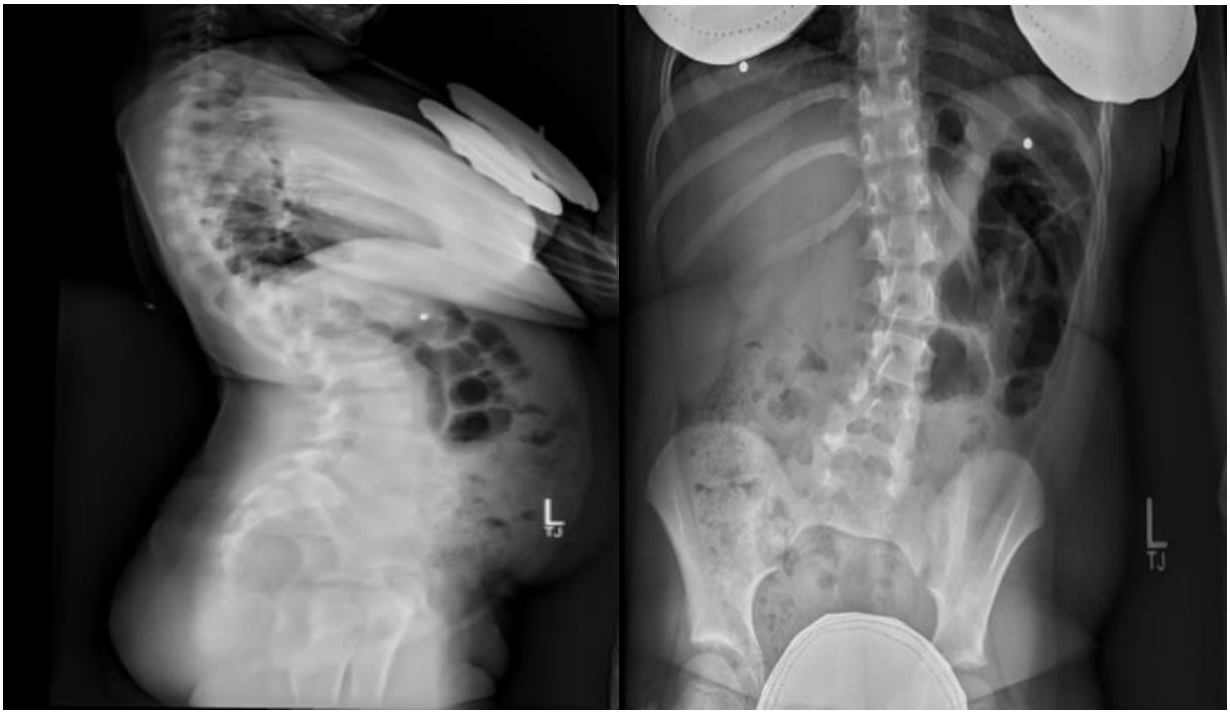

Figure 1. Abdominal radiographs.

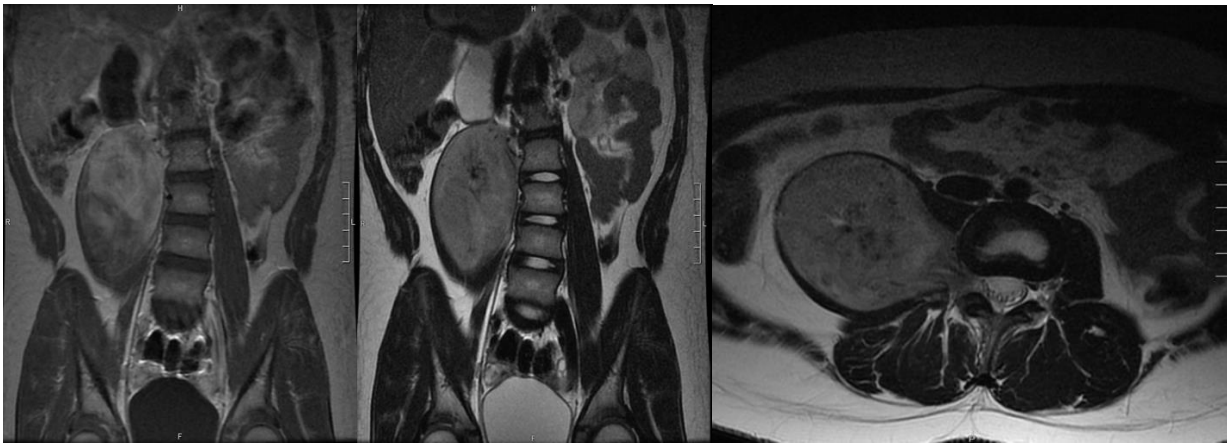

Figure 2. MRI of the spine.

