Some aspects of management Autoimmune Encephalitis

G.Chakhava MD, PhD, Prof. Multiprofile Clinic Consilium Medulla, D.Tvildiani Medical University,President of Georgian Association of Medical Specialties ; M. Beridze MD, PhD, Prof. The First University Clinic of Tbilisi State Medical University; S. Mikiashvili, MD, Aversi Clinic ; M. Demuria MD, LTD Multiprofile Clinic Consilium Medulla, Georgian Association of Medical Specialties; I. Rukhadze MD, PhD Central Republic Clinic; N. Tatishvili Prof. D.Tvildiani Medical University, Head of Neuroscience Dep. Central Children Hospital, President of Georgian Association of Pediatric Neurology; N. Kvirkvelia MD, PhD, Prof. Ivane Javakhishvili Tbilisi State University, P. Sarajishvili Institute of Neurology

ABSTRACT

Several types of encephalitis are associated with antibodies against neural proteins. The presentation and the course of disease may be different. Paraneoplastic syndromes are combinations of symptoms and signs resulting from damage to organs and tissues distant from the site of a neoplasm and its metastases. Despite initial optimism, the interest to these syndromes diminished due to poor response to therapy, especially those with paraneoplastic cerebellar degeneration or limbic encephalitis. At the same time, antibodies against cell surface antigens (NMDA and LG1 antibodies) are associated with better prognosis and response to therapy and less likely to be associated with cancer. We are presenting several case reports including clinical phenotypes, differential diagnosis and treatment options. We also reviewed detailed clinical profiles and laboratory findings of 3 cases in children with probable Bickerstaff's Brainstem Encephalitis (BBE).

PATIENTS AND CLINICAL DATA

Patients and clinical data

Patient1.(22276)

F. 16 years old.

Patient has developed headache and temperature since 29.11.13, accompanied by imbalance and gait disturbance and walking immobility. Strabismus accompanied by total opthalmoplegia. Double vision, slureed, scanning speech with nasal voice. Dysphagia,whitout nasal regurgitation. Flaccid paraplegia in lower extremities. 2.12.13 patient was admitted to the Hospital of infectious diseases, with the diagnosis of inflammatory polyradiculoneuropathy and the treatment was conducted by Solu-Medrol (methylprednisolone sodium succinate), Ceftriaxone , Zovirax (acyclovir) and symptomathic therapy.

Laboratory studies : CSF protein-0,33% , albuminocytological dissociation was not found, Needle electromyography (N Tibialis, N Peroneus L and R) defined motor axonal degeneration. CBC , urine and stool tests were normal. EKG and 24 h. Holter – normal.Serology – Enterovirus IgG 200U/ml. MRI detected no abnormalities.

Treatment : immunoglobulin IV (2,0 kg),Atarax 25 mg, physical therapy

Patient 2. (21485)

F. 5 years old

8.11.2013 patient came from school with headache. In the evening pain developed in the lower extremities, weakness, fell down while walking. 9.11.2013 pain started in the morning in both eyes,photophoby, pain of masticatory muscles during eating. Dysphagia when eating solid food, slurred nasal speech were present. 11.11.2013 she was admitted to Kutaisi hospital, where she spent 12 days with the diagnosis of bacterial meningitis. Conducted treatment: Vancover (vancomycin), Garde (meropenem), methylpred . 29.11. 2013 she was admitted to our hospital with bilateral facial nerve peripheral paresis, adynamia, without conscious disturbance.

Laboratory studies : MRI detected no abnormalities, CSF protein –0,066%, cytosis – 3. Needle electromyography (N. facialis , N. medianus, N peroneal, N tibialis)) defined axonal degeneration. CBC, EEG, electrolytes, glucose – normal.

Treatment : methylpred, Rocephin (ceftriaxone).

