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ORIGINAL ARTICLE

Intraspinal neural axis abnormalities in severe spinal deformity: a 10-year MRI review

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Abstract

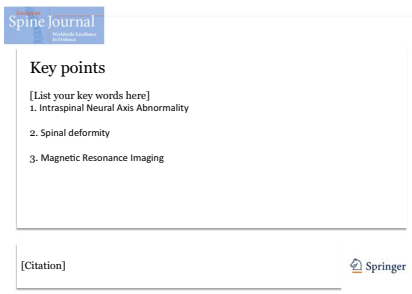
Purpose Documents indicated that the average prevalence of intraspinal neural axis abnormalities (INAAs) in presumed idiopathic scoliosis (PIS) patients was about 17.7%. However, paucity study focuses on the incidence of INAAs in severe spinal deformity (SSDs). In this study, we investigate the incidence of intraspinal neural axis abnormalities (IINAAs) and the clinical relevance in SSD at a single center.

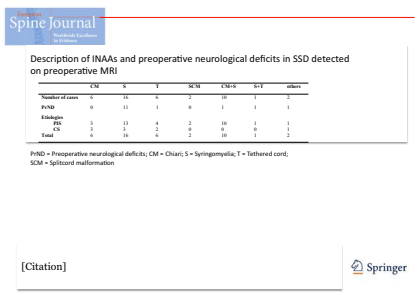
Methods All the patients with SSDs admitted for spinal surgery were evaluated from 2003 to 2014. Inclusion criteria: patients who present with coronal Cobb over 90° (and/or the sagittal Cobb ≥ 90°); patients with whole spine magnetic resonance imaging (MRI) done preoperatively; and patients with documented clinical findings preoperatively. Exclusion criteria: ankylosing spondylitis, adult onset scoliosis, scoliosis secondary to bone destruction, and spinal dysraphism.

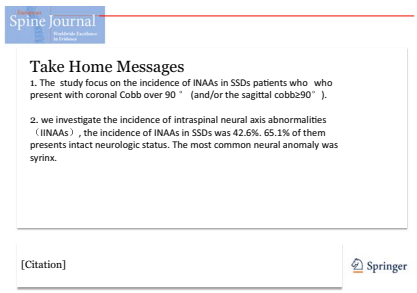
Results 101 patients fulfilled the criteria were included. 43 patients were detected with INAAs (42.6%, 43/101). The most common INAAs was syrinx (S) (16/43, 37.2%). Of which, 43.7% (7/16), 37.5% (6/16), and 18.7% (3/16) were spindle, slit, and swelling types, respectively. Most of them were located in thoracic (6/16, 37.5%) and cervical (5/16, 31.3%) region. MRI revealed Chiari malformation with syringomyelia (C + S) in ten patients (10/43, 23.2%), Chiari malformation (C) in 6 patients (6/43, 13.9%) and others in 11 patients (11/43, 25.6%). As to the etiology, most patients with INAAs were PIS (34/43, 79.1%). On clinical examination, 16 of 101 patients (16/101, 15.8%) had abnormal neurologic signs. 15 of 16 patients (15/16, 93.7%) with abnormal neurologic signs had INAAs on MRI. On the other hand, 28 of 43 patients (28/43, 65.1%) with INAAs on MRI presented neurologically intact. 28 of 85 patients (28/85, 32.9%) with neurologically intact were detected with INAAs on MRI.

Conclusion The incidence of INAAs in SSDs was 42.6%. 65.1% of them present intact neurologic status. The most common neural anomaly was syrinx. Preoperative whole spine MRI must be beneficial for SSDs even in the absence of neurological findings.

Graphical abstract These slides can be retrieved under Electronic Supplementary Material.







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Keywords Intraspinal neural axis abnormality · Spinal deformity · Magnetic resonance imaging

Introduction

With the development of magnetic resonance imaging (MRI), intraspinal neural axis abnormalities (INAAs), such as syringomyelia (S), Chiari malformations (CM), tethering of the cord (T), Chiari malformation with syringomyelia (C + S), and split cord malformation (SCM), are increasingly being found in patients with scoliosis. Through the previous studies, we are aware of specific curve patterns and clinical symptoms that warrant MRI for further evaluation because of the association with INAAs [1–3]. INAAs were shown in 20–38% of patients with presumed idiopathic scoliosis (PIS) (adolescent, infantile, and juvenile scoliosis) and 15–43% in patients with congenital scoliosis (CS) [4–9].

