Cervicothoracic Diastematomyelia in an Elderly with Normal Neurology: Report of a Case and Review of the Literature

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ABSTRACT
Diastematomyelia, an uncommon dysraphic state usually seen in children, is a rarity in adults and a medical curiosity in the elderly. Among 55 adults with diastematomyelia, only three elderly have been published so far. Cervicothoracic and cervical region are the least common site for the spinal cord splitting, so it is not a surprise to encounter only ten adults with diastematomyelia of this area reported in the past. On the other hand, while, pain and abnormal neurology is a guiding feature of diastematomyelia which point to the diagnosis, to remain asymptomatic throughout the life time till the eighth decade should be very rare event. Herein, a 72-year-old neurologically asymptomatic female patient with cervicothoracic diastematomyelia is presented containing a set of these rare unique features.

KEY WORDS: Diastematomyelia, Dysraphism, Geriatric, Split cord malformation

INTRODUCTION
Diastematomyelia is an uncommon dysraphic disorder usually seen in children. It is rare in adults. In most of these adults, the lesion was located in lumbar and lower two third of the thoracic region. Discovery of cervical spine diastematomyelia in adults is considered to be very rare. In this study a case of cervicothoracic Diastematomyelia in an Elderly is presented.

CASE REPORT
This 72-year-old female was admitted to our clinic because of the neck pain and discomfort in the last 2 weeks. Her neurological exam was quite normal, Plain radiography disclosed blocked vertebra and some degenerative changes in the lateral planes while the AP view showed increased interpedicular distance and a central bony spur through C6 to T2 (Figure 1A,B). Despite amelioration of the pain with NSAID’s, with regard to radiological findings the patient was sent for MRI which showed split cord malformation including two separate hemicords extending from C 5 to T3 rejoining into a single wide dural sac distally (Figure 2A,B). Subsequently the axial CT and coronal CT reformats could confirm and delineate the long bony cleft (Figure 3A,B). This anomaly was discussed with the patient and she was advised to perform regular neck exercises and come for seasonal
follow up. The patient was followed every three months for 18 months and has not shown any neurological findings since then.

**DISCUSSION**

Diastematomyelia is a spinal dysraphism characterized by the presence of an osseous, cartilaginous, or fibrous septum which splits the spinal cord into two distinct hemicords. It is regarded as type I if the two hemicords have their own dural sac and type II if both hemicords are within a single common dural envelop.

Most patients with diastematomyelia present in childhood and rarely exhibit in adult life. In 1990,
Russel et al made a careful survey of the English literature and could encounter only 45 adult cases with diastematomyelia (19). We updated the review and could find 10 additional cases excluding those associated with inclusion tumors (2,4,5,12,13,16,17,22,). In most of these adults, the lesion was located in lumbar and lower two third of the thoracic region while diastematomyelia developing above T2 vertebra remained exceptional in adults and was confined to 10 cases including the current case (2,3,8,10,14,15,18,20,24) (Table 1).

Concerning diastematomyelias of all locations and ages, the condition is three to four times as common in females as in males in various series. But with regard to adults with SCM above T2, females were only two times more common than males.

The review confirmed that the age of the adult patients with diastematomyelia above T2 ranged from 19 to 72 year with a mean of 33.6 years.

Discovery of diastematomyelia in the geriatric age group in compared to adults is seen less frequently and is regarded extremely rare. Only three patients with diastematomyelia in the elderly above the age of 65 has been published in the past, none in the cervicothoracic region (5,9,23) (Table 2).

Surprisingly, the number of diastematomyelias expected to be found either incidentally or as a result of neurology do not correlate with increased availability of modern radiological tools. Only two new cases of adult cervical diastematomyelia have been published in the last two decade (1,18).

In children with cervical diastematomyelia, a short neck, low posterior neckline and limited range of neck movement which are the result of associated Klippel-Feil syndrome other than cutaneous stigmas might be the sole clues for diagnosis. Cutaneous stigmas and scoliosis which are good markers for underlying diastematomyelia of the thoracic and the lumbar region are rare in cervical diastematomyelia.

Moreover, in pediatric age group with cervical diastematomyelia, the neurological symptoms may develop

