

# Asymptomatic Ganglioneuroma in Presumed Idiopathic Juvenile Scoliosis: A Case Report and Review of the Literature

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## Abstract

Idiopathic scoliosis accounts for the majority of scoliosis cases. The advent of Magnetic Resonance Imaging (MRI) gave new insights into possible causes for presumed idiopathic scoliosis, such as neural axis pathology. Although an increasing number of studies have dealt with this topic, the indications for MRI vary widely and controversy still exists. As an example of this problem, we present the case of a child with presumed idiopathic scoliosis and an otherwise normal physical examination, where routine MRI revealed a large thoracic ganglioneuroma.

## INTRODUCTION

By definition, idiopathic scoliosis is due to an unrecognizable cause and therefore is a diagnosis of exclusion. In occasional cases however, the label "idiopathic" is wrongly used when there may be a pathology underlying the scoliosis that has not been detected by appropriate investigations. Non-idiopathic causes for scoliosis are neural axis anomalies, such as tumors, Chiari malformations, syringomyelia or a tethered cord syndrome<sup>1,2,3,4,5</sup>. The association of neural axis anomalies and scoliosis is long established<sup>6,7</sup>, but was usually only considered if pathological neurological symptoms and signs were present. These abnormal clinical findings prompted further investigations, which in the past consisted of invasive procedures such as myelography and myelo-CT scans. Since Baker and Dove in 1983<sup>8</sup> reported a case of a progressive scoliosis as the presenting sign of an otherwise asymptomatic syringomyelia, a new interest in the early detection of incorrectly classified idiopathic scoliosis due to neural axis pathologies came up has been kindled. One major drawback in the investigation of the incidence for this entity was the invasive procedures needed to establish the diagnosis. In recent years MRI has become a standard diagnostic procedure, replacing myelography and myelo-CT in the investigation of neural axis conditions to a large extent. It has the advantage of being non-invasive and not exposing patients to ionising radiation and is therefore ideal as a screening investigation. Several studies

<sup>2,4,9,10,11,12,13,14,15,16,17,18</sup> have evaluated scoliosis using MRI and have shown that it is effective in detecting the presence of neural axis pathologies. We present the case of a clinically asymptomatic male child with presumed juvenile idiopathic scoliosis where the screening MRI led to the diagnosis of significant neural axis pathology.

## CASE

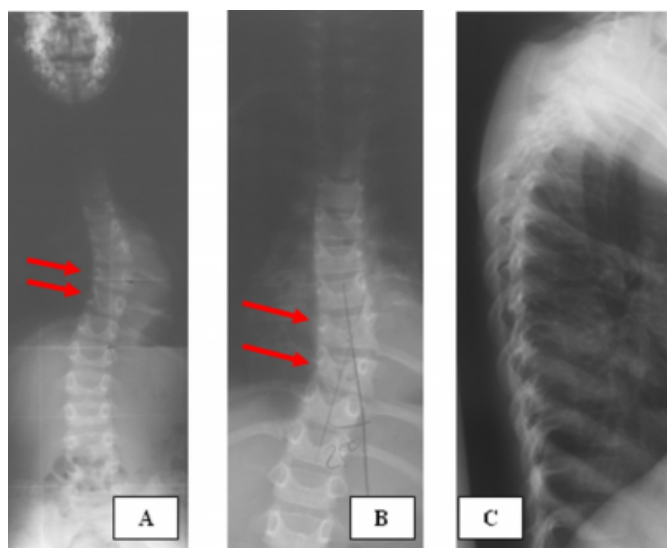
We describe the case of an 8 year and 9 month old boy, who had been in permanent medical care in a pediatric institution since one year of age because of a failure to thrive. At the time we saw him first he was 108.2cm high (about 14cm below 3rd percentile) and weighed 19.1kg (about 3kg below 3rd percentile). He had been thoroughly investigated but no medical explanations for his condition were discovered. Microsomia was mainly assigned to both a constitutional delay (pubertas tardas of the mother) and familiar short stature (body height of the mother 158cm and of the father 164cm).

