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Arteriovenous malformation of the spine as a cause of scoliosis

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Spinal arteriovenous malformations are rare abnormalities that are difficult to recognize and treat. They represent 3.3 to 11 percent of all spinal cord tumours. The signs and symptoms most commonly encountered consist of progressive upper and lower motoneuron deficits, eventually leading to severe crippling disability. Ten to twenty percent of these patients will present with an associated spinal deformity that must be differentiated from idiopathic scoliosis. The case presented in this paper illustrates the diagnostic difficulties when spinal arteriovenous malformation is associated with scoliosis.
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KEY WORDS: spine, arteriovenous malformation, scoliosis.

Les malformations artério-veineuses de la colonne vertébrale sont des anomalies rares difficiles à détecter et à traiter. Elles représentent de 3,3 à 11 % de toutes les tumeurs de la moëlle épinière. Les signes et les symptômes rencontrés le plus souvent consistent en des déficits graduels des montoneurones supérieurs et inférieurs, aboutissant éventuellement à un sérieux handicap. De 10 à 20 % de ces patients auront en plus une malformation de la colonne vertébrale à ne pas confondre avec une scoliose idiopathique. Les cas présenté dans cet article montre les difficultés de diagnostic quand une scoliose s'ajoute à la malformation artério-veineuse.
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MOTS-CLÉS : colonne vertébrale, malformation artério-veineuse, scoliose.

Introduction

Spinal arteriovenous malformation (AVM) is a rare abnormality representing 3.3 to 11 percent of all spinal cord tumours.[1] It is a tangle of dilated vessels forming an abnormal communication between the arterial and venous systems, located within the covering of the cord and/or the cord itself. The lesion is caused by a persistence of embryonic blood vessels within the cord and/or its surroundings.[2]

Scoliosis commonly occurs in patients with spinal vascular malformations. Aminoff[1] found a scoliosis or kyphosis in 22% of his 60 patients with spinal AVM; while Dublin and Djindjian[3] reported that 9% of their 175 patients with spinal AVM had an associated scoliosis. The exact etiology of the spinal deformity is not well understood. However, like spinal cord tumours, it is believed to occur secondary to neurological damage causing muscle imbalance, or secondary to compression and disturbed blood flow to the surrounding osseous structures leading to vertebral body asymmetry.[4] The following case illustrates the progression of scoliosis associated with spinal arteriovenous malformation.

Case report

A fourteen-year-old girl presented to the Royal University Hospital Scoliosis Clinic with a progressive scoliosis. This curve had been recognized two years previously by her family physician, who subsequently referred her to an orthopaedist. A 33 degree right thoracic curve was measured and she was prescribed a Milwaukee brace that she wore constantly for two years. Still, the curve continued to progress and the patient was referred to the Scoliosis Outpatient Clinic for a second opinion.

This active teenager denied back pain or any functional impairment. She was 18 months post-menarche and appeared quite mature. The past and family history were unremarkable.

On examination, she had a right thoracic curve with 2.5 cm rib hump and a left thoracolumbar curve with a 1.5 cm transverse process hump. The curves appeared flexible on side bending, but a 2.0 cm left trunk shift was observed. Two large café au lait spots were present, but no other stigmata of neurofibromatosis were identified.

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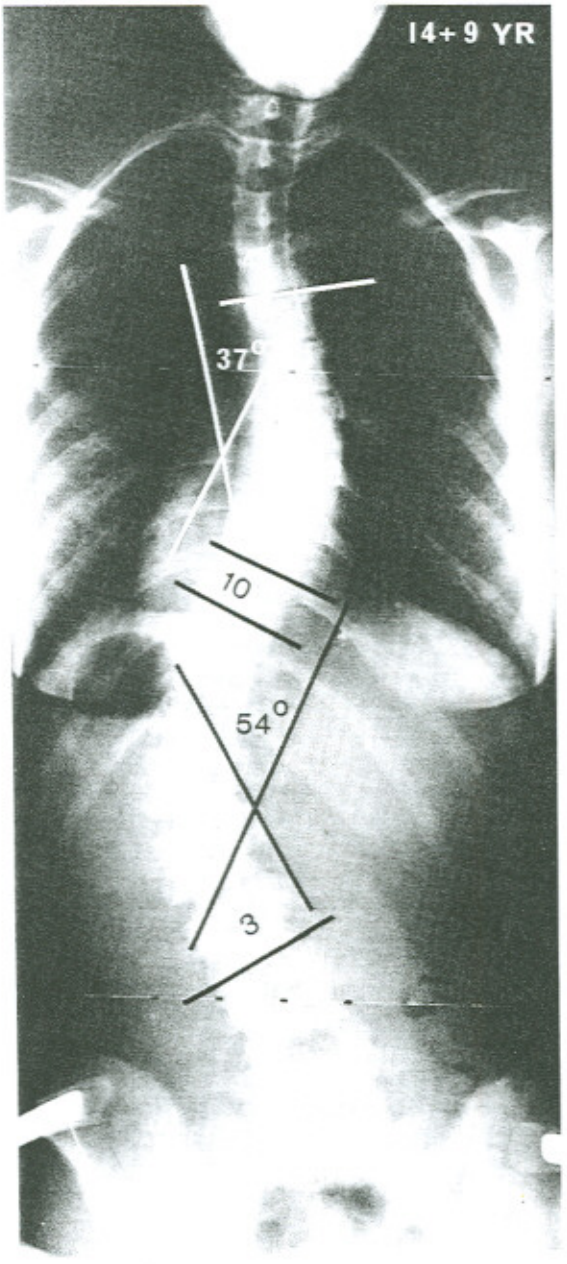

