Peripheral nerve disorders I

PP1247

POEMS syndrome – case report of rare clinical presentation with pathological spinal fracture

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Background and purpose: POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammapathy and skin changes) is a rare multisystemic disease, which belongs to plasma cell dyscrasia (monoclonal plasma cell proliferative disorder, almost always λ). Bone fractures in patients with POEMS syndrome are rarely described in literature. They are caused by bone lesions, which can be osteosclerotic, osteolytic with sclerotic shaft or “soap bubble” vision lesions. The aim of this article is to present a rare clinical presentation of POEMS syndrome in a woman with spinal fracture, probably as manifestation of osteosclerotic myeloma.

Case report: We have presented a rare clinical case of a female patient with pathological fracture of the thoracic vertebra. Our patient has developed spontaneous asymptomatic vertebral fracture that was initially misdiagnosed as hemangioma, but later histological result has lead us to the possibility of plasmacytoma. Serum protein electrophoresis and immunofixation revealed evidences of monoclonal gammapathy (λ light chains).

Conclusions: In patients with chronic and progressive demyelinating polyneuropathy of unknown origin, with systemic manifestation of organs, skin and endocrine glands one should always be thinking of monoclonal plasmacytoma. We report a patient with POEMS syndrome with an extremely rare clinical manifestation.

Disclosure: Nothing to disclose

PP1248

Neurogenic claudication-mimic due to severe aortoiliac disease

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Introduction: Symptoms arising from lumbar stenosis and arterial occlusive disease can demonstrate similarities that present a considerable diagnostic challenge. We experienced an atypical case of patient mimicking neurogenic claudication, whose symptom arose from severe aortoiliac occlusive disease.

Methods: Case report.

Results: A 48-year-old male had exertional bilateral pelvic pain and claudication for three months. He complained of severe aching and burning sensation around his buttock and thighs. After about 200~1,000 meter walking, he experienced pain in the gluteal region, followed by paresthesia and sensory loss. These sensory symptoms were not correlated with dermatome. Pulses of bilateral femoral, posterior tibial and dorsalis pedis arteries were weak. At rest, the right and left ankle/brachial indexes (ABI) were reduced. When he walked about 200m, bilateral dorsalis pedis pulses were not observed. After five minutes of rest, weak pulses of dorsalis pedis arteries reappeared. The CT angiography revealed long segmental chronic thrombotic occlusion involving the distal abdominal aorta and both common iliac arteries. After a successful aorto-bililac bypass operation, he could walk remarkably long distance without pain and sensory symptoms.

Conclusions: Many cases with claudication have typical manifestation of whether vascular or neurogenic type. But there exists a widely unknown third type of intermittent claudication, which causes leg pain with any muscular effort similar to the vascular type. In the future, by evaluating the clinical symptoms and physical examination in such cases, it can shed a light on the clinical marker of diagnosing neurogenic claudication mimic due to peripheral arterial disease.

Disclosure: In conclusion, we report a patient with peripheral arterial disease and neurogenic claudication mimicking symptom which resolved completely after a successful vascular surgery.
PP1249

The time course of pseudo-conduction blocks in a patient with vasculitic neuropathy

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Introduction: “Pseudo-conduction block” can be an early and warning electrophysiological finding of vasculitic neuropathies (VN). Here we report the time course of pseudo-conduction block in a VN patient.

Patient: 56-year-old female referred because of burning and weakness in hands and feet for two months. The neurological examination revealed asymmetric, multifocal motor weakness more prominent on the right hand with thenar atrophy. There was asymmetrical sensation loss in all modalities. Biceps, triceps reflexes were diminished whereas patella and Achilles reflexes were absent. Anti–nuclear antibody titer was 1/320, C-reactive protein and p-ANCA were highly positive. The cerebrospinal fluid examination was normal. First electrophysiological examination showed asymmetric axonal neuropathy. Also there were significant drops in the amplitude/area of compound muscle action potentials (CMAP) evoked by proximal electrical stimulation relative to distal stimulation at ulnar and tibial nerves. Initially these findings were suggestive for motor conduction blocks. We repeated electrophysiological examination during the following five days. Motor conduction blocks at these nerves were resolved, proximal and distal CMAP amplitudes became similarly low (Figure 1). Finally we decided these findings as pseudo-conduction blocks. The sural nerve biopsy showed perivascular inflammation and active axonal degeneration (Figure 2). Congo-red staining failed to demonstrate amyloid. Amyloid polyneuropathy was always suspected but the genetic analysis was not available at that time. The Val30Met mutation could be determined after 19 years when his son suffered from burning in the feet who is on tafamidis treatment now.

