

Patients with Spinal Cord Damage due to Uncertain/Unknown Etiology: The CP Phenomenon (Confusion and Perplexity). Case Study

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*The Sutton Law: Always perform at the outset the diagnosis test
or therapeutic maneuver most likely to establish the diagnosis”
When Willy Sutton, [1901-80] American bank robber, was asked why
he robbed banks, he replied „because that’s where the money is”.
The term was proposed by Petersdorf & Beesen, Medicine, 1961*

SUMMARY

Usually, most of the clients who are referred to departments of rehabilitation medicine, bear firm and sound diagnoses. We describe herewith 10 patients who developed spinal cord pathologies due to unknown or uncertain etiologies. We would like to share our thoughts with the readers.

Key words: unknown etiology, rehabilitation medicine, spinal cord damage

BACKGROUND

The classical Classification Types of myelopathy is described as follows: [1]:

- a. Traumatic (penetrating [2-3], closed, surgical, manipulation)
- b. Non-traumatic [4].
 1. Motor Neuron Disease (ALS, SMA, others)
 2. Spondylotic myelopathy (spondylosis, spondylolisthesis, spinal stenosis, disc herniation, C 1/2 instability)
 3. Infections, inflammatory (Acute Transverse Myelitis, Multiple Sclerosis, poliomyelitis, osteomyelitis, arachnoiditis, HIV, Guillain-Barre' syndrome (GBS) [5-6], epidural abscess, Tuberculosis [7], Caisson disease)
 4. Neoplastic diseases
 5. Vascular (ischemic, A-V malformation, Aneurismal bone cyst [8-9])
 6. Toxic, metabolic (post-irradiation, vitamin B12 def.,)
 7. Congenital and developmental (myelodysplasia, syringomyelia [10])
- c. Non specified

Our aims in modern Rehabilitation Medicine are:

- Correct & accurate diagnosis
- Early referral
- Comprehensive-integrative-multidisciplinary program (skilled team, modern equipment)
- Long term follow-up
- Long- term planning
- Re-integration into society & into the mainstream of life
- Functional restoration even without known etiology
- Successful rehabilitation process is accomplished with full cooperation of patient & family
- Implementing modern ethical principles

During our professional life, we encounter with patients whose diagnosis is not confirmed. Sometimes patients are referred to departments of Rehabilitation Medicine with disability without sound diagnosis or even unknown diagnosis. We would like to share our experience with the readers of this journal.

CASE REPORTS

1. A 67 year-old woman, experienced a sudden (incomplete) paraplegia below D6 (ASIA C), with no pyrexia, but with anemia, hyperlipidemia and sphincters paralysis. MRI studies just left us with an enigma: a lesion which might be leptomeningitis due to lymphoma? Sarcoidosis? Tuberculosis? malignancy? Cortico-steroid therapy was started with an apparent improvement. We were reported by the patient that

previously, her dog developed the same clinical picture and died. EMG of the patient showed cervical and lumbar myelo-radiculopathy. Bence-Jones protein was not found. Later she developed pneumonia and urinary tract infection. 2nd MRI showed some improvement. No neurological changes had occurred later on. She remained wheel-chair bound and with sphincters' paralysis.

2. A 68 year-old man who suffered from: hypertension, transient ischemic attacks, gall-bladder stones, anemia, iodine sensitivity, nephrectomy (malignancy), resection of lung's metastasis, resection of prostatic carcinoma, resection of pheochromocytoma and now, newly collapsed vertebra with incomplete paraplegia below D7. Full investigation did not reveal the accurate diagnosis. Although metastatic vertebral lesion was suspected, we could not confirm it. He was discharged walking with a cane and partially controls his sphincters.

3. A 54 year-old man who developed acute paraparesis, fever, rash, (and suffered from chronic hypertension, hyperlipidemia, ischemic heart disease, diabetes mellitus). He had signs of diabetic neuropathy in the upper limbs. Skin biopsy showed: leukocytoclastic vasculitis. EMG showed axonal polyneuropathy. Radiological investigation showed D6/7 spondylotic changes and D7/8 disc protrusion. Marked improvement was evident after cortico-steroid therapy. No proof of autoimmune disease was found. He remained wheelchair- bound.

4. A 48 year-old woman who developed acute paraparesis below L1. She was found depressed, alcoholic, obese, s/p operation for stomach stricture, and poor nutritional status. EMG showed axonal polyneuropathy. She partially controlled her sphincters. Severe neuropathic legs' pain were relieved by anti-depressants and anti-epileptics. She remained confined to wheel-chair. Spinal stroke was suspected. MRI study could not confirmed it.