Figure 1 The posteroanterior (PA) scoliosis view shows a 37 degree right thoracic curve measured from T5 to T10 and a 54 degree left thoracolumbar curve measured from T11 to L3.

The radiological examination consisted of a posteroanterior and lateral scoliosis view. A 37 degree right thoracic curve and a 54 degree left thoracolumbar curve were measured (Figure 1). Mild posterior scalloping of the lumbar vertebrae was seen on the lateral radiograph, but it was not associated with other anomalies (Figure 2). The radiographs showed a well-developed iliac crest apophysis (Risser IV) that indicated spinal maturity. Bracing was discontinued since the curve was unlikely to progress.

During the following six months, she developed back pain after activity. Her scoliosis progressed to 40 degrees in the thoracic spine and 59 degrees in the thoracolumbar spine. Supine lateral bending films were taken to evaluate curve flexibility. These revealed a stiff thoracic and a flexible thoracolumbar curve (Figure 3). The patient also complained of a tender subcutaneous lump over the right shin after brusing it two months previously. The radiographs of the right tibia and fibula were normal, except the fibula appeared gracile (Figure 4). She was sent to a neurofibromatosis clinic for further assessment and no evidence of the disease was found.

At the next six-month follow-up, the scoliosis had further progressed. Her upper and lower curves measured 43 and 68 degrees respectively (Figure 5). She did not have any neurological deficit, but her right calf was smaller than the left and her right lower extremity was hyper-reflexic with associated ankle clonus. A myelogram, followed by a CT scan, revealed an expansion of the cord between the second and eighth thoracic levels (Figures 6 and 7). The contrast column around the expanded cord appeared irregular. Neurofibromatosis was suspected and the patient was referred for a MRI (Figure 8). This showed mixed signal intensity within the expanded cord on the flow weighted gradient echo study. The appearance of this lesion was consistent with spinal arteriovenous malformation. It was not possible to determine if the AVM was intra- or extramedullary in location. A spinal angiogram showed an intramedullary AVM at the T4–6 spinal levels that was fed mainly by the right sixth intercostal artery (Figures 9 and 10).

The patient was next seen eighteen months following her initial visit. She had had two transient episodes of paraesthesia in her right arm and leg in the previous six months. There were no signs of progressive neurological deficit. Further progression of the scoliosis was observed. The right thoracic curve measured 45 degrees while the left thoracolumbar curve measured 73 degrees (Figure 11). The decision was made to obliterate the AVM prior to surgical correction of the scoliosis. She was referred to a major centre for embolization of the malformation. However, they were unable to introduce a catheter into the main feeding vessel of the lesion and felt that her neurological status was too good to attempt such a high risk intervention. It was decided to await further progression of the scoliosis or deterioration of her neurological status and then attempt embolization of the malformation.

