Case Report

Inflammatory multifocal encephalopolyradiculopathy : a case report

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We report a case of inflammatory disorder involving brain, cranial nerve and peipheral nerves following an attack of postviral meningo-encephalitis. Few cases has been reported involving inflammatory disorders affecting extensive nervous system sites like brain, cranial nerves, spinal cord and peripheral nerves and such presentation is usually associated with features indicative of malignancy or granulamatous lesions[1-3]. Here, we report a case of quadriplegia following an attack of viral meningo-enchephalitis with mixed presentation of spastic upper motor neuron lesion (affecting the brain and cranial nerve) and lower motor neuron lesions involving the peripheral nerves on the extremities.

Case report

A 11-year-old boy presented to us on July 2010 with complaints of weakness all four limbs following an attack of fever and loss of consciousness of one week duration, a month ago. He was treated as a case of meningo-encephalitis in Paediatric ward and discharged after 3 weeks. On examination, higher mental function (HMF) was affected (irritable, orientation to time, place and person affected, follows verbal command coma scale: M3E3V2). Speech aphasia (motor), Cranial nerves: Occulomotor nerve affected on left side. Motor system: Tone decreased on upper limb and left lower limb; DTR ++ on right biceps, triceps, supinator, knee and ankle, reduced on left knee, biceps, triceps, supinator, but not illicitable on left ankle. Plantar reflex: flexor on right and non-responsive on left side with no ankle clonus. Sensory system appears intact (though difficult to test in view of aphasia and HMF status). Spasticity modified Ashworth scale grade I to II present on right upper and lower limbs.

Bladder voluntary voiding with incontinence at times. He was managed with the provisional diagnosis of postmeningo-encephalitis sequelae (quadriparesis) with exercises to maintain range of motion of the upper and lower limbs, stretching exercises, regular change of posture to avoid pressure sore over and above to improve nutritional status.

He was readmitted in January 2011 for further evaluation, upright standing and gait training. Muscle wasting on left calf and thigh was present. Muscle tone on right lower limb was increased while on left lower limb hypotonia was observed. Muscle power on right upper limb and lower limb was 4/5 except for Knee extensor 3/5; that of left upper limb was 4/5 while that of left lower limb was 2/5 .Muscle stretch reflex ++ on right upper and lower limb; (-) on left ankle. Right plantar reflex is Babinski positive while left showed Babinski negative. There was also associated ill sustained clonus on right ankle. Spasticity of grade II in hip adductor, hamstring and gastrosoleus on right side. Thomas test positive at 30° on right side. Right hip abduction range was 40/60° (when hip and knee are kept in flexed position) but hip abduction range was 30/40° when measured with hip and knee extension compared with left side. Hip external rotation was full and internal rotation 30/45°

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Received on 01/08/2011, Revised on 21/08/2011,
Accepted on 22/08/2011

compared with left side. There was pelvic obliquity on physical examination with no limb length discrepancy. On radiological examination of pelvis (anteroposterior view), there was 33.3% migration index on right hip as compared to 17% on the left hip joint suggesting Ist degree of hip subluxation on right side[4]. Electrophysiological studies (6 months after attack) showed features of peripheral neuropathy changes on both upper and lower limbs peripheral nerves with more affection on left lower limb i.e lateral popliteal and posterior tibial nerves. In view of bizzaire neuological presentation, a search for viral antibodies for cytomegalovirus, human immunodeficiency virus, Ebstien –Barr virus were made with all serological reports showed negative. We put the patient on KAFO with ischial socket extension to compensate weak hip extensor on left side for upright standing and gait training inside parellel bar. Adductor tenotomy (bilateral), Soutter Yount's release, fractional lengthening of hamstring and posterior tibial neurectomy on right side were performed to manage spasticity and grade I hip subluxation on right side. The patient was ambulatory with left Knee Ankle Foot orthosis (KAFO) at present.

