# Split Cord Malformation Associated with Severe Scoliosis and Tethered Cord Syndrome in an Adult: Case Report and Literature Review

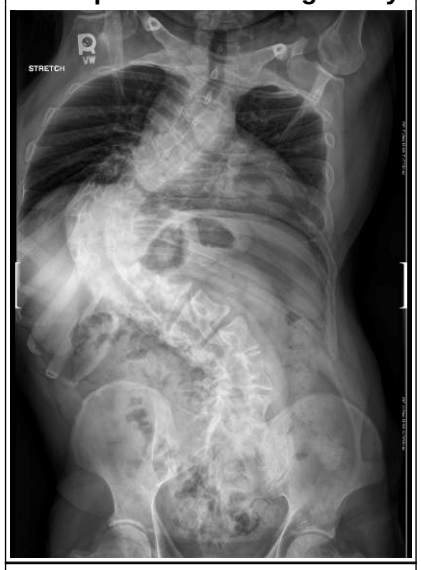
Kristopher Allen Lyon MD; Joseph Cleveland; David Garrett MD
Texas A&M College of Medicine - Scott & White Medical Center, Temple, Texas

CNS 2017 ANNUAL MEETING
BOSTON, MASSACHUSETTS
OCTOBER 7–11, 2017

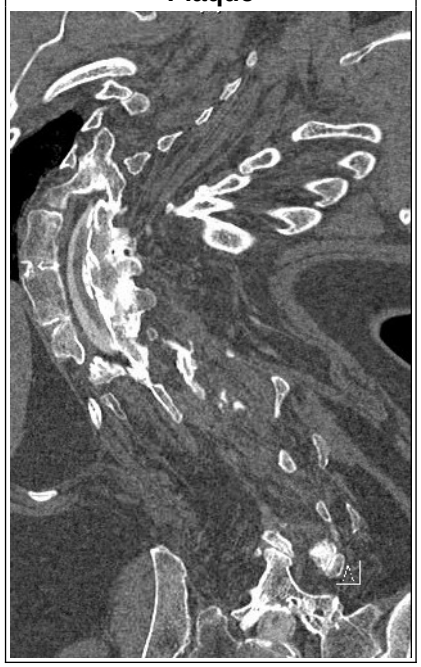
## Introduction

Split cord malformations (SCMs) are rare congenital abnormalities arising from a primary neurulation defect during development of the spinal cord. SCMs can lead to scoliosis and gait difficulties if left untreated. Traditionally, SCMs have been classified into two categories determined by the presence of a dural sac around each hemi-cord separated by a rigid septum (Type 1) or a single dural sac surrounding both hemi-cords separated by non-rigid tissues (Type 2). This case is one of the only known examples of a SCM with a single dural sheath, separated by a bony spicule in an adult.

**Pre-Operative Standing X-Ray**




**Coronal CT Myelogram Thoracic Spine with Calcified Plaque**



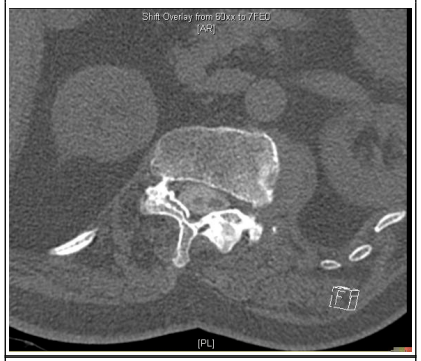
## Methods

We reviewed the clinical data and imaging from the index case and conducted a systematic review of the medical literature using PUBMED to determine the most common signs associated with SCMs to help clinicians screen patients for SCMs. Classification systems to accurately describe patients such as ours with unusual SCM pathology were explored.

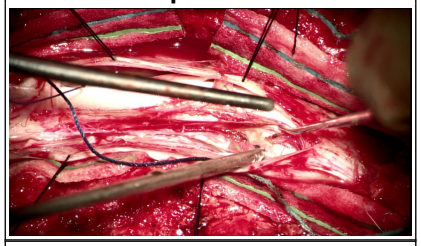
## Results

The most common sign associated with SCMs is hypertrichosis in up to 79% of patients. Up to 31% of patients with scoliosis can also have intraspinal abnormalities, with approximately 14% of these patients exhibiting a SCM. Type 1 SCMs can be further classified into Type 1a, 1b, 1c, or 1d based on the presence of a dividing bony spur and the location of the duplicated spinal cord in relation to it. Therefore, our patient is classified as Type 1b SCM, making him the only known reported adult to have this pathology.

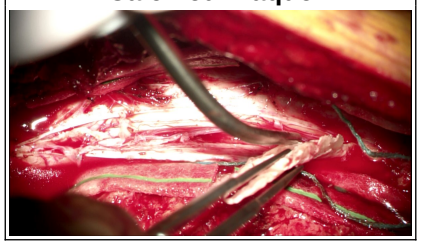
**Axial CT Myelogram Thoracic Spine with Two Distinct Spinal Cords**



**Intra-Operative SCM**



**Intra-Operative SCM with Calcified Plaque**



## Conclusions

As illustrated in the literature and our patient, prophylactic laminectomy to remove the septum causing a SCM could have prevented or minimized resultant scoliosis and neurologic sequelae. Given these findings, screening for SCM is recommended when congenital scoliosis or hypertrichosis is discovered in young patients.

## References

- Mahapatra AK, Gupta DK. Split cord malformations: a clinical study of 254 patients and a proposal for a new clinical-imaging classification. **J Neurosurg 103**(6 Suppl):531-536, 2005
- Pang D, Dias MS, et al: Split cord malformation : Part I : A unified theory of embryogenesis for double spinal cord malformations. **Neurosurgery 31:**451-480, 1992
- Yusuf I, Gonul M, et al: The diagnostic value of skin lesions in split cord malformations. **J Clin Neurosci 14:** 860 - 863, 2007
- Liu W, Zheng D, et al: Characteristics of osseous septum of split cord malformation in patients presenting with scoliosis: A retrospective study of 48 cases. **Pediatr Neurosurg 45:**350-353, 2009