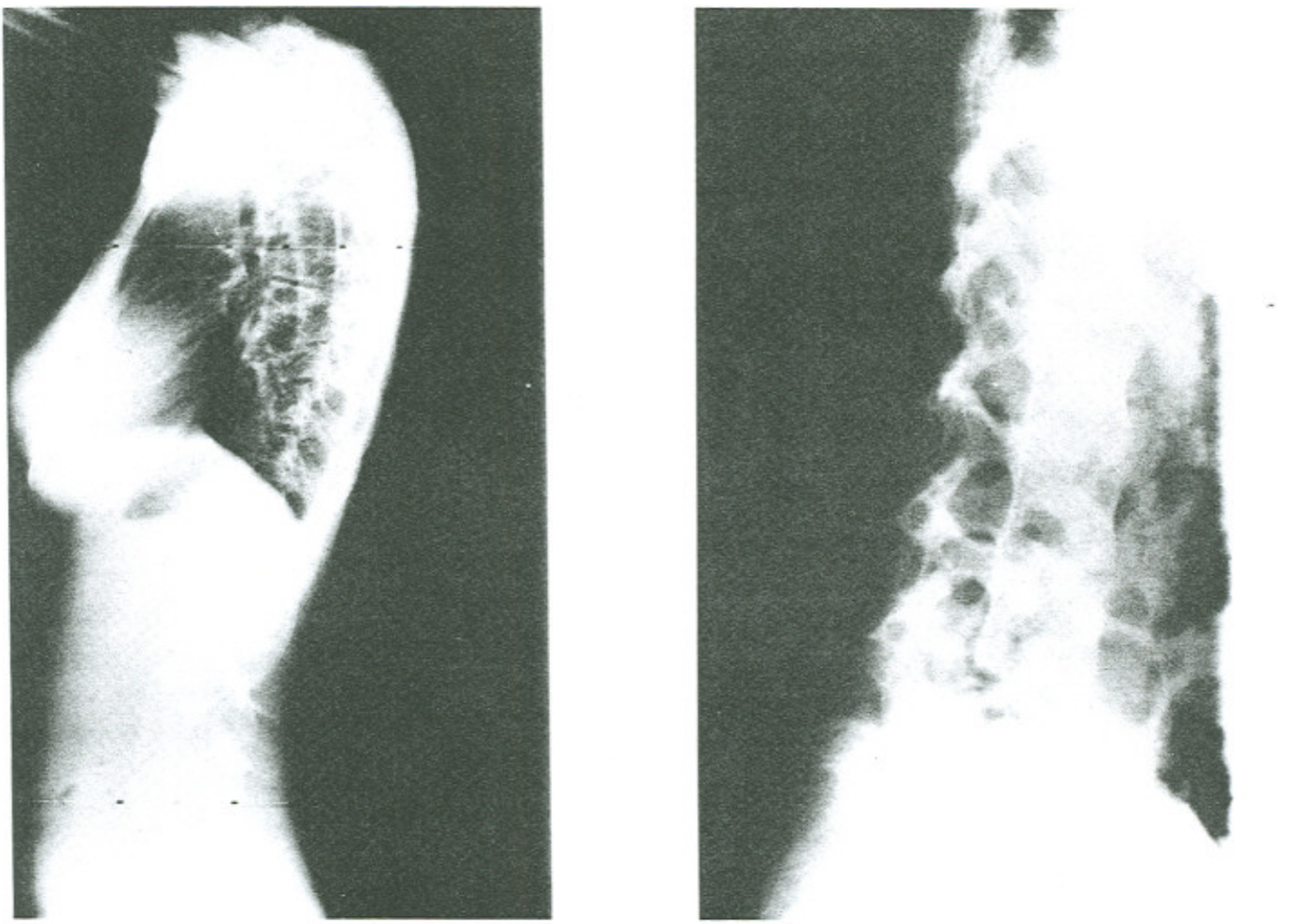

Figure 2 **(a)** The lateral scoliosis view shows flattening of the thoracic kyphosis and mild posterior vertebral body scalloping in the lumbar spine. **(b)** The sectional lateral lumbar view shows posterior scalloping of the lower lumbar vertebrae.

Discussion

Spinal arteriovenous malformation is a rare abnormality that is difficult to recognize and treat. Its presentation is highly variable, but the signs and symptoms most commonly encountered are combined upper and lower motoneuron deficit. The patient will present with pain (usually radicular in nature), motor and sensory deficit in the legs, and disturbance of micturition.[1] AVM can present at any age, but patients with intramedullary AVM tend to develop symptoms earlier than patients with extramedullary AVM. The average age of onset of intramedullary AVM is approximately 16 years younger than that of extramedullary (dural) AVM.[5]

The neurological deterioration of AVM patients tends to be slowly progressive, in a step-like fashion. It is often associated with remissions and exacerbations. This presentation can sometimes be confused with multiple sclerosis.[6,7]

The pathophysiology of symptoms in AVM remains obscure. Large subarachnoid haemorrhages might explain the symptoms; however these are rare and usually lead to sudden neurological deficit. The slow step-like deterioration could be explained by direct cord compression by the tangle of vessels or increased venous pressure within the cord that leads to venous congestion and cord infarction.[8]

AVM can cause spinal deformity. In this case, the scoliosis was first diagnosed as idiopathic. Yet, certain aspects of this case are atypical. While the progression of an idiopathic curve usually slows as the child approaches spinal maturity, in the presented case report the curve kept progressing rapidly. In