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 mediastinum or retroperitoneum.[4] 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.[1] 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.[9] 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.[8] 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.[10] 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.[9] 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.[11] However, MRI offers better characterization and is typically included in the work-up for retroperitoneal masses.[12,13] 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.[14] 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.[9] 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.

References

1.Geoerger B, Hero B, Harms D, Grebe J, Scheidhauer K, Berthold F. Metabolic activity and clinical features of primary ganglioneuromas. Cancer. 2001;91(10):1905-13.
2.Decarolis B, Simon T, Krug B, Leuschner I, Vokuhl C, Kaatsch P, et al. Treatment and outcome of ganglioneuroma and ganglioneuroblastoma intermixed. BMC Cancer. 2016;16:542.
3.De Bernardi B, Gambini C, Haupt R, Granata C, Rizzo A, Conte M, et al. Retrospective study of childhood ganglioneuroma. J Clin Oncol. 2008;26(10):1710-6.
4.Alexander N, Sullivan K, Shaikh F, Irwin MS. Characteristics and management of ganglioneuroma and ganglioneuroblastoma-intermixed in children and adolescents. Pediatr Blood Cancer. 2018;65(5):e26964.
5.Paasch C, Harder A, Gatzky EJ, Ghadamgahi E, Spuler A, Siegel R. Retroperitoneal paravertebral ganglioneuroma: a multidisciplinary approach facilitates less radical surgery. World J Surg Oncol. 2016;14(1):194.
6.Schulman H, Laufer L, Barki Y, Philip M, Mares AJ, Maor E, et al. Ganglioneuroma: an 'incidentaloma' of childhood. Eur Radiol. 1998;8(4):582-4.
7.Zugor V, Schott GE, Kuhn R, Labanaris AP. Retroperitoneal ganglioneuroma in childhood--a presentation of two cases. Pediatr Neonatol. 2009;50(4):173-6.
8.Qiu Y, Wang S, Wang B, Zhu F. Adolescent thoracolumbar scoliosis secondary to ganglioneuroma: a two case report. Spine (Phila Pa 1976). 2007;32(10):E326-9.
9.Wang X, Yang L, Shi M, Liu X, Liu Y, Wang J. Retroperitoneal ganglioneuroma combined with scoliosis: a case report and literature review. Medicine (Baltimore). 2018;97(37):e12328.
10.Yang Y, Ren M, Yuan Z, Li K, Zhang Z, Zhang J, et al. Thoracolumbar paravertebral giant ganglioneuroma and scoliosis: a case report and literature review. World J Surg Oncol. 2016;14:65.
11.Mut DT, Orhan Soylemez UP, Demir M, Tanik C, Ozer A. Diagnostic imaging findings of pelvic retroperitoneal ganglioneuroma in a child: a case report with the emphasis on initial ultrasound findings. Med Ultrason. 2016;18(1):120-2.
12.Silveira CRS, Vieira CGM, Pereira BM, Lopes EJ, Gerson G, Tavora DGF, et al. Magnetic resonance neurography in the diagnosis of a retroperitoneal ganglioneuroma: case report and literature review. Radiol Case Rep. 2018;13(2):380-5.
13.D'Eufemia P, Properzi E, Palombaro M, Lodato V, Mellino L, Tetti M, et al. Scoliosis secondary to ganglioneuroma: a case report and up to date literature review. J Pediatr Orthop B. 2014;23(4):322-7.
14.Sanchez-Galan A, Barrena S, Vilanova-Sanchez A, Martin SH, Lopez-Fernandez S, Garcia P, et al. Ganglioneuroma: to operate or not to operate. Eur J Pediatr Surg. 2014;24(1):25-30.