### Table 1

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Sex</th>
<th>Age</th>
<th>Location</th>
<th>Clinical Picture</th>
<th>Trauma</th>
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<tr>
<td>James</td>
<td>1960</td>
<td>F</td>
<td>56</td>
<td>C4-C5</td>
<td>Quadriplegia</td>
<td>?</td>
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<tr>
<td>Kuchner</td>
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<td>F</td>
<td>20</td>
<td>C5-C6</td>
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<tr>
<td>Beyerl</td>
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<td>38</td>
<td>C2-C3</td>
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<tr>
<td>Okada</td>
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<td>19</td>
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<td>Right upper Extremity atrophy</td>
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<tr>
<td>Simpson</td>
<td>1987</td>
<td>M</td>
<td>19</td>
<td>C6-C7</td>
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<td>-+</td>
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<tr>
<td>Wolf</td>
<td>1987</td>
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<td>27</td>
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<td>Quadriparesis</td>
<td>+</td>
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<tr>
<td>Ohwada</td>
<td>1989</td>
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<td>29</td>
<td>C6-T2</td>
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<tr>
<td>Rawanduzi</td>
<td>1991</td>
<td>F</td>
<td>33</td>
<td>C5C6</td>
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<td>-</td>
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<td>Armstrong</td>
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<td>37</td>
<td>C7-T2</td>
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<td>Present Case</td>
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<td>F</td>
<td>72</td>
<td>C5-T2</td>
<td>Intact Neurology</td>
<td>-</td>
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### Table 2

<table>
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<th>Author</th>
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<td>James &amp; Lassman</td>
<td>1962</td>
<td>67</td>
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<td>76</td>
<td>F</td>
<td>L3-L4</td>
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<td>Conti</td>
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<td>L1-L2</td>
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<tr>
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<td>2011</td>
<td>72</td>
<td>F</td>
<td>C5-T2</td>
<td>MRI</td>
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</table>
years later and even postponed till late teens ranging from mild to moderate in compare to the similar lesions in the lower levels. This discrepancy can be explained through the fact that developmental ascent of the spinal cord relative to the bony spinal column is less in the cervical region than the thoracolumbar region. This means that the cord is less affected by the ascent of the spinal cord in these regions in comparison to those in the lower regions. As a result, the neurological symptoms are expected to be less pronounced in former regions or even tending to remain occult till adult life. Late discovery is reasonable if we consider the absence of associated cutaneous stigmas in all reported adults with diastematomyelia above T2 vertebral. Remaining neurologically free till old age as was seen in the present case is therefore a medical curiosity. Nonetheless, in an adult subject with this pathology, sooner or later the neurology will be elicited either gradually by meeting the relatively fixed spinal with increased flexion extension of the neck against the septum resulting in tension and stretching on the neural tissues or with acute onset, precipitated by an obvious trauma.

A careful search of the literature showed that In adults with cervical or cervicothoracic diastematomyelia, exhibition of neurological symptoms had been with acute weakness of the extremities preceding with trauma in one third of the patients (3,10,20) while in the remaining two thirds, the neurological symptoms were subtle but progressive, varying from upper extremity weakness to quadripareisis (1,14,15,24).

Actually, there are no reports in the literature on how frequently asymptomatic diastematomyelia occurs in the general population. In a review of the literature we could find only a few case reports of the patients with occult and neurologically silent diastematomyelia of all locations (1,4,7,15). Probably in such rare instances, the hemicords should have been relatively free around the septum and therefore not easily affected by ascent of the cord or daily movements.

The value of plain radiographs in cervical diastematomyelia should not be ignored. The condition might be suspected with demonstration of multiple congenital anomalies in the cervical spine. Anomalies such as Klippel Feil, irregular laminar fusion, hemivertebra, spina bifida, increased interpedicular distance as well as the craniovertebral anomalies can lead to the discovery of underlying split cord malformation (10).

MRI is the procedure of choice for evaluation of the entire cord and the hemicords in the vicinity of the cleft. Associated anomalies such as syringomyelia, Arnold Chiari malformation, tethered cord and inclusion tumors can be best seen with the aid of this tool (5,13,18,20,21). However, MRI cannot differentiate the bony spur from fibrous cartilage emphasizing the value of CT and Metrizamide CT as an imaging method of choice in detection of the bony spur, duplication of the dural tube and the spinal cord (3,5,11,14,16,18,22,24).

The number of reported adults with cervical and cervicothoracic diastematomyelia is too small to formulate a definitive treatment protocol. However, surgery is strongly considered if progressive neurological deficits are detected. Dramatic improvement over a period of time after surgical intervention can be expected in such instances (3,10,14,15,18,20,24).

On the other hand, management of asymptomatic subjects remain controversial for the lack of natural history applicable to this age group (6). With concern about the clinical consequences of tethering of the cord, prophylactic surgery of diastematomyelia in adults even with minor neural symptoms is advocated by some authors, but most of the surgeons propose observation and monitoring of neurological function in asymptomatic patients with diastematomyelia (1,4,6,7). The same is true in the present case and the detection of a cervicothoracic diastematomyelia in an asymptomatic old woman in the eight decade of life indicates a need for observation rather than surgery.

In conclusion, discovery of cervical spine diastematomyelia in adults is considered to be very rare. It might only remain occult and undetected till old age in extremely rare occasions (9,23). In contrast to young adults and middle-aged asymptomatic subjects with high susceptibility to trauma which might support the concept of prophylactic surgery, such a lesion remaining silent and occult till the elderly will undoubtedly not necessitate surgical intervention.
REFERENCES

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