Conclusion: Although motor conduction blocks generally suggest acquired demyelinating neuropathies, it might be an earlier evidence of Wallerian degeneration. Serial nerve conduction studies are helpful to distinguish these two entities.

Disclosure: Nothing to disclose

PP1250

Three families with hereditary amyloid polyneuropathy with three different mutations

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Introduction: Transthyretin (TTR)-related hereditary amyloidosis is a rare autosomal dominant systemic disorder which can present with progressive, axonal sensory, autonomic or motor neuropathy called familial amyloidotic polyneuropathy. We report cases from three families with clinical and nerve biopsy findings.

Case presentations:
Case 1: 54-year-old man presented with numbness in his hands and feet for two months. The neurological examination revealed glove-stocking sensation loss and diminished Achilles reflexes. Routine laboratory tests were normal. Three months later, his examination showed bilateral weakness in lower extremities. Electrophysiological examination showed mixed neuropathy prominent in sensory nerves. The nerve biopsy revealed moderate fiber loss (Figure 1). Congo-red staining failed to demonstrate amyloid. Amyloid polyneuropathy was always suspected but the genetic analysis was not available at that time. The Val30Met mutation could be determined after 19 years when his son suffered from burning in the feet who is on tafamidis treatment now.
Case 2: 18-year-old female referred because of constipation, orthostatic hypotension. His father got the diagnosis of hereditary amyloidosis with Thr49Ser mutation on TTR gene due to autonomic symptoms and heart failure; died at the age of 38. Her neurological examination was normal except minimal vibration loss.
Case 3: 30-year-old female admitted with heart failure, progressive leg weakness, incontinence and orthostatic hypotension. She had 4 relatives including her sister with familial amyloidosis (Figure 2). The genetic analysis showed Glu54Lys mutation. She did not have any improvement despite three months tafamidis trial.

Conclusion: We present three families with variable clinical features due to three different mutations.

Disclosure: Nothing to disclose
PP1251

Small fiber neuropathy in patients with chronic graft-versus-host disease after allogeneic hematopoietic stem cell transplantation - preliminary results of a prospective study

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Introduction: Neurologic complications in chronic graft versus host disease (cGVHD) after hematopoietic stem cell transplantation (HSCT) may include peripheral neuropathy. A significant number of cGVHD patients experience painful muscle cramps and neuropathic pain (NeP). Small fiber neuropathy (SFN) may be the cause of NeP and it can be diagnosed with quantitative sensory testing (QST).

Methods: Ten patients with cGVHD were examined and QST was performed (Pathway CHEPS). The patients also filled the Pain Detect questionnaire with final goal to diagnose the NeP and to validate the NeP treatment.

Results: Six out of 10 patients reported pain and muscle cramps at the time of presentation and 4 of them met the criteria for NeP according to Pain Detect questionnaire. In all patients with NeP the QST disclosed affection of C and A-delta fibers (elevated threshold for pain, heat and cold sensation). In five non-NeP patients QST showed affection of only A-delta fibers.

Conclusion: Neuropathy in cGVHD may have various causes and patterns. SFN may be the cause of NeP. According to our preliminary results, it is possible that A-delta fibers in cGVHD are affected first, in the period before NeP, while the affection of C and A-delta fibers presents later, after patients develop NeP. Neurologic complications and pain in cGVHD may have major impact on the functional status, quality of life and long term outcomes of cGVHD patients. Early recognition and proper diagnostics of SFN and NeP may contribute to the better treatment of pain in cGVHD patients.

Disclosure: Nothing to disclose

PP1252

Severe motor neuropathy with monoclonal gammopathy and high titers of antibodies against GM1 and GD1b gangliosides: response to treatment

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Introduction: Neuropathy associated with monoclonal protein (MP) and antibodies against gangliosides (Ab-G) GM1 GD1b asialo GM1 present with severe motor neuropathy. We hereby report.

