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# Segmental Myoclonus in a Child with Spinal Cord Tumour

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## ارتجاج عضلي قطعي في طفلة تعاني من ورم في الحبل الشوكيَ

*روش لال كوك ، براتب شاند*

**خلاصة** : نعرض هنا لحالة فريدة لطفلة عمرها سبع سنوات ظهر عليها إرتجاج عضلي قطعي عنقي . وقد أظهر التصوير بالرنين المغناطيسي للعمود الفقري ورماً بالحبل الشوكي العنقي .

ABSTRACT: A Seven year female child presented with cervical segmental myoclonus. Magnetic resonance imaging spine revealed a cervical cord tumor. A brief discussion about spinal myoclonus, a rare entity, is given.

Myoclonus is an involuntary, brief and sudden muscle jerk that occurs synchronously or asynchronously, symmetrically or asymmetrically and is focal or generalised. It may be rhythmic or arrhythmic. Based on clinical manifestations and site of origin it is divided into cortical, brainstem and spinal types (Marsden, Hallet and Fahn, 1982). Clinical features, electrophysiological findings, electroencephalogram (EEG) and imaging studies help in differentiating the three types. The spinal myoclonus is further classified into spinal segmental myoclonus and propriospinal myoclonus (Brown et al., 1991). In spinal segmental myoclonus (SSM) Myoclonus is limited to a few adjacent spinal segments while in propriospinal myoclonus the involuntary muscle jerks propagate up or down from the originating site of myoclonus in the spinal cord through slowly conducting polysynaptic pathways (Brown et al., 1991; Chokroverty et al., 1992; Kapoor et al., 1992).

Although Campbell and Garland (Campbell and Garland, 1956) are credited with first description of SSM, Penfield and Jasper (Penfield and Jasper, 1954) described this earlier in a patient with glioma of the lower thoracic spinal cord. Several diseases are associated with SSM such as tumours (Penfield and Jasper, 1954; Renault et al., 1995) trauma (Birbamer et al., 1993) infections, arteriovenous malformation, cysticercosis and demyelination (Kapoor et al., 1992). There may be no cause detected in some cases (Chokroverty et al., 1992). An association with hyperglycorrhachia has been seen recently (Bass and Lewis, 1995). Self limited cervical segmental myoclonus in association with familial dystonia has been reported in two children (Kyllerman et al., 1993).

What initiates the generation of SSM is not known. Hypotheses include local irritation, inflammation, hypoxia and deranged neurotransmitters. The latter mechanism is postulated on total control of myoclonus with valproic acid and benzodiazepines, suggesting GABAergic and serotoninergic mechanisms respectively in some cases.

We report a seven years old female child who presented as spinal segmental myoclonus with cervical cord tumour . She presented in 1994 with complaints of abnormal movements of both hands since the age of nine months (about 4 years duration). These movements were present during day and night without spread to any part of body. The movements had been gradually increasing with time. Six months ago, she had started with urinary frequency and precipitancy. Her developmental milestones were normal. Three brothers and four sisters were healthy. Examination revealed a cheerful co-operative girl with a height at 10$^{th}$ centile and weight at 5$^{th}$ centile. Higher functions, cranial nerves including palatal function were normal. There was drooping of left shoulder. There were sudden flexion movements of interphalangeal joints, thumb, palm and lower forearms occuring almost every second with some asynchrony between right and left hands. There was mild pronation and ulnar deviation of both hands. No abnormal

movements in upper arm, chest, abdomen, diaphragm or lower limbs were noted. All deep tendon jerks were elicitable and plantar reflexes were down going. She felt reduced pin prick on chest and back from dorsal sixth to tenth. There was no spinal bruit. The gait was normal. The complete blood count, liver and renal functions, thyroid function tests, serum ceruloplasmin and growth hormone levels were normal. X-rays cervical spine were normal.

Myelo-CT Scan of the spine showed widening of the cord shadow from cervical 5 to thoracic 1 region, suggesting intramedullary lesion. Electromyogram of the hand muscles showed spontaneous myogenic bursts occurring almost 60-80 times per minute. T sensory nerve conduction studies of med nerves were normal in both hands. All evc tests (visual, brainstem auditory, and so were normal. Magnetic resonance imaging was normal. MRI spine showed enlarge cervical cord from cervical 5 to thoracic (F widening was homogenous, smooth and shape. Signal of the cord on all sequence approximated likeness of normal cord astrocytoma.