Severe spinal deformity (SSD) is a relatively uncommon condition; most often presents with severe, rigid, angular kyphoscoliosis or kyphosis with severe clinical symptoms, which often arises surgical management. In contrast to the more common PIS, the treatment of SSD patients has been identified the challenges [10]. The mechanism responsible for development of spinal deformity and neurological deficit in these disorders, such as higher tension of the spinal cord or compromises cells within the cord and ischemia at the site of tethering, is postulated to be unique for each [11]. As a result, all of these conditions in a patient potentially place them at a greater risk of developing iatrogenic neurological complications during corrective surgery. The INAAs often need to be addressed first to be able to treat the SSD patients more effectively and/or safely. Existing literature has clearly documented the prevalence of INAAs and current indications for MRI prior to surgical correction for PIS or CS [1–9]. However, there is paucity study about INAAs in the population with SSD patients.

Therefore, the current study analyzed the experience at a single center treating patients with SSD during 10 years in an attempt to (1) demonstrate the prevalence of INAAs in population with SSD patients and (2) to determine which clinical and radiographic characteristics correlate with INAAs on MRI in SSD patients.

Materials and methods

Institutional Review Board approval was obtained to perform this retrospective study. A review of the orthopedic, and radiology databases was performed at a single institution to identify patients with SSD admitted for spinal surgery were evaluated from 2003 to 2014. Inclusion criteria for the study were: patients who present with coronal Cobb over 90° (and/or the sagittal Cobb ≥ 90°); patients with whole spine MRI done preoperatively; and patients with documented clinical findings preoperatively. Exclusion criteria were: ankylosing spondylitis, adult onset scoliosis, scoliosis secondary to bone destruction due to infection, tuberculosis, tumour, previous operation, and trauma. Patients with clinically obvious spinal dysraphism were also excluded.

MRI study

INAAs were examined for the MRI analysis. T1W1 and STIR weighted sagittal, and T1W1 coronal and axial images of the cervical, thoracic, lumbar, and sacrococcygeal regions of the spine were examined using 1.5-T magnetic resonance equipment. A T2W1 sequence and T2W1 and STIR sequences were used to differentiate among syringomyelia, tethered cord. The position of the conus medullaris was confirmed, and the presence or absence of S, CM, T, C + S, and SCM was determined by the two orthopedic surgeons independently, and any patients whose examination demonstrated abnormal findings were referred to neurologists or radiologist for further evaluation. Each case of S was classified into one of the three groups: "swelling type," "spindle type," or "slit type" [12]. The location of syringomyelia in the region of spinal cord was also described with "craniocervical," "cervical," "cervicothoracic," "thoracic", or "holocord". The criterion used for the diagnosis of CM-I was herniation of at least one cerebellar tonsil 5 mm or more below the foramen magnum. Tonsillar herniation between 1 and 5 mm is termed tonsillar ectopia. The level of conus medullaris was also documented [13].

Etiology study

The patients were divided on clinico-radiological features into three broad groups according to the type of scoliosis as the prevalence and relation of intraspinal anomalies are known to be different in each type of scoliosis, CS, PIS, and others scoliosis. PIS included all patients who were diagnosed to have 'idiopathic' scoliosis at first presentation to the outpatient department.

Clinical examination

On neurologic examination, the presence of neurologic symptoms and abnormal neurologic signs such as sustained hyperactive reflex, unilateral superficial abdominal reflex, muscle atrophy, motor weakness, sensory loss, and bowel and/or bladder function was investigated. Any neurologic change during the preoperative skull-femoral traction was

also considered as a neurological deficit. Neurologic examination was performed separately by at least two spinal specialists. If any abnormalities were found, the patient was retested by another physician and a judgment was made by consensus.

