ABSTRACT

A 5-year-old female presented with acute tetraparesis and areflexia. Initial imaging and cerebrospinal fluid analysis were suggestive of acute disseminated encephalomyelitis (ADEM). Minimal clinical response with intravenous steroids prompted further work up. Nerve conduction studies suggested acute motor sensory axonal neuropathy, a rare variant of Guillain-Barré syndrome (GBS). Repeat imaging was compatible with demyelination of the peripheral nervous system indicating concomitance of ADEM and GBS. The patient suffered severe motor deficits and neurogenic pain without bowel and bladder dysfunction. Slow but significant functional recovery was noted after intensive inpatient rehabilitation followed by continued rehabilitation via home health services.

CASE DESCRIPTION

The patient is a 5 year old female presented to our acute care hospital with sudden onset of weakness in extremities 1 week after a middle ear infection. Neurological exam at admission revealed decreased areflexia, right eyelid ptosis, left lateral nystagmus, dysarthria in lower extremities, facioid tetraparesis affecting more prominently bilateral lower extremities and areflexia. Bowel and bladder functions were noted to be intact. At baseline, the patient had achieved normal developmental milestones at appropriate ages.

Acute care Hospital Course

Cerebrospinal fluid analysis on day 1 revealed leukocytosis with a WBC of 74µl, elevated protein level (117 mg/dl), and elevated myelin basic protein level (10 ng/ml). MRI of the brain on day 2 showed (Image 1A) increased signal on T2 weighted images within the thalamus and posterior aspect of the pons. MRI of the spine (Image 1B) indicated mild to moderate swelling of the spinal cord with intrinsic T2 hypointense signal from C1 through the conus medullaris. These findings were consistent with ADEM. She was treated with 5 days of IV steroids which was started on day 3. Minimal clinical response to IV steroids prompted treatment with additional 5 days of intravenous immunoglobulin. Subsequently, nerve conduction studies (NCS) performed on left upper extremity on day 18 due to persistent areflexia showed decreased amplitude with normal latency. These findings were suggestive of acute motor sensory axonal neuropathy, a rare variant of GBS. Repeat MRI of the brain on Day 21 indicated interval resolution of the brain parenchymal lesions, since the prior studies and enhancement of the 3rd cranial nerve in the post contrast series. MRI of the spine (Image 2) showed improvement in previously seen spinal cord swelling and enhancement of multiple cervical and thoracic spinal nerve roots indicating the cauda equina. These findings along with NCS findings were compatible with a diagnosis of GBS suggesting concomitance of ADEM and GBS. The patient was discharged to our inpatient rehabilitation facility on day 22 from acute care hospital.

Inpatient Rehabilitation Facility Course

On admission to the inpatient rehabilitation facility, she exhibited severe neurogenic pain in extremities and persistent tetraparesis. She was continent of bowel and bladder. During a 27 day inpatient rehabilitation stay, the patient received intensive physical and occupational therapy to maximize functional recovery. Gabapentin was started for neuropathic pain with gradual titration of dose. At discharge, significant improvement was noted in her functional status (see table 1). She was discharged to home with continued rehabilitation services via home health.

OUTCOMES

Day 21 (Initial assessment)

Ceri: able to sit in supine position with excellent effort after 2 mod supervision.

Day 22 (IVF assessment)

Ceri: able to sit independently and ambulate 10 ft (transfer chair) with excellent effort.

Day 48 (IVF discharge assessment)

Ceri: able to sit independently and ambulate 125 ft (transfer chair) with excellent effort.

Day 105 (Follow up)

Ceri: able to sit independently and ambulate 125 ft (transfer chair) with excellent effort.

CONCLUSION

Limited evidence in the literature reports prolonged and incomplete recovery. Early diagnosis of this combined condition is important to initiate early aggressive medical interventions and early involvement of rehabilitation services which may expedite the recovery process resulting in better functional outcomes. Multidisciplinary team work, early and intensive inpatient rehabilitation, and continuum of care appear to be critical in such cases.

REFERENCES


DISCUSSION

ADEM and GBS typically occurs in the pediatric population after a viral-induced autoimmune response. The combination of these two diseases causes broad spectrum of neuromuscular abnormalities.2-6 Current scant literature reports persistent residual deficits during a follow up period ranging from 2 to 12 months in combined GBS and ADEM or transverse myelitis.7 In these retrospective studies, overall poor prognosis was reported in combined ADEM and GBS during follow up compared to isolated ADEM or GBS.8,9

Image 1: MRI on Day 2

1A. Brain MRI T2 weighted image of the brain shows patchy increased signal within both thalamus. No abnormal enhancement was present.

1B. Cervical spine MRI Sagittal T2 STIR sequence shows mild spinal cord swelling and hazy diffuse increased signal.

Image 2: MRI spine Day 21

2A. L spine MRI Sagittal T1 weighted post contrast image with fat saturation. Extensive enhancement and thickening of the nerve roots of the cauda equina.

2B. Axial T1 weighted post-contrast image shows that there is enhancement of cervical nerve roots.

Follow up

On recent follow up 6 months after discharge from inpatient rehabilitation, the patient was able to ambulate with bilateral ankle foot orthosis without the use of an assistive device.