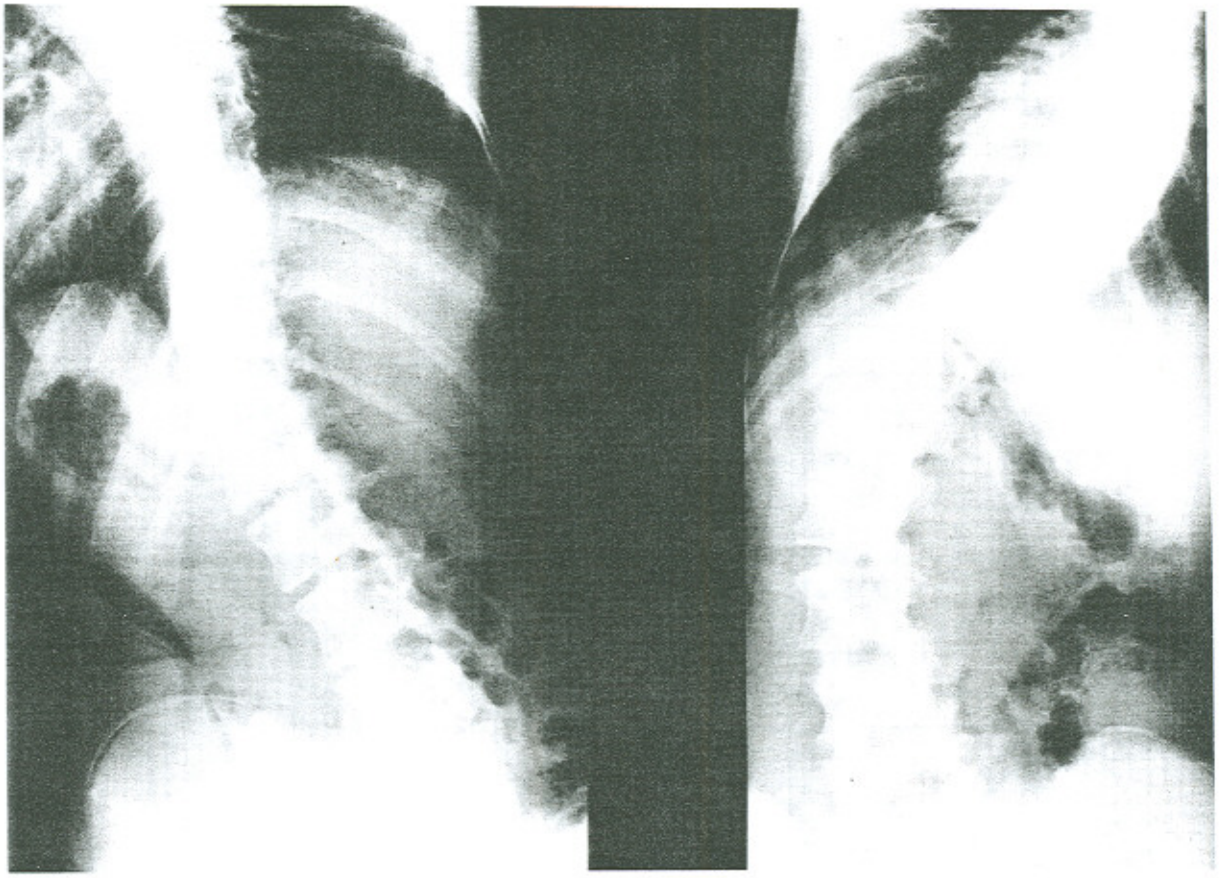

Figure 3 The supine lateral bending films show a stiff right thoracic curve and a flexible thoracolumbar curve. The iliac crest apophysis is not fused.

idiopathic curves, the primary curve is usually fixed while the smaller compensatory curve is flexible. However, in the above case, the exact opposite was found. Furthermore, the neurological abnormalities present in this case are uncommon with idiopathic scoliosis.

The above clinical presentation should raise suspicions of spinal cord pathology in the differential diagnosis. Several red herrings were present in this case. The cause of the scoliosis was initially believed to be neurofibromatosis because of the presence of two café au lait spots associated with posterior vertebral body scalloping and a gracile fibula. The CT scan seemed to confirm this suspicion, but the MRI and angiographic studies showed an intramedullary AVM.

The natural history of spinal AVM is disastrous. Within six years of the onset of symptoms, the majority of patients are severely disabled.[6,7] Operation or embolization of the malformation should follow the diagnosis as soon as possible, since patients with relatively mild neurological deficits show convincing improvement.[5] However, the decision to treat is complicated by the high risk of creating additional deficit during the surgery. This is especially true in cases where the lesion is intramedullary.[3]

Conclusions

Spinal arteriovenous malformations are rare vascular lesions leading to severe disability. Occasionally, the neurological deterioration can be preceded by the development of a secondary spinal deformity, such as scoliosis. A diagnosis of idiopathic scoliosis should not be considered until every other cause for the curvature has been ruled out.

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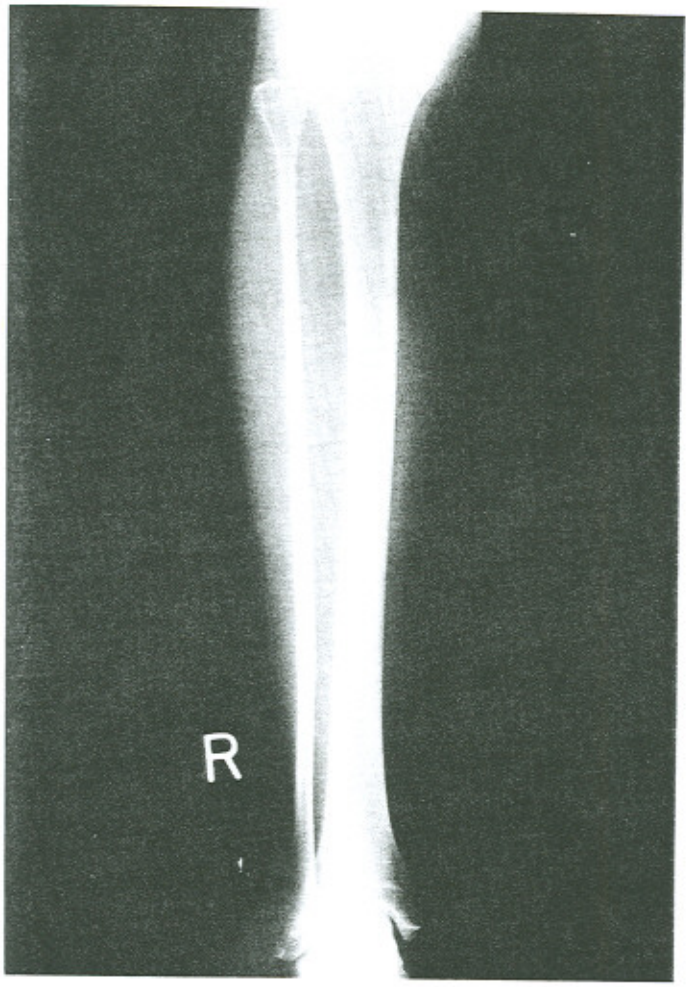