Results

Demographics

A total of 101 patients fulfilled the criteria were included. The mean age was 18.9 years (range 10–45 years). There were 58 female and 47 male patients. The major curve of scoliosis was 108.9 ± 25.5 preoperatively, and the segmental kyphosis was 89.8 ± 31.1.

Intraspinal neural axis abnormalities

When the overall patient group was considered, there were 7 different INAAs in 43 (43/101, 42.6%) patients (13 males and 31 females) on MRI. The most frequent INAAs was S, with a 37.2% rate (16/43, 37.2%); other pathologies were CM + S in 10 (10/43, 23.2%), CM in 6 (6/43, 13.9%), and others in 11 (11/43, 25.6%). AS to the S, 43.7% (7/16), 37.5% (6/16), and 18.7% (3/16) were swelling, spindle, and slit types, respectively. Most of them were located in thoracic (6/16, 37.5%) and cervical (5/16, 31.3%) region (Table 1).

Etiology

When the etiologies were considered, there were 78 PIS patients (78/101, 77.22%), 21 CS patients (21/101, 20.80%), and 2 others patients (2/101, 198%) in all the SSD patients. As to the SSD patients with INAAs, there were 34 PIS patients (34/43, 79.06%) and 9 CS patients (9/43, 20.93%). In PIS with INAAs patients, S was the most common INAAs, with a 38.2% rate (13/34); other pathologies were CM + S in 10 (10/34, 29.4%). In CS with INAAs patients, CM (3/9, 33.3%) and S (3/9, 33.3%) were more common than the other INAA (Table 1).

Physical findings

On clinical examination, 16 of 101 patients (16/101, 15.8%) had abnormal neurologic signs. 15 of 16 patients (15/16, 93.7%) with abnormal neurologic signs had INAAs on MRI, including 3 patients with swelling type of S developed transient neurological deficit during traction who had complete recovery as traction weight was reduced. The INAAs in patients with neurologic signs were S in 11, CM + S in 1, T in 1, S + T in 1, and others in 1. On the other hand, 28 of 43 patients (28/43, 65.1%) with INAAs on MRI presented neurologically intact, which included 5 S, 9 CM + S, 6 CM, 5 T, 2 SCM, and 1 T + CM + S. 28 of 85 patients (28/85, 32.9%) with neurologically intact were detected with INAAs on MRI (Table 1).

Discussion

To our knowledge, this is the first study of SSD and analysis of associated INAAs in the SSD population. A higher risk of neurological complications has been reported during surgery of scoliosis associated with INAAs [14]. There have been contradictory report about routinely obtaining MRI prior to surgical correction. All these studies aimed at PIS (juvenile, adolescent, and infantile), or CS to rule out INAAs [1–9]. However, the SSD patients should be distinguished from more mild-or-moderate spinal deformity requiring surgical treatment, because the preoperative assessment, evaluation, and neurologic risk for these curves can be substantially different from the more common moderate PIS curves. Xie et al. indicated that the association with INAAs is one of independent risk factors for neurologic deficits during surgical procedure to treat SSD patients [15]. To prevent potential neurological complications, INAAs need to be addressed before the treatment of SSD patients. Documenting the incidence, varieties of INAAs in SSD patients will help the surgeons to choose patients requiring further investigation.

All the patients, in this study, were SSD patients, whose coronal Cobb' angle of major curve was 108.9 ± 25.5, and the segmental kyphosis was 89.8 ± 31.1. The mean age was 18.9 years (range 10–45 years). We report a more frequent of INAAs (42.6%) than in other studies before, which

Table 1 Description of INAAs and preoperative neurological deficits in SSD detected on preoperative MRI

Type of INAAs	Chiari (C)	Syringomyelia (S)	Tethered cord (T)	Splitcord malformation (SCM)	C + S	S + T	Others
Number of cases	6	16	6	2	10	1	2
Preoperative neurological deficits	0	11	1	0	1	1	1
Total	6	16	6	2	10	1	2

PrND preoperative neurological deficits, *CM* Chiari, *S* syringomyelia, *T* tethered cord, *SCM* splitcord malformation

indicated the average prevalence of INAAs associated with PIS or CS was about 15–40% [1–9]. Most patients in this study were PIS patients (78/101, 77.22%) and CS patients (21/101, 20.80%). As to the SSD patients with INAAs, most them were also PIS patients (34/43, 79.06%) and CS patients (9/43, 20.93%), although the exact nature of the relationship between these malformations and scoliosis remains to be defined. However, PIS is a diagnosis of exclusion; that is, other organic causes must have been ruled out by physical examination or imaging. Therefore, perhaps, the reason for this result was that some of these patients with INAAs were not sought health care and/or diagnosed with MRI at their early onset until final severe curve magnitude.