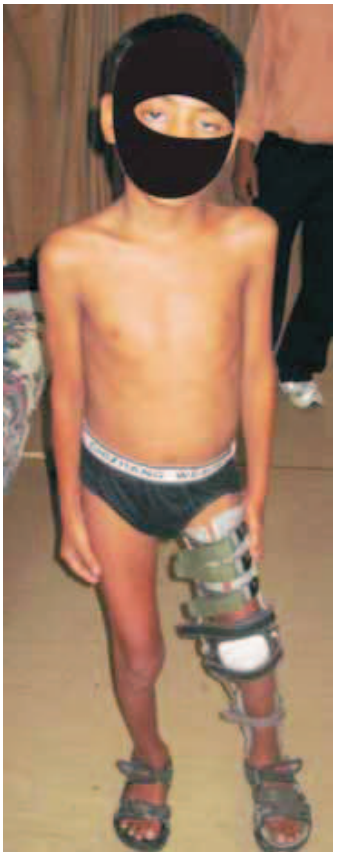
Discussion

Sandroni *et al*[1] reported a case of subacute paraparesis, diplopia and perioral paresthesia after an attack of viral illness with probable diagnosis of acute inflammatory demyelinating polyradiculopathy. The patient rapidly progressed to paraplegia with T10 sensory loss with asymmetric upper extrimity weakness along with facial weakness, bilateral deafness and visual impairement. Extensive diagnostic tests revealed no evidence of infective, neoplastic or paraneoplastic causes with special emphasis for vasculitis, paraneoplastic diseases, cytomegalovirus, human immunodeficiency virus, Ebstein-Barr virus, immunoglobin M, West Nile virus and other viral diseases. All viral serologies were negative. Electromyography and conduction studies showed absent or delayed F-waves with increase blink reflex R1 latency suggesting a polyradiculopathy. Cerebrospinal fluid examination showed mild elevation of protien but no pleocytosis. MRI showed marked enhancement of cauda equina, thoracolumbar nerve roots, multiple cranial nerves and leptomeninges. She was managed with intravenous immunoglobulin and high dose intravenous corticosteroid therapy and showed improvement of craniobulbar symptoms by 4 months and remarkable recovery of neurological deficits occurred. After one year her symptoms were localised to left foot weakness and paresthesia, mild difficulty in emptying bladder and minor visual symptoms.

The present case also presented with quadriplegia and coma following a febrile viral illness. He was managed as postencephalitis sequelae with weakness of all four limbs with involvement of superior rectus palsy (ptosis) on left side. On follow up, the upper motor spasticity was observed on right lower limb with spasticity on hip adductor, hamstring and gastrosoleus with ileo-psoas spasticity and lower motor weakness on left lower limb with quadiceps power grade 2/5. Hip subluxation was confirmed on right side on radiological examination following work up for pelvic obliquity with migration index on right hip 33%. Soft tisuue surgery as mentioned above along with tibial neurectomy was performed and patient is ambulatory with left KAFO to support lower motor weakness of quadriceps.

Several studies confirmed that the incidence of hip displacement in children with cerebral palsy is approximately 33% and is directly related to gross motor function classification system (GMFCS), but not related to the type of movement disorder.[5,6] Scrutton *et al*[7] also confirmed the high incidence of hip displacement in children with cerebral palsy. The authors have emphasised the need for regular clinical and radiological hip surveillance. Hip surgery in patients with cerebral palsy has been simply classified as preventative, reconstructive or salvage[8]. Preventative surgery refers to procedures pertaining to adductor releases which are designed to prevent progressive hip displacement in children who show early signs of hip displacement on hip radiographs. When preventative surgery fails or children present with very displaced or dislocated hips, reconstructive surgery may be employed. Turker *et al*[9] also reported adductor tenotomy to be effective in management of hip subluxation in spastic cerebral palsy in long term follow up.

Early intervention can be effective in management of hip dislocation and clinically, hip at risk for progressive subluxation are hips with flexion contracture of more than 20 degrees and abduction less than 30 degrees[10]. The three parameters considered for adductor tenotomy



for management of hip subluxation are: i) hip flexion contracture more than 20 degrees[10], ii) hip abduction range less than 30 degrees[10] and iii) (radiologically) hip migration index more than 25 degrees[4]. The above three parameters are for typical spastic cerebral palsy children. The present case reported herein is not a typical spastic cerebral palsy (as the timing for insult to brain occurred at the age of 10 years, but presented with an already subluxed hip on the right side due to adductor spasticity). Moreover, flexion deformity of hip on right side was 30 degrees (more than 20 degrees) and hip migration index on right side was 33% (more than 25 %) , which are significantly higher than the cut off values mentioned above. Due importance was given to these two factors while considering for abductor tenotomy on right side while accepting the fact that hip abduction range was more than 30 degrees on right side (i.e. 40 degrees).

Hence, the adductor tenotomy on right side was performed to prevent further progression of the already subluxed right hip. And, we are following up the case for any improvement or deterioration of the hip migration. In regards to the left adductor tenotomy, it was performed to release the adductor tightness / contracture secondary to positioning and lack of exercise compliance.

The present case with spastic subluxation of the right hip was propmtly managed with adductor tenotomy and is being followed up.

Conclusion

The evolution of extensive neurological involvement indicated a fulminant process affecting brain, cranial nerves and peripheral nerves, which was unlikely of an acute inflammatory demyelinating polyradiculopathy. The presentation of upper motor neuron and lower motor neuron features cannot be explained by single granulomatous or malignant process which are also most unlikely in regards to the age of the patient as in this case. The case has been highlighted in view of the extensive neurological affection of both upper and lower motor neuron paralysis including the cranial nerve affection. And also to highlight the importance of hip surveillance and timely management of hip at risk in spastic children.

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