Methods: A 75-year-old male presented subacute diffuse motor neuropathy. Electrophysiological study (EDX) revealed a mixed pattern. Nerve biopsy disclosed rare signs of demyelination. IgIV and steroids were inefficient. Plasma exchanges (PE) stabilized temporarily symptoms. Immunologic testing revealed an IgM lambda MP with monoclonal expansion in the blood and bone marrow, and positive Ab-G. In spite of treatment associating Cyclophosphamide, Fludarabine and Rituximab, the patient developed tetraplegia and died. A 67-year-old male presented chronic multifocal motor neuropathy. EDX revealed demyelinating signs without conduction blocks. Protein level was 0.91g/L in CSF. Nerve biopsy disclosed rare signs of demyelination. IgIV were inefficient. PE and steroids allowed improvement with relapse at weaning. Immunological testing revealed an IgG kappa MP and Ab-G. After 6 pulses of cyclophosphamide a severe relapse occurred with tetraplegia and respiratory failure, leading the patient to ICU. Treatment associating Rituximab and PE improved the patient’s condition 4 months later regaining walking with aid and no respiratory assistance.

Results: In both cases presentation was that of a disabling motor neuropathy with MP and similar Ab-G profile. Disease resisted to different treatments, chemotherapy was undertaken because of the severity of neuropathy and hypothesis of an immunological or infiltrative mechanism.

Conclusions: In the setting of severe motor neuropathy, testing for MP and Ab-G is important to adapt specific treatment and reverse dramatic course.

Disclosure: Nothing to disclose
PP1253
Deletion of ADAM10 in axons impairs axonal outgrowth and remyelination in the PNS

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Introduction: An emerging body of evidence suggests that disintegrin and metalloproteinases (ADAMs) play an essential role in primary development and myelination of the peripheral nervous system (PNS). ADAM10 is a membrane-anchored metalloproteinase with both proteolytic and disintegrin characteristics. Previous in vitro studies propose that during the process of myelin formation, ADAM10 is highly upregulated and appears to be critically involved in axonal outgrowth that is a requirement for myelination in the PNS.

Methods: To further address the importance of ADAM10 in these processes in vivo we generated a cell specific Cre-loxP-mediated knockout model of ADAM10 either in myelin-forming Schwann cells (P0-cre, ADAM10-/-) or in motor neurons (Mnx-cre,ADAM10-/-). We performed a sciatic nerve crush, which is widely accepted as a valid model for peripheral nerve regeneration. To determine the level of clinical impairment continuous clinical and electrophysiological examination was conducted. To quantify the degree of post-traumatic axonal degeneration and remyelination semi-thin sections were generated.

Results: Under physiological conditions P0-cre, ADAM10-/- as well as Mnx- cre, ADAM10-/- did not present with any clinical, electrophysiological or histological phenotype when compared to wildtype mice. However, traumatic peripheral nerve lesion by sciatic nerve crush revealed a significant reduction of nerve fibres, especially of small calibre fibres in Mnx- cre, ADAM10-/- mice.

Conclusion: Our data suggest that axonal ADAM10 is important for neuronal outgrowth and thus represents a prerequisite for successful regeneration and consecutively remyelination. ADAM10 might play a crucial role in the interaction between axons and Schwann cells during the process of remyelination.

Disclosure: Nothing to disclose

PP1254
Efficacy and tolerability of different brand of IVIg in the maintenance treatment of chronic immune mediated neuropathy

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Introduction: Treatment with high dose intravenous immunoglobulin (IVIg) is effective in chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) and multifocal motor neuropathy (MMN). Not all brand of IVIg are however licensed for their use in these neuropathies. We now analysed the efficacy and tolerability of different brand of IVIg in the maintenance treatment of CIDP and MMN.

Methods: We reviewed the reports of six patients with CIDP and seven with MMN treated with IVIg from 2009 to 2013. Two patients with CIDP and two with MMN started ex novo their treatment while 9 continued the treatment initiated 2-169 months before (mean 45). In all patients we measured the MRC score in the six most affected muscles before each infusion, the monthly dose and brand of IVIg and adverse events.