5. A 46 year-old woman who was operated in Russia 15 years ago (temporo-parietal lobectomy for seizures control). She suffered from tonic-clonic seizures, and mental distractions, and was treated by Phenobarbital. Now she was admitted in coma after a few days of fever: she was artificially ventilated, and treated with valproic acid and Tegretol. Upon gaining consciousness, she was found to have flaccid tetraparesis. EMG showed axonal polyneuropathy. High protein levels were found in her CSF. IVIG, and CS were started. Did she suffer from GBS? intensive care neuropathy (ICN)? She was referred to our rehabilitation ward: she was weaned from ventilator, fully conscious, distal 4 limbs hypoesthesia, tetraparesis, and pseudo-seizures. EEG showed normal pattern.

She gained full sphincters' control but developed joints' contractures. Oddly, later the muscles' tone turned to be spastic.

6. A 66 year old healthy woman was found unconscious. ER diagnoses were: diabetic keto-acidotic coma, respiratory insufficiency, UTI, staph sepsis, and atrial fibrillation. CSF study showed high protein level. Renal insufficiency was treated with hemodialysis. Upon gaining consciousness, she was found to have flaccid tetraparesis, bilateral ptosis, mild cognitive disturbances and distal hypoesthesia. Brain MRI was normal. EMG showed mixed polyneuropathy. GBS? IC-Neuropathy? CS therapy was given with mild clinical improvement. Five month later, cognitively she was found intact, but her muscles' tone turned to be spastic ! Eventually she was able to walk with a walker and partially dependent upon others in ADL.

The following two cases were described fully in this journal, suggesting a new syndrome [11]:

7. A 22 year-old woman who suffered from mild hypothyroidism, and positive hepatitis C serological test, consumed excessively alcohol and slept in unusual posture overnight. The next morning she was admitted with a clinical picture of rhabdomyolysis, anterior compartment syndrome, macrocytic anemia, thrombocytosis and flaccid paraparesis. EMG was found normal. Finally she was discharged with the same neurological picture, normal sphincters' control and she walked with two short leg braces and two crutches.

8. A 38 year-old man who is known to have a diet controlled DM, L5/S1 disc protrusion, hypothyroidism, fell asleep, while sitting/leaning, in front of his PC.

Upon waking up in the next morning he was rushed to the hospital and later was diagnosed as having: paraparesis, rhabdomyolysis, anterior compartment syndrome, and renal insufficiency. Hemodialysis was started. Sural nerve biopsy did not reveal any pathology. CSF was normal. He remained paraparetic. No autonomic nervous system involvement was noted. Metabolic complication? a combination of DM, hypothyroidism, sitting in a bizarre posture and muscles destruction due to bilateral compartment syndrome?

9. A 30 year-old homeless alcoholic man was found in a stupor. Initial diagnoses were: rhabdomyolysis, diarrhea, renal insufficiency, pulmonary emboli, staph viridens/pseudomonas sepsis, and hypertension. He was referred to a IC/dialysis unit. Upon gaining consciousness he was found tetraparetic and had no control over his sphincters. MRI showed central pontine myelinolysis (due to quick serum sodium level correction?) EMG showed severe axonal poly- neuropathy. GBS? ICN?. He was admitted to rehabilitation department with: cognitive disorders,

pressure sore, joints' contractures, spastic tetraparesis, impaired deep sensation below C5, and totally dependent in all ADL. Nine months later, he was discharged fully recovered!

10. A 33 year- old woman developed tetraplegia while being in a plasmapheresis [PP] treatment. She is known in the last five years to suffer from mild form of Gaucher disease, and essential monoclonal cryoglobulinemia. She was treated with CS, endoxan and marbthera. When mononeuritis multiplex was suspected, PP was re-started. During this last PP treatment, asymmetrical tetraplegia below C5 has appeared. Then, she developed perforation of small bowel and laparotomy was performed.

In the Rehabilitation department, she was found with incomplete tetraplegia below C5 on the right side and below C6 on the left. She gained some (assisted) ability to walk and gained control over her sphincters. She remained confined to her wheel-chair. Cryoglobulinemia with peripheral neuropathy in a patient with Gaucher disease who later developed Tetraplegia during PP treatment, is an unknown entity.

DISCUSSION

Special problems which arose during the rehabilitation process of these patients:

1. All cases suffered from non traumatic disabilities, and no financial compensation could be claimed, as is mandatory in road traffic, military or work-related accidents.
2. After the inpatient rehab-program is over, the national health and/or social security infrastructure gives actually little "rehab-maintenance " therapies and follow-up. The patients with disabilities of unclear etiologies, are expecting at least, a long-term treatment and follow-up.
3. These patients and their families were sometimes reluctant to fully cooperate during the rehabilitation process, facing uncertainty and confusion regarding diagnosis (and prognosis).
4. Absence of (substantial) financial support through social service networks, throw these disabled patients into a vicious circle of unemployment, social isolation and sometimes despair.
5. How much effort should be put in order to find out the correct diagnosis only for academic reasons?

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