Figure 4 The anteroposterior radiographic view of the right tibia and fibula shows a gracile-looking fibula.

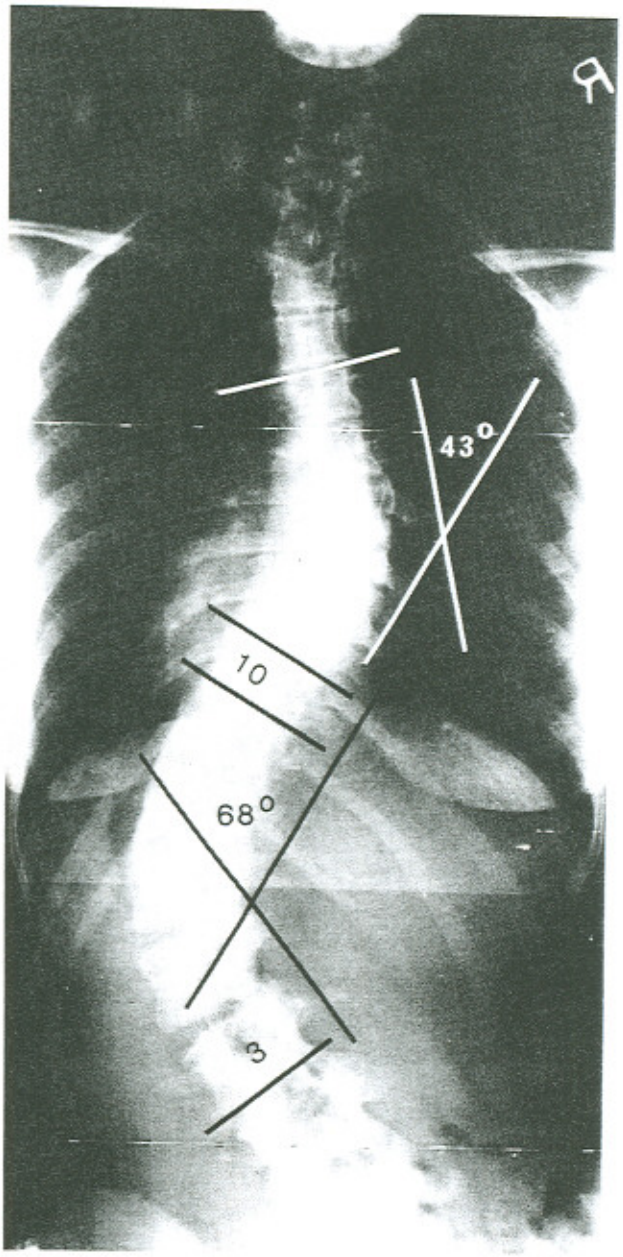


Figure 5 Twelve months after the initial visit, the curves have progressed to a 43 degree right thoracic curve and a 68 degree left thoracolumbar curve on the PA scoliosis view.

