

Acute Asymmetrical Painless Upper Limb Flaccid Paralysis in a Child - Solving an Enigma- A Case Report

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Abstract

Objective: Child with acute asymmetrical painless upper limb flaccid paralysis secondary to anterior horn cell disease was treated successfully with steroids.

Material: 9 year girl presented with weakness and tingling sensation of both upper limb since 3 weeks. Weakness of both upper limbs, left more than right was noted with inability to comb hair and difficulty in writing. On examination, there was extensor weakness of both upper limbs (distal more than proximal) with no muscle wasting with diminished upper limb reflexes. Differentials like non-polio like illness, anterior horn cell disease, brachial plexitis, traumatic neuritis and Guillain-Barre Syndrome were considered.

Methods: CT cervical spine shows hypo dense areas in spinal cord from C4 to C6. MRI spine showed hyper intensities in cervical cord in the region of anterior horn cell disease extending from C4 to C6 vertebral bodies. Nerve conduction study and Electromyography were normal. Stool culture for polio virus was negative. CPK and CSF were normal. CSF culture was negative.

Results: She received oral steroids for 3 weeks with physiotherapy and occupational therapy. Weakness of upper limb was significantly improved over 8 weeks and she was completely recovered over next 3 months.

Conclusion: The anterior horn of spinal cord contains motor neurons which primarily affect axial muscles. The illness can be most likely caused by viruses like West Nile Viruses, Enteroviruses, Epstein Barr and Herpes simplex viruses. Anterior horn cell disease should be suspected in patients who have acute, painless, asymmetric weakness, even if unaccompanied by fever, meningoencephalitis, sensory loss or headache with characteristic neuroimaging findings.

Keywords: *Motor Neuron Disease; Anterior Horn Cell Disease; Snake Eye Sign; Acute Flaccid Paralysis*

Objective

Child with acute asymmetrical painless upper limb flaccid paralysis secondary to anterior horn cell disease was treated successfully with steroids, physiotherapy and occupational therapy.

Material

9 year old girl of Indian origin born of nonconsanguineous marriage presented with gradual onset of weakness and tingling sensation of both upper limb and neck since 3 weeks prior to admission. Weakness of left upper limb was more than right side. There was inability to comb hair and difficulty in writing. On examination, extensor weakness of upper limb was more than flexor weakness with involvement of distal muscles more than proximal muscles. Though no apparent wasting was noted, her upper limb reflexes were diminished. Her pain, temperature, vibration and joint position sense was normal. Considering the presentation and history differentials like non-polio like illness, anterior horn cell disease, brachial plexitis, and Guillain - Barre Syndrome were considered.

Methods

Her Full blood count, CPK, Liver and renal profile was normal. Her cerebral spinal fluid for sugar, protein was normal and culture was negative. Her CT cervical spine shows hypo dense areas in cord from C4 to C6. Nerve conduction study was normal. Stool culture for polio virus was negative.

