Literature Review of Thoracic Myelopathy: Causes of Acute Worsening

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Abstract

Thoracic myelopathy is defined as a dysfunction of the spinal cord secondary, but not limited to, fracture, compression, and ischemic changes. Several studies have reviewed patients with thoracic myelopathy but incidence rates are unclear due to low sample size and improper diagnosis. Idiopathic thoracic spinal cord herniation (ISCH) and ossification of the ligamentum flavum (OLF) is reported to be more prevalent among the middle-aged and females, however males predominated in larger reviews of patients with thoracic spinal stenosis (TSS) and surgical treatment of OLF secondary to thoracic myelopathy. Common causes of thoracic myelopathy include ossification of the ligamentum flavum, posterior spur, and disc herniation. Possible etiologies for acute worsening of the spinal cord include iatrogenic cord insult, transient ischemia, reperfusion of neural tissue, microthrombi of spinal arterial supply, neuropraxia, and sudden decrease in blood pressure. Congenital kyphosis has also been implicated as a cause of thoracic myelopathy. Treatment varies widely from non-operative conservative measures to prompt surgical treatment with decompression via laminectomy, fenestration, and herniotomy. Treatment depends on the degree of mechanical stress leading to degenerative changes of the facet joints and thoracolumbar intervertebral discs. There is a paucity of literature on thoracic myelopathy secondary to acute worsening of the spinal cord and further clinical research is warranted to accurately diagnose and treat patients.

INTRODUCTION

Thoracic myelopathy is defined as a dysfunction of the spinal cord in the thoracic region secondary, but not limited to, fracture, compression, and ischemic changes.

INCIDENCE

Surgical intervention for patients with thoracic myelopathy has reported an incidence of 0.9 per 100,000 inhabitants between the years 1988-2002 [1]. Cervical spondylotic myelopathy, the most common cause of cervical myelopathy, has a reported incidence between 1.06 and 4.04 per 100,000 inhabitants [2,3]. Rates of myelopathy in the thoracic and lumbar regions are highly variable and considered rare due to unfamiliarity with the condition and misdiagnosis. Fewer than 650 cases of thoracic myelopathy have been reported. Only 89 cases of ventral spinal cord herniation have been reported since 1974 with the vast majority occurring in thoracic regions [4]. Among 95 patients with thoracic myelopathy with ossification of the ligamentum flavum (OLF), 7 patients (7.4%) were found to have spinal cord kinking (SK), a rare phenomenon associated with idiopathic spinal cord herniation [5]. Summers et al, reviewed 171 patients with idiopathic thoracic spinal cord herniation (ISCH), a major cause of thoracic myelopathy [6]. Between September 2004 and April 2008, 19 Chinese patients were reported to have ossification of the ligamentum flavum (OLF) secondary to thoracic myelopathy [7]. Between 2005 and 2012, 427 Japanese patients were reported to have thoracic spinal stenosis (TSS) [8]. Incidence of thoracic spinal cord compression with neurological impairment in patients with congenital kyphosis has been reported to be between 10-12% [9,10].

PREVALENCE

Idiopathic thoracic spinal cord herniation (ISCH) and ossification of the ligamentum flavum (OLF) is reported to be more prevalent among the middle-aged and females [6,11], however males predominated (1.4/1) in a larger review of 427 patients who underwent surgery following diagnosis of thoracic spinal stenosis (TSS) [4].

ETIOLOGY

There is a paucity of literature on predictive measures for thoracic spinal cord herniation with fewer than 200 cases [12].
Sato et al., performed an epidemiological review on thoracic myelopathy in a 1998 study of 81 residents of Northwest Japan. The most common causes of thoracic myelopathy include ossification of the ligamentum flavum (OLF) (64%), posterior spur (20%), disc herniation (19%), ossification of the posterior longitudinal ligament (OPLL) (16%), calcification of the ligamentum flavum (1%) and degenerative spondylolisthesis (1%) [4,13]. Wang et al., reviewed 95 Japanese patients with thoracic myelopathy secondary to OLF to gain a better understanding of the pathogenesis of spinal kinking, of which 7 patients were diagnosed. Spinal kinking was radiologically located at the end of the spinal cord between vertebral bodies T11 and L1 where the conus medullaris tip is compressed by OLF [4] and is typically a sign of spinal herniation [1]. Ascending myelopathy following thoracic spinal cord injury (SCI) is a rare cause of thoracic myelopathy but may provide insight into the disease progression with focus on spinal venous drainage and intraspinal pressure [11]. Idiopathic thoracic spinal cord herniation (ISCH) is a known but rarely encountered cause of thoracic myelopathy [14].

**ACUTE ETIOLOGY**

Acute thoracic myelopathy may be secondary, but not limited to, fracture, cord compression, and ischemic changes. Onset of myelopathy secondary to thoracic spinal stenosis may be triggered by acute events such as a fall [4]. A traumatic postoperative neurological deterioration was observed in a case study of 3 patients with severe mid thoracic stenosis that underwent decompression in the absence of spinal cord trauma. Possible etiologies include iatrogenic cord insult, transient ischemia, reperfusion of neural tissue, microthrombi of spinal arterial supply, neuropraxia, and sudden decrease in blood pressure [15]. Postoperative transient ischemia as a result of reperfusion following spinal decompression has also been suggested as an etiology for neurological degeneration in patients with chronic compression lesions [16]. Acute thoracic disc herniation can disrupt blood flow leading to anterior spinal artery syndrome (ASAS), the most common cause of spinal cord infarction. In the case of a 36-year-old female with acute onset of myelopathy, the role of diffusion-weighted imaging (DWI) in early recognition of ischemic changes proved useful [17]. Severe kyphosis can contribute to myelopathy by having a stretch effect on the spinal cord with subsequent reduction in blood perfusion [18], however it is also suggested that sudden onset of neurological degeneration may not correlate with kyphosis angulation. Further, the degree of neurological symptoms in kyphotic patients may not correlate with the severity of spinal cord compression [19].