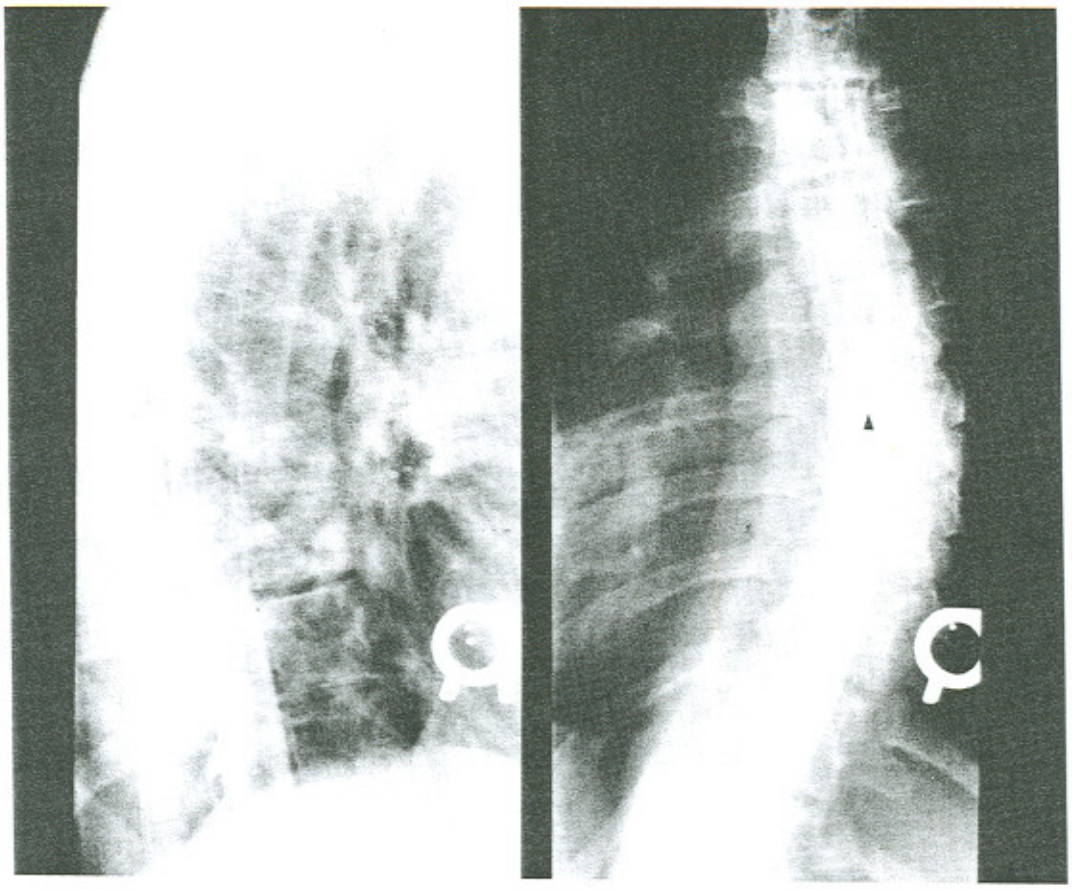


Figure 6 A myelogram of the upper (left) and lower (right) thoracic spine shows an expansion of the cord with circumferential thinning of the contrast media between the second and eighth thoracic levels. The lower border of the anomaly can best be seen on the lower thoracic myelogram. (arrow)

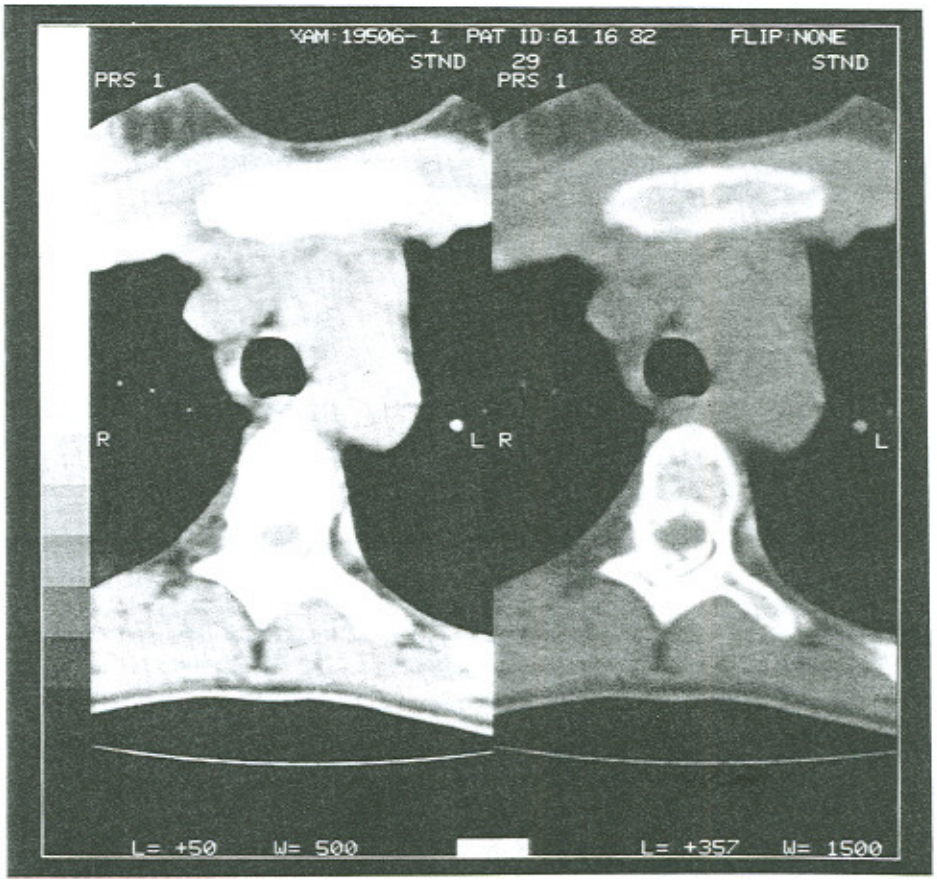


Figure 7 The soft tissue window (left) and bone window (right) of a contrast-enhanced CT scan taken through the sixth thoracic level show irregularities of the cord outline and thinning of the contrast column around the enlarged spinal cord.