Table 1.1 - clinical signs and symptoms

#/ clinical manifestation	Patient 1.	Patient 2.	Patient 3.
Opthalmoplegia	+	-	-
Gait disturbance/ walking difficulties	+	+	+
areflexia	-	-	+
Limb weakness and pain	-	+	+
Flaccid tetraplegia	-	-	+
Flaccid paraplegia	+	-	-
Dysphagia	+	+	-
Slurred/ nasal speech	+	+	-
Strabismus	+	-	-
Double vision	+	-	-
Photophoby	-	+	-
Headache	+	+	-

Patient 3. (1088)

F. 13 years old

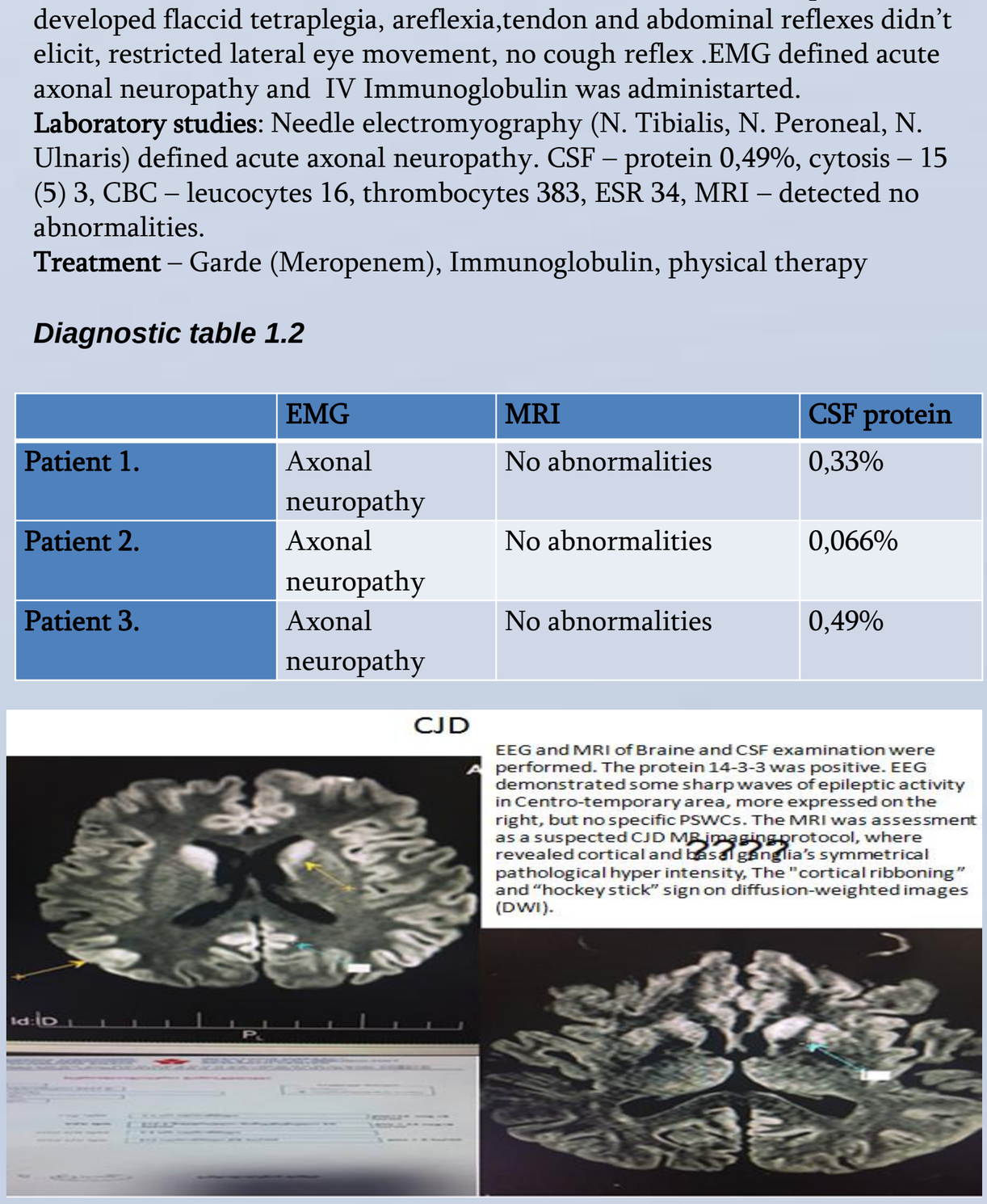
Symptoms started with numbness, pain in lower extremities, walking difficulties, this condition worsened rapidly, low back pain developed, movement in upper extremities worsened. With these symptoms the patient was admitted to Batumi Hospital.Disease progressed rapidly and because of respiratory insuficciency she was admitted to ICU department at our hospital in 19.01.2013 where mechanical ventilation was started. In ICU patient developed flaccid tetraplegia, areflexia,tendon and abdominal reflexes didn't elicit, restricted lateral eye movement, no cough reflex .EMG defined acute axonal neuropathy and IV Immunoglobulin was administarted.

