



Paraspinal Ganglioneuroma in an Adolescent with Painless Scoliosis: Case Report and Literature Review

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ABSTRACT

Adolescent scoliosis secondary to ganglioneuroma is a rare entity with a clinical presentation mimicking adolescent idiopathic scoliosis that can lead to delayed or misdiagnosis. We present this case with a literature review to illustrate the importance of careful evaluation of the clinical and radiological findings in reaching a proper, timely mannered diagnosis. Clinical and radiological evaluation with a keen eye for signs of the atypical curve and radiological abnormalities on plain films and getting more detailed imaging with CT and MRI would lead to earlier diagnosis of secondary causes of scoliosis and direct proper treatment plan.

Keywords: Paraspinal ganglioneuroma, Scoliosis, Radiological findings, Diagnosis

INTRODUCTION

Although most common scoliosis cases are idiopathic painless conditions other possible causes of scoliosis are congenital [1], neuromuscular [2], degenerative [3], syndromic [4], adolescent idiopathic [5] and infection should be excluded by proper clinical and radiological evaluation especially in presence of pain or neurological complaint [6]. Here we present a rare case of thoracolumbar scoliosis secondary to ganglioneuromatumor. Ganglioneuroma is a rare benign tumor derived from the primordial neural crest. Ganglioneuroma [7] arise from ganglion cells of the posterior mediastinum, retroperitoneum, cervical spine, and adrenal glands. Most commonly reported to be found in the posterior mediastinum and retroperitoneum but it can be found anywhere along with the sympathetic nervous tissue.

CASE HISTORY

A 15-year-old female presented with a complaint of painless back deformity. She had no history of pain, limitation of activity or any neurological symptoms. Unremarkable systemic review. No past medical illness and no family history of a similar condition. Upon examination: no dysmorphic features, positive Adam's test, no tenderness to palpation of the back, the normal neurological exam for power, sensation, and reflexes. Plain scoliosis films showed the right thoracolumbar curve with the apex at T11 with a compensatory lumbar curve.

There was widening of intercostal space between right 11th-12th ribs, right 11th rib head dislocation in addition to the elevation of right hemidiaphragm (Figure 1). Upon these findings, further imaging with CT and MRI was done and it showed large right paraspinal mass extending from the level of T7-T8 to lower T12 vertebral bodies. It is extending to and widening T10-T11, T11-T12 and to a lesser extent T9-T10 right intervertebral foramina. There is a focal posterior extension to back muscles through intercostal space, with space widening. The Dysmorphic appearance of the right eleventh rib is seen. The lesion shows low T1 and heterogeneous intermediate T2 signal intensity with fat signal intensity interspersed within the lesion. It shows heterogeneous enhancement. Significant mass effect on the right hemidiaphragm, right kidney, and liver (Figure 2). Histopathology diagnosis with core biopsy came as ganglioneuroma. The patient is planned for two-stage treatment with tumor resection followed by scoliosis surgery.

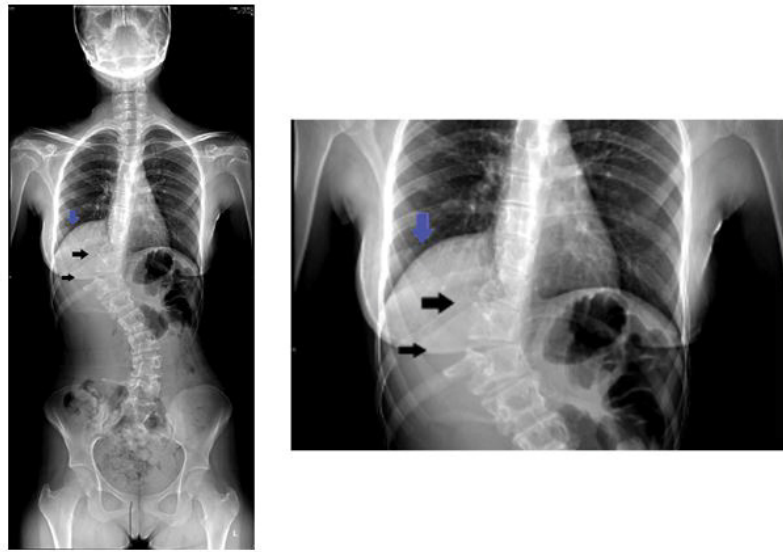


Figure 1 Right thoracolumbar curve with the apex at T11 with a compensatory lumbar curve. There is widening of intercostal space between right 11th-12th ribs (small black arrow), right 11th rib head dislocation (large black arrow) in addition to the elevation of right hemidiaphragm (blue arrow)