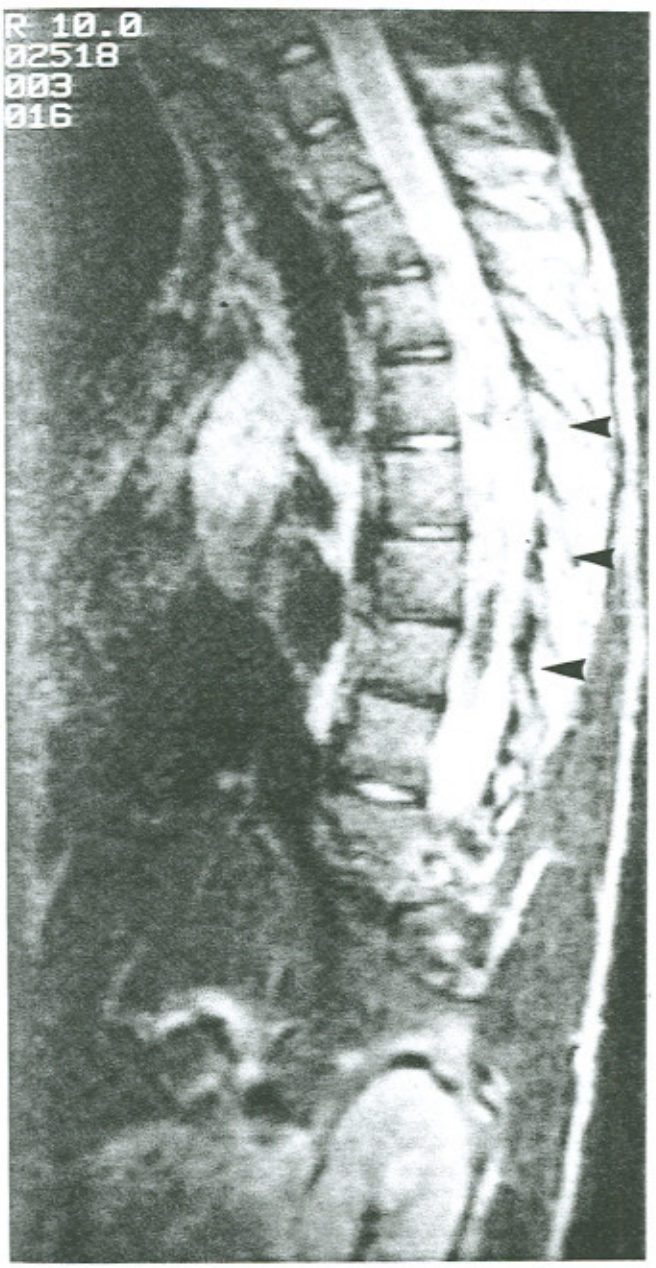


Figure 8 A sagittal section thoracic MRI scan shows mixed signal intensity within the enlarged cord. (arrows) This is suggestive of a spinal arteriovenous malformation, most likely intramedullary in location.

