Mononeuritis Multiplex Complicating Systemic Lupus Erythematosus

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Systemic Lupus Erythematosus (SLE) may have different neurological manifestations. Mononerits multiplex is the most common type of peripheral nervous system involvement in adult population, but case reports in pediatric population are sparse. We are reporting a case of pediatric SLE, presenting with polyarthritis and subsequently developing mononeuritis multiplex, identified by NCV.

Key words: *Mononeuritis multiplex, Nerve conduction velocity test, Systemic lupus erythematosus*.

SLE is a rare autoimmune disease in pediatric population. Prevalence of neuro-psychiatric manifestation in pediatric population is 28% [1]. Although isolated involvement of peripheral nervous system is very rare [2,3]. We are reporting a case presenting with fever and arthropathy where neuropathy was identified later and SLE confirmed as the etiology.

CASE REPORT

An 11-year old girl presented with fever and polyarthritis, involving right ankle, knee, elbow, wrist, metacarpophalangial and left knee joint for 10 days. The patient was nonambulatory due to severe pain and swelling in the joints**.** There was no significant past history and no weakness in the limbs. On examination pulse rate was 110/min, BP 102/84mmHg, temperature 101ºF. Respiratory, cardiovascular and nervous system examination was normal. Skin rash was absent. Investigations revealed hemoglobin 7.5gm%, MCV 81.7, MCH 27.0, MCHC 33.1, total leukocyte 9900 (neutrophil 70%, lymphocyte 28%, eosinophil 2%), without any abnormal cell. ESR was 122mm in 1st hour and CRP 0.6 mg/dL. Blood sugar, serum urea, creatinine and LFT were normal. ASO titer was 1:160. Blood for Dengue IgM was negative. Blood culture showed no growth. Routine urine examination was normal and culture revealed no growth. ECG showed sinus tachycardia. Chest *X*-ray was normal. *X*-ray of all involved joints was normal.

After exclusion of common causes of fever with polyarthritis, *i.e.* Rheumatic fever, Rheumtoid arthritis, Leukemia, Dengue etc. we thought of possibility of connective tissue disorder like SLE as there was fever with nonerosive polyarthritis with involvement of small joints with anemia and raised ESR and normal CRP. We tested for Anti nuclear antibody (ANA) which was positive in 1:640 dilutions with a characteristic homogeneous & speckeled pattern. Anti ds DNA was positive in 1:10 dilution. Antiphospholipid antibody, both IgG and IgM were positive. Oral prednisolone in a dose of 2mg/kg/day was started. 10 days later fever subsided completely and arthritis diminished. That time when the patient became ambulatory we noticed that the girl was walking with a high stepping gait with toe walking in right side. Power of flexors and extensors of right ankle joint was diminished with hypotonia. Ankle and knee jerks were diminished and planter response was absent in right side. There was sensory loss in the lateral aspect of right foot and leg. Atrophy was absent. Left lower limb and both upper limb examination were normal. Sensorium and cranial nerves were not involved. Nerve conduction velocity test showed: Right Common Peroneal Nerve (CPN) & Posterior Tibial Nerve (PTN) were inexcitable, there was distal latency in left PTN, left CPN showed decreased amplitude with normal conduction velocity and right median nerve showed decreased amplitude with normal conduction velocity. In presence of involvement of more than 2 nerves in different site in different phases of evolution diagnosis of mononeuritis multiplex was confirmed. Nerve biopsy from right sural nerve showed features of peripheral neuropathy, without any vasculitis in epineural vessels and there was demyelination of nerve fibers. One month after starting treatment prednisolone gradually tapered and now patient is on 10 mg prednisolone per day. In last 6 months follow up patient developed atrophy in right calf muscle and there is no further deterioration of neurological function (clinically and in repeat NCV testing) or involvement of any other system.

DISCUSSION

American College of Rheumatology (ACR) subcommittee in 1999 proposed 19 neuropsychyatric syndromes to be present in SLE, 7 of these involve peripheral nervous system [4]. Neuropathy associated with SLE is the least described entity in pediatric population [2,3]. Previous

case reports mentioned involvement of both sensory and motor nerves after some interval of time from the diagnosis. They were treated with multiple drugs [2,3]. To the best of our knowledge, presence of mononeuritis multiplex at the time of initial diagnosis of pediatric SLE has never been reported before. Involvement of peripheral nervous system in SLE may be in the three forms. Mononeuritis multiplex is the most common presentation. Acute and chronic inflammatory demyelinating neuropathy and symmetrical distal sensory motor neuropathy are other presentations. Mononeuritis multiplex develops due to vasculitic insult to vasa-nervosum. There is Wallerian degeneration of nerve fibers secondary to ischemic infarction due to occlusion of blood vessels caused by leukocytoclastic vasculitis [5]. A positive association of antiphospholipid antibody with possibility of development of this type of vasculitic neuropathy is suggested [3]. Bodi, *et al.* [6] in their study on sural nerve biopsy in SLE neuropathy patient, found that endoneurial immune complex deposition also plays an important role in the demyelinating process and axonal damage seen in peripheral neuropathy. In this child, presence of neuropathy was confirmed clinically, and by NCV testing and nerve biopsy; although vasculitis of vasa nervosa, a common finding in nerve biopsy, was absent. This may be a result of poor preservation of specimen during transportation. Neuropathy may develop at any time during the course of disease. Different scoring systems like Neurologic symptom score, Neurologic disability score are used to study neuropathic involvement in SLE [7]. Neuropathy may present in SLE without any symptom. As in our case, there was only right lower limb symptomatology, but significant axonopathy in right median nerve was detected by NCV testing. Neuropathy in SLE needs early aggressive treatment. In this regard, NCV may be considered as a screenig tool for early detection of neuropathy, altough more studies are needed prior to adopting routine NCV in all cases of SLE. Features of axonopathy, i.e. decreased amplitude with normal conduction velocity is most commonly found in NCV [7].

There is no consensus guideline for treatment of neuropathy associated with pediatric SLE. Steroid is effective in most of the situation. Gabapentin, carbamazepine, azathioprine, cyclophosphmide etc. are used [3]. Pulse cyclophosphamide therapy is mentioned to be used with good result in adult patient [8]. Because of good response with steroid alone in our patient, cyclophosphamide was not used.

To conclude, high index of suspicion should be there for development of neuropathy in pediatric SLE patient, even when no other manifestation of SLE is present. Aggressive treatment should be instituted using steroid, with or without immunosuppressive agent.

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