Results: Patients were treated with IVIg for 25-60 months (mean 48) with a monthly dose of 70 g (range 20-160g including starting dose). Patients were treated with IgVena, Gammagard, Kiovig and Flebogamma for a variable period of time. Minor and transient side effects were equally observed with each therapy. Two MMN e two CIDP patients increased the monthly dose for disease progression but this was not related to the change in IVIg. No variation in the MRC sumscore and in IVIg dose was observed in the other patients despite the change of IVIg.

Conclusions: Chronic maintenance treatment with IVIg in our patients with MMN and CIDP was not associated with a different tolerability or efficacy despite the use of different brands of IVIg.

Disclosure: Nothing to disclose
PP1255

Acute intermittent porphyria (AIP) presenting as acute neuropathy: a laboratory and genetically confirmed observation

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Introduction: AIP is an inherited metabolic disorder and one of the main differential diagnosis for acute motor neuropathy.

Methods: Case report.

Results: A 32-years old previously healthy female patient presented with recurrent heavy abdominal pain. Antibiotics and analgesics had been given without success. The patient lost weight and developed general weakness. The patient presented with progressive proximal symmetrical flaccid tetraparesis (BMRC 3 prox., 4 dist.), normal DTR, no sensory loss. She reported general hyperalgesia and diffuse pain. Severe tachycardia was considered to be a sign of autonomic involvement. The tetraparesis progressed within 4 days (BMRC 1 prox., 2 dist.). The combination of abdominal symptoms and neuropathy suggested AIP, hematin therapy was initiated. Over 2 months the patient gradually recovered (BMRC 3 prox., 4 dist.).

Findings:
- 24-hour urine analysis: porphyrin (3,300 µg; <150µg), aminolevulinic acid-U (90mg/l; <10mg/l) and porphobilinogen (142mg/l; <1.7mg/24hr)
- Genetic examination: confirmation of AIP (mutation c.973C>T). Family history for AIP negative.
- NCV: mild axonal neuropathy with preserved sensory conduction velocities. F-waves preserved.
- SSEP, MEP, cerebral MRI: normal.

Conclusions: This is a case report of a family negative AIP presenting with an acute motor neuropathy and abdominal symptoms. Abdominal pain is often the initial symptom followed by symmetric or asymmetric neuropathy with proximally accentuated tetraparesis. Also a diffuse pain syndrom, autonomic symptoms, mental state changes and seizures are reported. The duration of an attack may be days to weeks, usually with a complete recovery.

Disclosure: Nothing to disclose

PP1256

Guillain-Barré syndrome associated with rapid immune reconstitution following autogeneic hematopoietic stem cell transplantation

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Introduction: A patient followed for Hodgkin disease (HD) presented a Guillain-Barré syndrome (GBS) after hematopoietic stem cell transplantation (HSCT). In this context, a post transplant immune reconstitution is discussed.

Observation: A 21-year-old man, followed for HD with recurrence at 18 months, enjoyed an autologous HSCT. After 57 days, the patient presented neurologic trouble with tetraparesia, abolition tendon reflexes and facial diplegia. Acute polyradiculoneuropathy was confirmed by electromyogram; cerebrospinal fluid (CSF) examination revealed normal cell count, elevated protein levels at 2.1 g/l. No cell lymphoma was found. No tumor or infectious argument was otherwise founded. Plasma exchange (PE) were made before a neurological worsening, complicated by acute respiratory distress syndrome and autonomic dysfunction with cardiac arrest recovered. Thereafter, 5 monthly courses of intravenous immunoglobulin (IVIG) were performed to clinical improvement. Unfortunately this development was hampered by mechanical respiratory complication that led to the death of the patient, the 237th day of evolution. The final diagnosis of GBS, occurred in a context of post transplant immune reconstitution in a patient treated for LH, was retained.

Discussion: With a frequency of less than 1%, GBS is a rare complication of HSCT. The literature review collects 31 cases, 10 occurred after autograft. The average installation time is 7 weeks, so that chemical toxicity can not be discussed. Etiopathogenic mechanisms remain obscure.

Conclusion: GBS is possible after HSCT indicated for the treatment of LH. Despite the use of immunomodulatory treatment, the prognosis is dark for 1/3 of cases.