The boy presented to us with an asymptomatic scoliosis that had been noted by the pediatricians looking after him. A connection between the scoliosis and the microsomia was not suspected at the time though. Clinically he showed no pathological findings such as neurological deficit, pain or cutaneous abnormalities. Radiographs (Fig. 1) showed a left-sided thoracic curve with a Cobb-Angle of 25°. Due to slight wedging of the ninth and tenth thoracic vertebrae and the absence of other deformities, the diagnosis of idiopathic

juvenile scoliosis was made and we recommended an MRI of the spine as a routine screening investigation. The scan (Fig. 2,3) revealed a large paravertebral tumor on the left with intraspinal extensions through the neural foramen at T8/9 and T9/10.

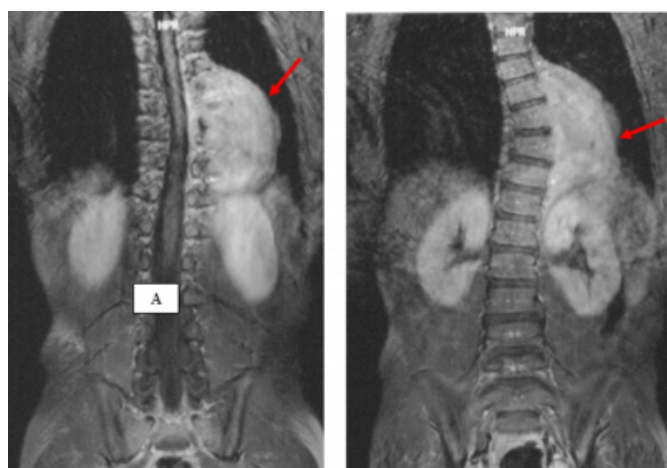
**Figure 1**

Figure 1: A x-ray spine A.P., B thoracic A.P., C thoracic lateral



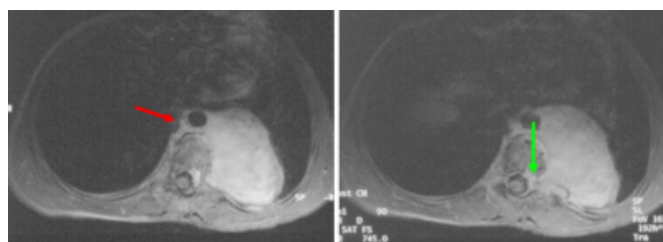
**Figure 2**

Figure 2: Sagittal MRI with left paravertebral tumor ( red arrow ).



**Figure 3**

Figure 3: Axial MRI. Demonstrates encapsulation of the aorta by tumor ( red arrow ) and the intraspinal growth through the neural foramen ( green arrow ).



The presumed diagnosis was neuroblastoma with a differential including ganglioneuroma. A biopsy was performed at a pediatric oncology clinic showed that the tumor was a ganglioneuroma. The patient underwent resection of the extraspinal intrathoracic parts of the tumor in two sessions (due to its size). In a further procedure, much of the intraspinal tumor was resected by a neurosurgical team. A complete resection of the intraspinal part was not feasible due to bleeding and the risk of neurologic sequelae. Postoperative recovery was uneventful, with no neurologic or systemic complications. Adjuvant therapies were not performed and the boy is now under permanent oncology follow up.

## DISCUSSION

Ganglioneuroma is a rare, benign tumor originating from the ganglion cells of the sympathetic nervous system and adrenal medulla. It is seldom associated with scoliosis. Cote et.al. <sup>19</sup> reported a case of a 12 year old girl with normal clinical examination, initially diagnosed as having adolescent idiopathic scoliosis. A tumor was found during surgical correction of the scoliosis. Rigault and Padovani <sup>20</sup> reported three cases, one with a rapidly progressive paraplegia and two having preceding resection of tumors as infants who then developed recurrent tumors only discovered during surgical correction for progressive scoliosis. The management of neural axis pathologies found coincidentally during scoliosis surgery as described in these reports can lead to considerable postoperative neurologic complications. Therefore many clinicians have searched for a better way to detect clinical features suspicious for neural axis pathologies before embarking on surgery. Citron et. al. <sup>3</sup> stated that pain and neurological deficits should lead to diagnostic tests beyond plain radiographs, such as protein estimation in the cerebrospinal fluid, CT and as standard method contrast myelography. Similarly, Schwend et.al. <sup>17</sup> reported that the combined symptoms of pain and a