The syringomyelia is either a fluid-filled, gliosis-lined cavity within the spinal cord parenchyma or it is a focal dilatation of the central canal. In the setting of CM or T, the disturbance in normal cerebrospinal fluid (CSF) is caused by the downward herniation of the cerebellar tonsils or metabolic and ischemic changes in the distal spinal cord, obstructing normal CSF flow from the cranial to spinal compartment during the normal cardiac cycle, which is a communicating syrinx with swelling, spindle types. S without CM is described as after segmentally occluding the central canal of the spinal cord, that an increase in arterial pressure may cause CSF to enter directly into the parenchyma of the cord via perivascular spaces and cause an enlarging noncommunicating syrinx with slit type [16–18]. S usually occurs most commonly the CM and T. Some insight is provided by studies of the imaging prevalence of CM, which have attempted to define the imaging prevalence of syringomyelia among those patients with CM. Kelly et al. described that scoliosis associated with the presence of a CM is up to 20% of patients, and even more frequently associated with CM in the setting of S, with rates as high as 60% [19–22]. Most syringomyelia, in the scoliosis patients associated with CM or T, CM-associated syringes tend to occur in the cervical spinal cord (15–21%) and cervicothoracic spinal cord (12–25%) [23, 24]. In our study, the most common INAAs included S (37.2%), CM (13.9%), or a combination of the two (23.2%). As to the types of S, most of them were spindle, slit types, and were located in thoracic and cervical region, noted in this study.

Pressure from an asymmetrically expanding syrinx may be imparted to the medial nuclear group of cells and irreversible damage by the syrinx to nerve cells may allow the scoliosis to progress and present with neurologic abnormalities on examination. Some previous studies suggested that routine preoperative MRI is not indicated in IS unless the patient has neurologic deficits or symptomatic one [1–3]. Actually, many patients with scoliosis secondary to INAAs also had normal neurologic findings. Inoue et al. mentioned that 18 (9%) of 204 patients without abnormal neurologic signs had INAAs on MRI [25]. Ozturk et al. also showed that 20 (8%) patients who were neurologically intact had INAAs on MRI [26]. Some studies even have indicated that the incidence of MRI abnormalities in IS with a negative history and physical examination was up to approximately 20% [27, 28]. The current study indicated that most of the SSD patients with INAAs (28/43, 65.1%) are asymptomatic. On the other hand, 15 of 16 SSD patients (15/16, 93.7%) with abnormal neurologic signs had INAAs on MRI. According to Belmont et al., an abnormal finding on the history or physical examination demonstrated an accuracy of 71%, sensitivity of 56%, specificity of 76%, positive predictive value of 42%, and negative predictive value of 85% for the diagnosis of an intraspinal anomaly [29]. Such more frequent of INAAs in our research may influence orthopedic surgical treatment in SSD patients by dictating the use of spinal shortening strategy, because these in theory interfere with spinal cord tension more than PIS patients, by neurosurgical treatment before correction surgery, and perhaps by discouraging aggressive curve correction. It is also suggested that patients with SSD patients are indicated for routine preoperative MRI study even with normal physical examination and asymptomatic.

Our study, therefore, offers additional evidence, from a single center, to suggest that the incidence of INAAs in SSDs was 42.6% and most of them present intact neurologic status. To prevent iatrogenic morbidity and ensure the efficacy of the treatment, preoperative whole spine MRI must be beneficial for SSDs even in the absence of neurological findings.

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Compliance with ethical standards

Conflict of interest No benefits in any form have been or will be received from a commercial party related directly or indirectly to the subject of this manuscript.

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