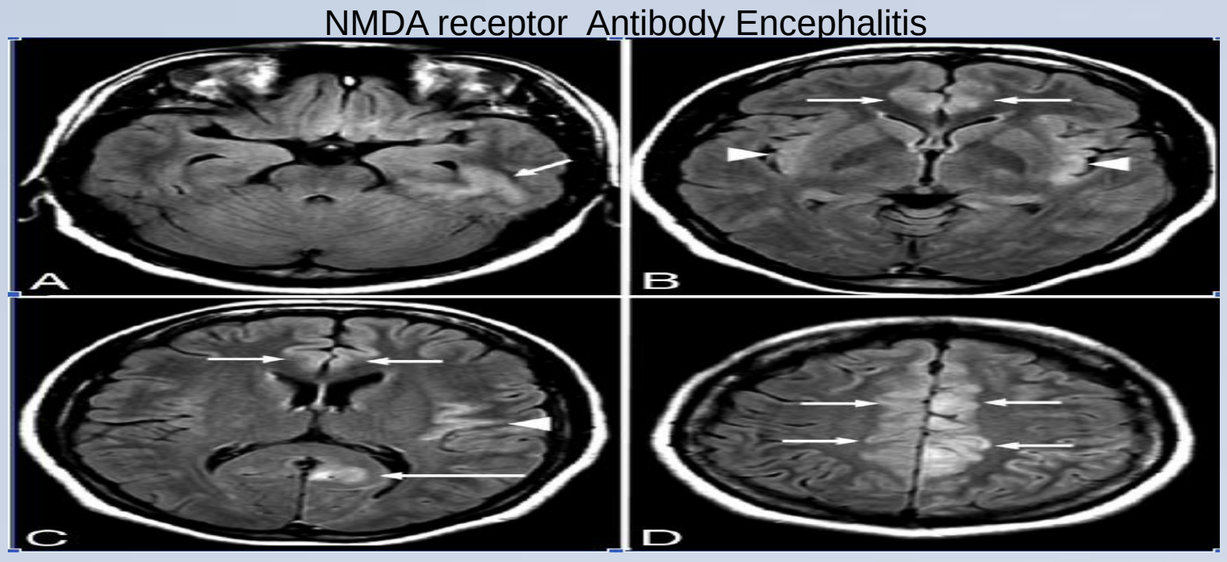
Laboratory studies: Needle electromyography (N. Tibialis, N. Peroneal, N. Ulnaris) defined acute axonal neuropathy. CSF – protein 0,49%, cytosis – 15 (5) 3, CBC – leucocytes 16, thrombocytes 383, ESR 34, MRI – detected no abnormalities.

Treatment – Garde (Meropenem), Immunoglobulin, physical therapy

Diagnostic table 1.2

	EMG	MRI	CSF protein
Patient 1.	Axonal neuropathy	No abnormalities	0,33%
Patient 2.	Axonal neuropathy	No abnormalities	0,066%
Patient 3.	Axonal neuropathy	No abnormalities	0,49%





CONCLUSIONS

There are an increasing number of immunocompromised patients with potentially atypical presentation of infectious encephalitis.

There is a higher likelihood of detection of "exotic" infections in Western countries due to increased international travel.

Testing of antibodies' panel is recommended in patients with encephalitis of unknown etiology. It is important to know caveats and link clinical phenotypes to antibody panel testing results.

The field of autoimmune neurology has broadened beyond neurology and psychiatry, intensive care physicians and pediatricians should be alert to the illnesses of this spectrum.

Differentiation among infectious, antibody-mediated, and other types of encephalitis is important for differentiated treatment.

Prospective population-based studies are needed to evaluate the impact of different immunotherapy in autoimmune encephalitis and to standardize the different diagnostic tests in order to improve the management of these complex disorders.

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