Disclosure: Nothing to disclose
PP1257

Two cases with different involvement of hypoglossal nerve
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Introduction: Isolated hypoglossal nerve palsy is rare and may be due to an intracranial or extracranial space–occupying lesion, head and neck injury, carotid artery dissection, vascular abnormality, idiopathic, infection, autoimmune disease or neuropathy and vaccination. We report two cases of isolated unilateral XII th nerve palsy one of them cause proved to craniocervical trauma and other one due to local infection.

Methods:
Case 1: 54 years old woman had admitted with hypoglossal atrophie and paralysis. Cause of palsy appeared to be cervical trauma to hypogossal nerve. Her cranial MRI and other radiologic investigation were normal. Her lingual electromyographic investigation revealed acute, sub-acute partial axonal damage at the right side. Her findings were lost after 3 months.

Case 2: We report a 48-year-old woman who presented with headache spreading from servical to the head and dysphonia. The left side of the tongue was atrophic and deviated to the left side on protrusion and fasciculations were noted. Cranial and neck MRI revealed that a lesion at the inferior of canalis hypoglossus next to foramen jugularis. Concentric needle electromyography of the left side revealed fibrillation and positive sharp waves with decreased recruited motor units.

Results: All patients have family history, clinical history and electrophysiologic studies compatible with CMT. Five patients have a demyelinating pattern and the last 4 an axonal pattern. Most patients presented with previously unsuspected CMT (8 of 9). Five patients received vinca alcaloides and 4 taxanes. Five patients have a mild clinical improvement after treatment discontinuation.

Conclusions: In the hypoglossal palsy, all possible causes must be thought. Two cases prognosis were good which one of them using antibiotherapy, other one spontaneously.

Disclosure: Nothing to disclose

PP1258

Neurotoxic chemotherapy in Charcot-Marie-Tooth disease: a case series
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Introduction: Disabling peripheral neuropathy (PN) is a major dose-limiting side effect of many chemotherapeutic agents (CA). The main risk factors are the type of chemotherapy and the cumulative dose. Pre-existing neuropathy, especially Charcot Marie Tooth (CMT) neuropathy is a generally accepted risk factor and can determine early and severe chemotherapy-induced neuropathy. This study reports 9 patients with CMT, all treated for cancer and worsened after “safe dosages” of CA.

Methods: Review the last 15 years of patients with cancer and clinical and electric evidence of CMT in the neurological clinic of the Salpêtrière hospital. Nine patients have been collected and have received CA.

Results: All patients have family history, clinical history and electrophysiologic studies compatible with CMT. Five patients have a demyelinating pattern and the last 4 an axonal pattern. Most patients presented with previously unsuspected CMT (8 of 9). Five patients received vinca alcaloides and 4 taxanes. Five patients had a mild clinical improvement after treatment discontinuation.

Conclusions: PN is a major side effect in patients with cancer receiving CA. Pre-existing CMT neuropathy is often unknown. Regarding our case series, CMT with axonal neuropathy pattern is as risky as demyelinating one. Vinca alcaloides and Taxanes seem to be the most dangerous CA for deterioration or revealing CMT. Even if CMT is a quite rare condition, prior to use CA, it seems relevant to screen for family history or clinical signs of CMT and refer to a neurologist if necessary.

Disclosure: Nothing to disclose
PP1259

Report of two unusual cases of unilateral isolated hypoglossal nerve palsy

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Introduction: Isolated hypoglossal nerve palsy is a rarely seen condition. Here, we present two unusual cases of hypoglossal nerve palsies subsequent to dental complications.

Cases: Patients were female (21 and 35 years old respectively). They both had a history of difficulty in swallowing, disturbed speech and loss of volume on the right half-side of their tongues. The second patient had also diagnosis of bruxism and trigeminal neuralgia. Oral pantogram of the first patient showed a small exostosis on the apex of the right bottom impacted wisdom tooth with a surrounding radiolucency suggesting inflammation around it. Removal of the tooth resulted in the improvement of function of the hypoglossal nerve. The second patient had a history of full mouth dental reconstruction for the last 18 months. Oral pantogram showed radiolucency suggesting inflammation that lies under the right bottom dental bridge. Removal of the dental bridge resulted with improvement of both trigeminal neuralgia and function of the hypoglossal nerve.

