Clinical Pathologic Conference

Name of the patient: 林XX Presented by：兒童腦神經內科 藍士耘醫師
Chart Number: 1033**** Discussed by：兒童腦神經內科 林光麟醫師
兒童過敏氣喘風濕科 李文益 醫師
Date of admission: 2014/11/14 兒童放射診斷科 勞允澤 醫師
兒童外科 明永青 醫師
婦產科 邱健泰 醫師
解剖病理科 薛 純 醫師
Moderator：解剖病理科 孫建峰 醫師

Chief Complaint: acute onset of personality change for 5 days

Present Illness:

This 13-year-1-month-old girl with appropriate development and regular vaccination had no previous underlying disease. She had developed acute onset of agitation, disorientation and self-talking since 5 days before admission. There was no fever, headache, seizure, recent infection or trauma history. She was brought to local hospital and admitted to psychiatric ward. The initial impression was acute psychosis. Antipsychotics were applied including Olanzapine, Lorazepam, Lendormine and Biperiden. However, intermittent crying and screaming during sleep was noted. In addition, conscious disturbance, drooling, dysphagia, urinary retention and involuntary movement developed. There was also facial and mouth twitching. Extrapyramidal symptom due to antipsychotics was impressed. Due to severe side effect, the family refused to take medications, and she was discharged against medical advice. Then she was transferred to our hospital.

By the way, while emphasized by her mother, the girl was stressed out by the overwhelming tests at school and homework after 7th grade. Her school grades regressed and she was scolded by the teacher.

Past History:

- Admission history: before this admission, in psychiatric ward for 3 days under the impression of acute psychosis
- Operation history: hernia
- Medication history: denied
- Chronic Illness: denied

Personal History:

- Birth history: G2P2A0, term baby via NSD, no perinatal insult
- Newborn screen: no abnormal finding
- Growth: 157cm (50th to 75th percentile), 45kg (25th to 50th percentile)
- Development: as milestones
- Vaccination: as scheduled
- Travel history: denied
- Allergy: denied

Family History: No hereditary disease in her family

Physical Exams: T: 37.7’c P: 108bpm R:20/min BP: 128/78 mmHg

General Appearance: acute ill-looking

Consciousness: E4V1M5

HEENT: sclera: not icteric; conjunctiva: not injected; throat: not injected; no oral ulcer

Neck: supple without LAP
Chest: Smooth breathing pattern, clear and symmetric breathing sound
Abdomen: soft and mild distended; dullness on percussion; no palpable mass
Extremities: involuntary movement; symmetric peripheral pulse
Skin: no rash

Neurologic exams: Pupil: 3+/3+; EOM: full/full; Gag reflex: +; DTR: ++/++; Babinski sign: plantar flexion/plantar flexion; others could not performed

Lab Exam: (at admission)

WBC: 12.9K/uL	AST: 19 U/L	Hb: 14.7 g/dL
Ca: 9.1 mg/dL	Hct: 43.7 %	Na: 145 mEq/L
MCV: 84.7 fL	K: 3.8 mEq/L	Plt: 313K/uL
Cl: 111 mEq/L	Seg: 85.4 %	CRP: 4.7 mg/L
Lym: 8.4 %		

Course and Treatment:

After admission, symptoms deteriorated. Abnormal breathing pattern was noted on the next morning. Thus she was transferred to PICU within 24 hours. Under the initial impression of encephalitis, we performed blood culture, CSF culture, serial viral serology and CSF work-up. Vancomycin, claforan, oseltamivir, acyclovir and doxycycline were all applied. Status epilepticus was impressed Conscious disturbance, persistent facial and oral twitching were observed. Conventional antiepileptic drugs including luminal and continuous midazolam were prescribed for suspicion of status epilepticus. Intubation was performed to protect the airway. IVIG and methylprednisolone pulse therapy were also applied.

Later, anti-NMDA receptor Ab was positive both in serum and CSF. Abdominal sonography on 4th day in PICU showed pelvic mass favoring teratoma. Teratoma with rupture was impressed on abdominal CT. 2 days later, laparoscopic salphingo-oophorectomy was performed. Final pathology was immature teratoma, grade 3. Ascites was also positive for NMDAR Ab.

1st cycles of chemotherapy, JEB for malignant germ cell tumor was completed on 2014/11/27-29. Further serum NMDAR Ab was positive persistently. The plasma exchange was performed on 2014/12/16, 18, 20, 22 and 24 smoothly. Orofacial dyskinesia improved, and seizure frequency also decreased step by step. 2nd cycle of JEB was completed on 2014/12/25-27. Elective tracheostomy was done on 2014/12/19 due to extubation failure.

NMDAR Ab waS negative in serum after plasma exchange. The patient was back to ordinary ward on 2015/01/03. Currently, infection control and neurological condition monitoring are critical problems. The patient is under linezolid for VISA sepsis, and under amikacin and fluconazole for E.coli plus fungal UTI. Current conventional anti-epileptic drugs include luminal and clonazepam. She is still in confusion and intermittent irritability..

Point of Discussion:

1. Manifestation of acute psychosis and neurological symptom in anti-NMDA receptor encephalitis
2. Association of teratoma with anti-NMDAR encephalitis
3. Treatment and prognosis of anti-NMDAR encephalitis