Mild scalloping of the posterior vertel the corresponding region reflecting a long st was noted. The child was put on sodium about a year without relief of myoclonus. T advised surgery which was refused by pare has been no followup since then.

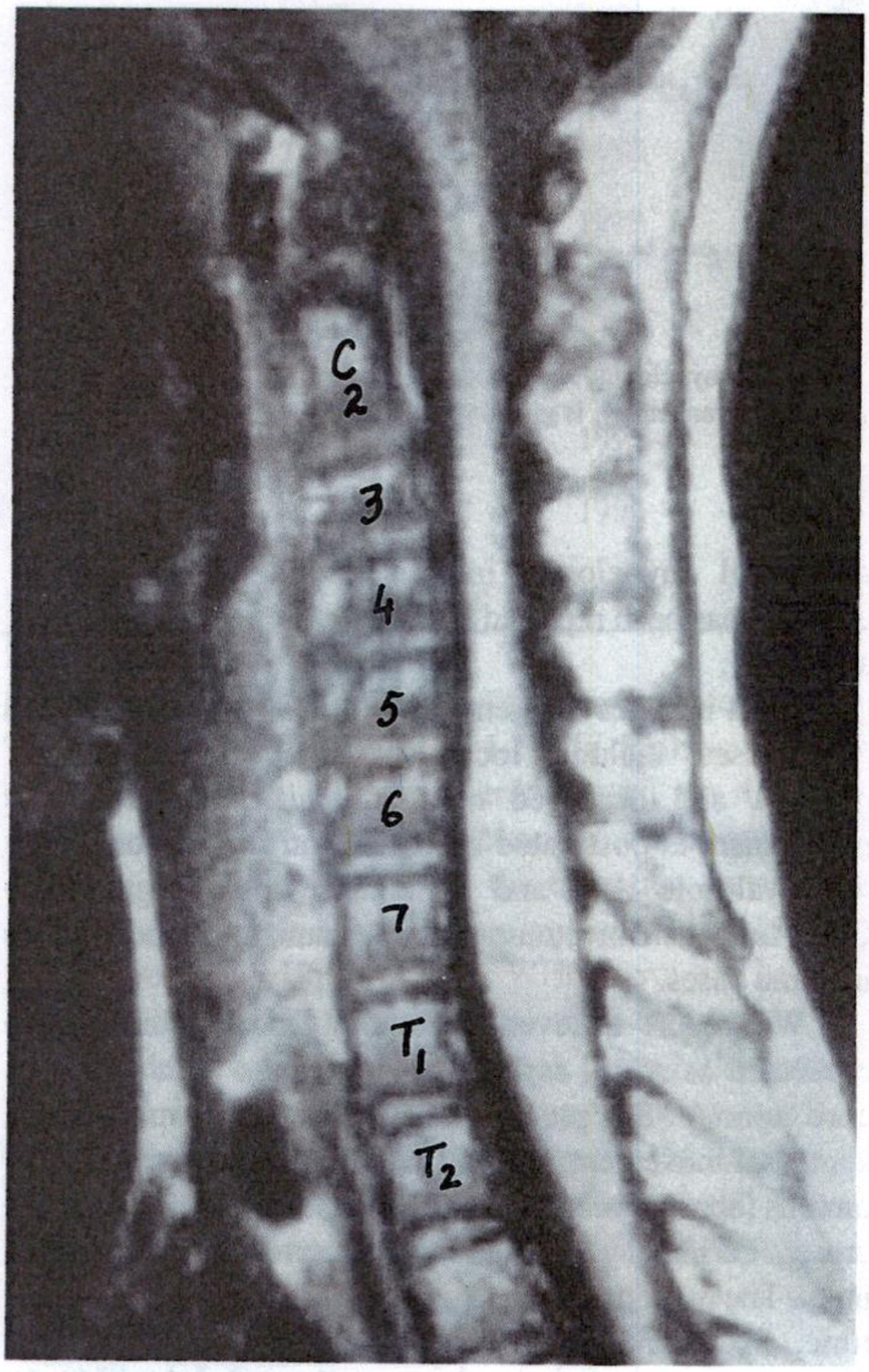


**Figure 1.** T1 MRI Scan showing spindle like enlargement of the cervical spinal cord from cervical; 5 to thoracic 1 segments with vertebral scalloping.

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