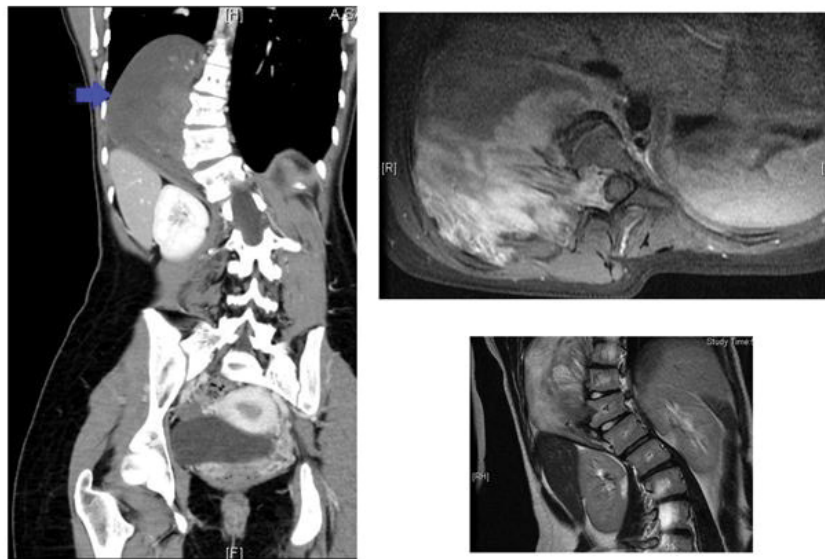


Figure 2 Large right paraspinal mass extending from the level of T7-T8 to lower T12 vertebral bodies. It is extending to and widening T10-T11, T11-T12 and to lesser extent T9-T10 right intervertebral foramina. There is the right lateral epidural extension at the same levels. There is focal posterior extension to back muscles through intercostal space, with space widening. Adjacent osseous scalloping and reactive changes are seen involving adjacent vertebral bodies and ribs. The Dysmorphic appearance of the right eleventh rib is seen. The lesion shows low T1 and heterogenous intermediate T2 signal intensity with fat signal intensity interspersed within the lesion. It shows heterogeneous enhancement. There is a significant mass effect on the right diaphragm, liver and right kidney

DISCUSSION

Ganglioneuroma is a rare benign tumor arising from the neural crest cells. The reported incidence of GN is one per million population. The reported mean age for diagnosis is approximately 7 years with slight female predominance ranging from 1.13:1 to 1.5:1. The most common locations are the posterior mediastinum (41.5%), retroperitoneum (37.5%), adrenal gland (21%) and neck (8%) [7-11].

The tumor is mostly asymptomatic and discovered incidentally upon evaluation for other conditions [10]. In some instances, it can cause local mass effect and patient present with symptoms related to the location and size of the mass (e.g. scoliosis [12,13], abdominal pain [14,15], shortness of breath [16], vomiting [16], Back pain [17], neurological symptoms). Hormonally active forms have been reported and it may present with hypertension diarrhea and virilization which can be explained by secretion of catecholamines, vasoactive intestinal polypeptides, or androgenic hormones by the tumor [8,18].

In the reported cases of ganglioneuroma with scoliosis deformity, the tumor is reported to be most commonly located adjacent to the apex of the deformity, on the convex side and asymptomatic [9,19].

The mechanism by which ganglioneuroma lead to scoliosis is largely unknown but it is hypothesized that: tumor mass cause destruction of the side and front vertebrae causing scoliosis [7]; scoliosis and tumor occurring simultaneously [8]; the mass stimulates the growth plates on the convex side [9]; local mass effect of the tumor cause weakness of the paraspinal muscles on the tumor side [8].

The staged surgical treatment with tumor resection followed by spinal deformity corrective surgery has recurrence rate and excellent prognosis especially in case complete tumor resection was performed with low recurrence on long term follow up [9,19].

CONCLUSION

Scoliosis secondary to ganglioneuroma can be misdiagnosed as idiopathic scoliosis due to a similar presentation as asymptomatic deformity. Clinical and radiological evaluation with a keen eye for signs of the atypical curve and radiological abnormalities on plain films and getting more detailed imaging with CT and MRI would lead to earlier diagnosis of secondary causes of scoliosis and direct proper treatment plan.

DECLARATIONS

Conflicts of Interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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