neurological deficit was a primary indication for MRI scanning. In contrast, Davids et.al.<sup>21</sup> found no neural axis pathology in 20 patients with presumed idiopathic scoliosis who complained of spinal pain and therefore underwent MRI scanning.

There is controversy regarding minor neurologic deficits as indicators of underlying pathology. Zadeh et.al.<sup>22</sup> described an absent superficial abdominal reflex as an indication for further investigations, whilst Evans et.al.<sup>1</sup> believed the abdominal reflex was not a reliable marker. In general most authors stress the importance of a thorough neurological examination to detect a possible neurogenic origin for scoliosis. However, a normal neurologic examination may not necessarily exclude a neurogenic cause since scoliosis can be the only presenting sign of neural axis pathology in an otherwise normal examination<sup>5,12</sup>.

Spiegel et.al.<sup>23</sup> suggested that some atypical curve patterns such as left thoracic should be followed by MRI. Evans et.al.<sup>1</sup> in contrast found that only 50% of the neural axis pathologies in their study were associated with left sided curves and Mejia et.al.<sup>13</sup> performed a prospective study of left thoracic curves and found only 7% with a neural axis abnormality.

One possible reason for the diverse conclusions of these studies could be the age of the patients. Evans et.al.<sup>1</sup> examined juvenile scoliosis patients, whereas Mejia et.al.<sup>13</sup> had only two out of 29 patients below the age of eleven. Results seem to be more consistent if the subset of "juvenile" children according to James'<sup>24</sup> age classification is examined. Lewonowski et.al.<sup>4</sup> found a 19.2% incidence of neural axis pathologies in 26 consecutive patients who were less than eleven years of age. Similar results were reported by other authors for infantile and juvenile scoliosis<sup>1,2,18</sup>. However, the same incidence does not exist in cases of adolescent scoliosis. Winter et.al.<sup>10</sup> saw no need for routine MRI in typical adolescent scoliosis. Other authors have found similar results<sup>9,11,25</sup>. Jaramillo et.al.<sup>26</sup> believed that MRI was beneficial in juvenile, infantile and congenital scoliosis, whereas it is unnecessary in a typical case of adolescent scoliosis. Morcuende et.al.<sup>16</sup> concluded that the onset of a scoliosis before the age of 10 years was suspicious for an atypical scoliosis. They found a 10% incidence of neural axis pathology on MRI in these young patients, although the most predictive indicators were neurologic abnormalities and severity of curve despite immaturity.

Davids et.al.<sup>21</sup> believed pain, abnormal neurological findings or atypical curve patterns were the indications for MRI in their adolescent patients. They also added the absence of the typical apical segment lordosis as an indicator and found this to be the most valuable single indicator for abnormal findings. Ouellet et.al.<sup>27</sup> also found this pattern to be suspicious for neural pathologies in their patients.

## CONCLUSIONS

It seems that there are some risk factors, either singly or in combination that might predict the presence of neural axis pathologies in presumed idiopathic scoliosis. Their significance is only partially understood and further research is necessary. Open questions such as the clinical significance of minor neurological abnormalities, cost effectiveness of investigations, the ideal screening technique, the impact of neural axis pathologies on the clinical course of the scoliosis and what are the criteria for screening with special regard to the child's age, must be answered before final recommendations can be made. Nevertheless, in our clinical practice, we feel that MRI screening in infantile and juvenile scoliosis patients is justified. In adolescent patients possible risk factors for neural axis pathologies should be carefully looked for before MRI scanning is considered necessary.

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