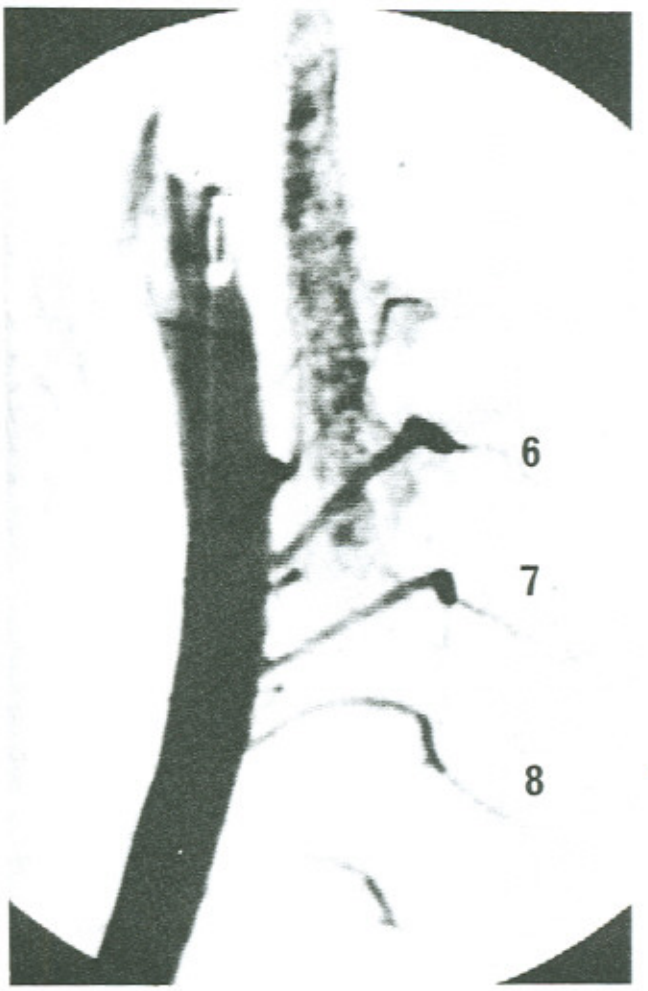
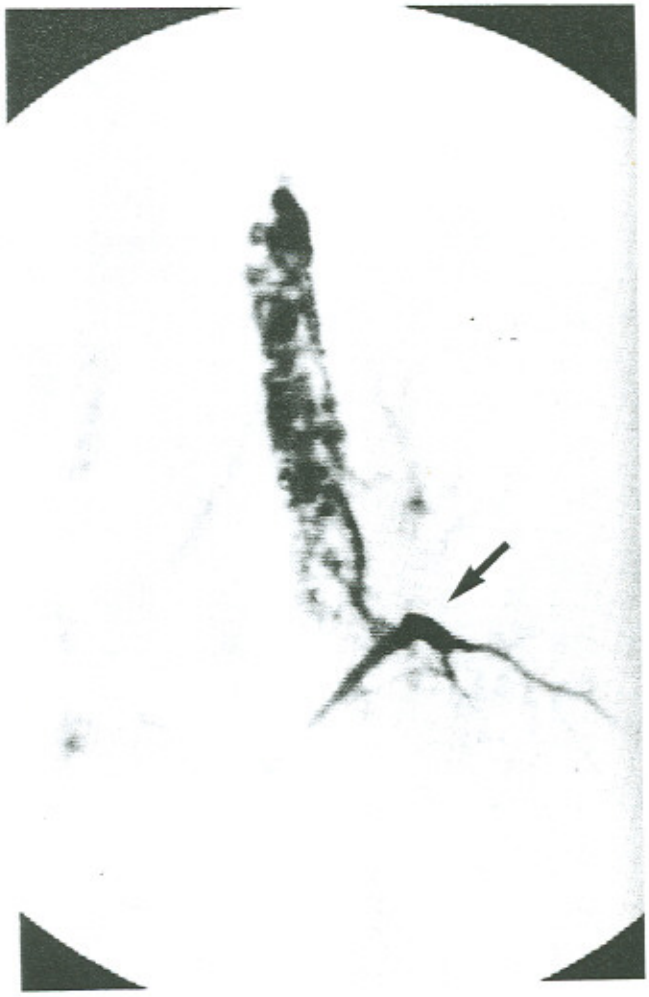


Figure 9 Spinal angiography shows an intramedullary AVM located superiorly to the large right sixth intercostal artery.



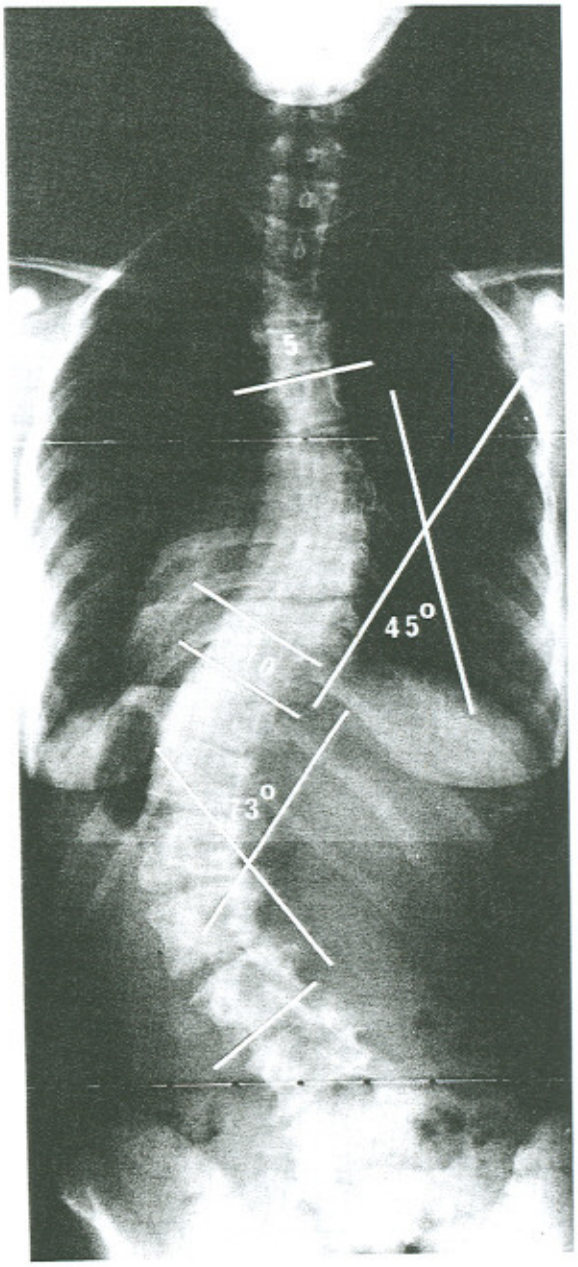


Figure 11 The last radiological examination shows further progression of the scoliosis. The thoracic curve measures 45 degrees and the thoracolumbar curve measures 73 degrees.

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