Conclusion: Inflammation around the hypoglossal nerve or itself during its course from the angle of the mandible through the sublingual region may result with the injury of this cranial nerve. Therefore, the physicians coming across with isolated hypoglossal nerve palsy should consider the possible unusual dental causes as the differential diagnosis.

Disclosure: Nothing to disclose

PP1260

Significance of ultrasound muscle echointensity in diagnosis of carpal tunnel syndrome

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Introduction: High echointensity (EI) of muscles in ultrasonography (US) is suggested as evidence of secondary muscle damage resulting from neuropathy. We performed this study to investigate reliability of new ultrasonographic measurement to evaluate secondary muscle changes resulting from carpal tunnel syndrome (CTS).

Methods: Forty-five hands from 30 patients with CTS and 20 hands from 11 normal healthy subjects were recruited. The hands of patients were divided into three subgroups based on the Canterbury grading. Transverse US images of thenar and hypothenar muscles were obtained.

Results: The EI ratio was significantly higher in the patient group compared with the control group (p=0.044), whereas the inhomogeneity ratio was not significantly different. Comparison among the three subgroups of patients showed significant differences in both EI ratio (p=0.045) and inhomogeneity ratio (p=0.014). The presence of denervation potentials in EMG showed a very strong correlation with EI ratio (p=0.000).

Conclusions: The new ultrasonographic method to detect secondary muscle damage resulting from neuropathy was proven to be effective. Both EI and inhomogeneity ratios measured by US could be diagnostic parameters for CTS.

Disclosure: Nothing to disclose
PP1261

Ulnar nerve instability around the elbow in healthy subjects: ultrasonographic and electrophysiologic findings

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Introduction: Ulnar nerve (UN) instability during elbow flexion may be a potential cause of ulnar neuropathy, which might be related with snapping of medial triceps muscle. This study is designed to evaluate the relationship between ulnar nerve instability and snapping of the medial triceps muscle during elbow flexion using ultrasonography and nerve conduction study (NCS) of the UN.

Methods: Forty-two elbows of 21 healthy subjects were recruited. Dynamic ultrasonography was performed in three positions of the elbow: extension, 90 degree flexion, and full flexion. The horizontal distance from the apex of medial epicondyle (ME) to the margin of ulnar nerve and medial triceps muscle (ME_UN and ME_TB, respectively) were measured. The ulnar nerve instability was classified into three types according to the degree of movement: no dislocation, subluxation and dislocation. Ulnar NCS was done.

Results: In 90 degree elbow position, no dislocation and subluxation were 35 (83.3%) and 7 elbows (16.7%), and in full flexion, no dislocation, subluxation and dislocation, (47.6%), 17 (40.5%), and 5 elbows (11.9%). Pearson correlation coefficients between UN instability and snapping of medial triceps muscle in 90 degree and full flexion of elbow were 0.547 and 0.781 (p-value, <0.001), respectively.

Conclusions: Ulnar nerve instability is increased with elbow flexion, which might be related with the snapping of median triceps muscle. It is important to recognize ulnar nerve instability during elbow flexion as a potential cause of ulnar neuropathy, and also to perform the ulnar motor NCS considering the ulnar nerve instability.

Disclosure: This study was supported by a Korea University grant (K1326792).

PP1262

Case report: acute inflammatory demyelinating polyneuropathy with bilateral extensor plantar reflexes

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PP1263

A helping hand

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PP1264

Searching for an etiology for mononeuritis multiplex

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PP1265

A severe chronic form of (“A”)MSAN during pregnancy

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PP1266

Electrophysiological study of the sciatic nerve in spontaneously hypertensive rats: is there a gender difference?

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PP1267
Study of late responses' parameters in carpal tunnel syndrome
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PP1268
Livedoid vasculopathy associated with peripheral neuropathy successfully treated with warfarin
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PP1269
Guillain-Barré syndrome following acute brucellosis: a case report
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PP1270
Recurrent severe Guillain-Barré syndrome: a case report
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PP1271
Ulnar nerve entrapment neuropathy at the elbow: relationship between the electrophysiological findings and neuropathic pain
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PP1272
A case of rhabdomyolysis presenting as unilateral hemiplegia
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PP1273
Neuralgic amyotrophy: a specific cause of bilateral shoulder pain
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PP1274
Post-irradiation neuromyotonia of the hypoglossal and spinal accessory nerves
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