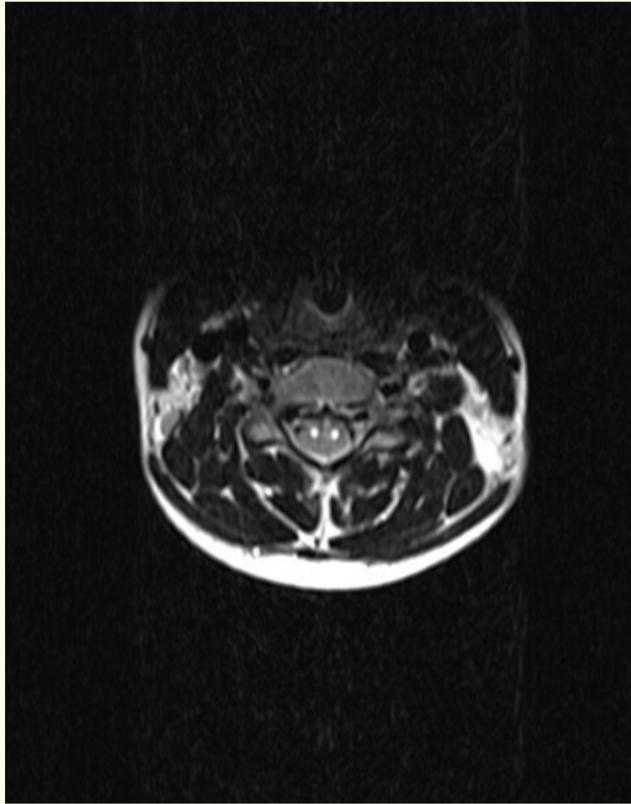


Figure 1: MRI spine Axial T1W image showed hyper intense lesion in cervical cord appearance typically resembling “Snake eye sign” (two small hyper intensities - one in each half of the cord on axial MRI images in the background of normal gray spinal cord) in the region of anterior horn cell extending from C4 to C6 vertebral bodies.



Figure 2: MRI spine T2 FLAIR confirmed the above finding. Whole spine screening with T2W no significant abnormalities.

Results

She received oral Prednisolone 1 mg/kg/day for 14 days followed by tapering of it over next 2 weeks. She was given physiotherapy and occupational therapy. Weakness of upper limb was significantly improved over 6 - 8 weeks. On follow up after 3 months she was able to comb her hair independently and was able to write. No further focal neurological deficit was noted.

Discussion

Motor neuron diseases (MND) are heterogeneous group of neurological disorders which are characterised by progressive degeneration of motor neurons. These disorders are usually uncommon with an incidence of approximately 2 cases per 100,000 populations [1]. The diseases like amyotrophic lateral sclerosis, Spinal muscular atrophy non-Polio like illness, progressive muscular atrophy, Charcot-Marie-Tooth disease are some examples of it. Clinical picture depends upon the type of the disease.

Etiopathogenesis is thought to be multifactorial. The presentation can be triggered due to various interactions between genetic and environmental factors [2]. The anterior horn cell disease can be most likely caused by viruses like West Nile Viruses, Enteroviruses, Epstein Barr and Herpes simplex viruses.

Anterior horn cell disease is a type of MND primarily affecting axons of the anterior horn of spinal cord. There are degenerative changes in the affected motor neurons namely anterior horn cells of the spinal cord [3]. Since the axons of the ventral portion of the spinal cord are concerned with motor function of the skeletal muscle first symptom of this disease usually is weakness of the corresponding muscles [4]. The disease affecting anterior horn cells of the spinal cord may result in highly selective weakness of the group of muscles. Symptoms vary depending upon the level of involvement of anterior horn cell.

The diagnosis is based upon clinical features and characteristic imaging findings. Nerve conduction velocity is usually normal as the pathological process involves axons and myelin is mostly unaffected.

Neuroimaging plays important part in making diagnosis of the patients and may show T2 hyper intensity in the area of anterior horn cells of the spinal cord – “Snake eye appearance” (two small hyper intensities - one in each half of the cord on axial MRI images in the background of normal gray spinal cord) is not exclusive to anterior horn cell disease and may also be seen in resolved cord contusion, Hopkins syndrome and radiation myelopathy But considering the typical clinical presentation with the particular appearance of spinal cord on axial images comprises the diagnosis of anterior horn cell disease [5].

Treatment usually consists of steroids, physiotherapy and occupational therapy. No definitive treatment is available for these disorders. Recent treatment modalities which are being investigated are stem cell transplantation and gene therapy although many questions remain about the utility of these recent therapies.

Conclusion

With eradication of poliomyelitis from many parts of the World the cases of non-polio acute flaccid paralysis are being increasingly identified and reported. Anterior horn cell disease is one such cause of acute flaccid paralysis.

It should be strongly considered in children who present with acute onset of muscle weakness, even if unaccompanied by fever, meningoencephalitis, sensory loss or headache. Characteristic imaging features on MRI typically resembling “Snake eye sign” in the region of anterior horn cell usually confirms the diagnosis.

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