**KNOWN PATHOLOGICAL CHANGES**

Acute spinal cord injury generally follows a two-step process including primary and secondary mechanisms. Primary mechanisms involve initial mechanical injury and can result in severe ligamentous injury, fracture, and ruptured discs leading to compressive forces on the spinal cord. These initiate a cascade of complex and inter-related secondary mechanisms including vascular changes, ionic and neurotransmitter imbalance, and apoptosis. Ischemia may present due to dramatic reduction in blood flow and has been experimentally shown to persist for at least 24 hours in rats and monkeys. Apoptosis proceeds via a caspase cascade with initial impact to local neurons but then spreading to down or upstream oligodendrocytes [20]. Radiographic changes can also be assessed following spinal cord injury, namely atrophy, myelomalacia, and cystic changes [21].

**TREATMENT**

Treatment of thoracic myelopathy varies widely from non-operative conservative measures to prompt surgical treatment [4,7,9]. Steroid treatment with methylprednisolone within 8 hours of trauma is widely used, however recently both its safety and efficacy has been disputed [22]. Lamrectomy is considered the gold standard for surgical treatment of OLF but other decompressions are available. Fenestration utilizes a longitudinal resection of the lamina and ligamentum flavum at the middle of the spinal canal. Herniotomy can be completed via a transversoarthropedicectomy approach with lateral herniated mass excision. The decision for surgical treatment largely depends on the degree of mechanical stress and degeneration to the facet joints and thoracolumbar intervertebral discs [1].

A review of 132 Japanese patients with thoracic myelopathy with secondary OLF underwent surgery between 1988 and 2002. Postoperative outcomes depended on the preoperative severity of myelopathy. The postoperative Japanese Orthopaedic Association (JOA) scores from patients with severe preoperative myelopathy were significantly lower than those of patients with only mild to moderate preoperative myelopathy [23,24]. A case study of a 61-year-old man with a history of progressing thoracic myelopathy with ventral herniation of the spinal cord, details the unfamiliarity clinicians are with the condition leading to under- and misdiagnosis and inadequate surgery. In this context it is crucial to differentiate between thoracic spinal cord herniation and posterior arachnoid cysts. Misdiagnosis of thoracic spinal cord herniation was reported to occur in 21% of cases resulting in wrong surgical therapy in the majority of misdiagnosed cases. Unfamiliarity is attributed to the rare presentation of the disease despite advances in medical imaging, however the authors recommend being watchful for common initial presentations that may lead to myelopathy to prevent misdiagnosis and provide adequate treatment. Patients with thoracic myelopathy most commonly present with motor and sensory deficits in the lower limbs. Since these symptoms are often similar to those of other cervical and lumbar spine disease, this might explain frequent delays in treatment. Furthermore, thoracic spinal stenosis from pathological changes, such as OLF, OPLL and thoracic disk herniation often coexist with lumbar and cervical spine disease [25]. The disease process is highly variable. Generally, the onset is insidious but may be triggered by acute events. Conservative measures, commonly steroid administration of methylprednisolone, may be more favorable if patient presents asymptomatic [7]. Summers et al. reviewed 78 publications with 171 patients treated after diagnosed with ISCH and concluded that management should be individualized, but
without further treatment evaluations, an adequate treatment strategy has not been formulated [5,6]. Taher et al., presented 3 patients who experienced initial worsening of myelopathy following thoracic decompression surgeries but whose condition later resolved beyond baseline scores [15]. Possible etiologies are aforementioned. Intraoperative loss of motor evoked potential (MEP) occurred in 2 of the patients after completed decompression [12]. Miscusi et al., observed two treatment cohorts, minimally invasive spinal surgery (MISS) and traditional open surgery, of oncological patients with spinal thoracic metastases causing acute myelopathy. Though no neurological differences were reported between the two treatment methods, MISS patients showed improvements important to maintain quality of life for oncological patients with short or midterm life expectancy [26].

OUTCOMES

In a retrospective review of 427 patients diagnosed with thoracic spinal stenosis (TSS) secondary to thoracic myelopathy, patients reported with pre-operative motor (81%) and sensory (64%) deficits in the lower limb [4]. Brown-Sequard syndrome and spastic paraparesis has also been reported as a major symptom in idiopathic thoracic spinal cord herniation (ISCH) [3,10], characterized as an atypical myelopathy. Clinical improvements were reported within 3 months following surgical treatment of ISCH for all 5 patients in a case study [27]. In a larger study of 19 patients with OLF, significant improvements were charted comparing pre- and post-operative Japanese Orthopedic Association (JOA), Oswestry Disability Index (ODI), and Visual Analog Scale (VAS) scores [3].

CONCLUSION

Acute worsening of the thoracic spinal cord is not well understood due to the rarity of myelopathy and its unfamiliarity; however certain presentations should raise suspicion of onset of disease to avoid misdiagnosis and inadequate treatment. Further clinical research is warranted to identify methods for proper diagnoses and early, adequate treatment.

REFERENCES

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